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With the valuable participation of many who believed in this project, we are pleased to deliver to our readers the 10th issue of our Scientific Journal CEREM Goiás, already considered one of the most important and respected medical-scientific publications in Goiás and Brazil. We remain firm and determined to contribute more and more to the serious and transparent dissemination of the scientific work carried out by resident physicians and medical residency services in our region.

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RESPIRATORY FAILURE AFTER ACCIDENTAL PHRENIC NERVE BLOCK IN REGIONAL ANESTHESIA OF THE BRACHIAL PLEXUS VIA INTERSCALENE

ISABELA ALCÂNTARA ROCHA[1]; DAVYD FONSECA[1]; LARISSA MANZAN DE ALCÂNTARA BORGES[1]; MATEUS FERREIRA DE SIQUEIRA E SILVA[1]; GUSTAVO SIQUEIRA ELMIRO[1,2]; GIULLIANO GARDENGHI[1,2,3,4]

ABSTRACT

Objective: To describe a form of ventilatory complication resulting from regional anesthesia of the brachial plexus, accidental blockade of the phrenic nerve and its consequences on the respiratory system. Methodology: This is a descriptive study, in the format of a case report, carried out in Goiânia-GO. Data were collected through analysis of electronic medical records. The collected information was discovered with the pre-existing literature through the search of correlated articles in the PubMed/Medline and SCIELO database. Case report: A 42-year-old patient with severe sequelae and total limitation of movement of him right arm because of a car accident, attended for care of the left rotator cuff via arthroscopy, received general balanced anesthesia and regional anesthesia guided by ultrasound via interscalene and who in the post-anesthesia recovery room evolved with acute respiratory failure as a result of the right frantic nerve block. The patient was maintained on ventilatory support until the end of the effect of regional anesthesia and recovery of respiratory function. Conclusion: The patient reported here presented significant ventilatory dysfunction and diaphragmatic paralysis, due to possible involvement of the phenic nerve, after performing an interscalene block for arthroscopic surgery of the brachial plexus. Thus, he could have benefited from a supraclavicular blockade, which provides satisfactory analgesia and less chance of blocking the phrenic nerve, or in the case of maintaining the choice for the interscalene route, the blockade could have been performed with the lowest volume and concentration of local anesthetics.

KEYWORDS: ANESTHESIA, CONDUCTION; ANESTHETICS, LOCAL; ANESTHESIA RECOVERY PERIOD; PHRENIC NERVE; RESPIRATORY INSUFFICIENCY; POSTOPERATIVE COMPLICATIONS.

INTRODUCTION

Peripheral nerve blocks have seen a great resurgence of interest in the last decade, especially with the advent of ultrasound. Nerve blocks evolved from an art that only a few physicians could master to something with more objective results and transferable skill, due in large part, as already mentioned, to the introduction of ultrasound guidance. Peripheral nerve blocks are today a main component of perioperative multimodal analgesia [1,2]. In particular, for upper limb surgery, brachial plexus blocks (interscalene, supraclavicular, infraclavicular, and axillary approaches) have been consistently associated with anesthesia, with better time efficiency, faster recovery, fewer adverse events, better analgesia, and greater acceptance. of the patient [3-4].

Interscalene brachial plexus block is well established in shoulder surgeries, as it offers adequate postoperative anesthesia and analgesia. Traditionally, it provides a significant postoperative analgesic benefit. However, when used as a single-dose approach, it is limited by its duration [5], not lasting longer than 24 hours, even when using longer-acting local anesthetics such as bupivacaine [6].

In the context of shoulder surgery, interscalene nerve block is the most commonly used regional anesthetic technique [7,8], however, unintentional phrenic nerve block may be associated with this route of regional anesthesia. The blocked phrenic nerve generates ipsilateral diaphragmatic paralysis, which is temporary and usually asymptomatic in healthy patients. However, patients with limited ventilatory function or a disorder of the contralateral phrenic nerve may have severe respiratory symptoms.

The present study presents the case report of a patient who developed severe acute respiratory failure in the immediate postoperative period of shoulder surgery, and through this report we sought to explore, based on the pre-existing literature, forms of treatment and prophylaxis for this condition.

CASE REPORT

Male patient, 42 years old, ASA II, controlled hyperten-

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sion, smoker, with severe sequelae and total limitation of right arm movement resulting from brachial plexus injury after an automobile accident in 2002, with rotator cuff syndrome and chronic shoulder pain left due to loosening of the synthesis material installed for the treatment of a fracture of the left proximal humerus in 2018.

Having verified the loosening of the internal fixation plate of the left humerus with maintenance of fracture alignment, the orthopedics team indicated surgical treatment for removal of the material and repair of the rotator cuff, which surgery took place in the morning on March 9, 2022.

In the operating room, the patient was properly monitored with a pulse oximeter with plethysmographic curve, cardioscopy and non-invasive blood pressure, with stable vital signs before anesthetic induction and received venipuncture with a 20G needle in the right upper limb.

The patient was submitted to balanced general anesthesia: Pre-oxygenation under facial mask with oxygen at 100% 6L/min for 3 minutes, for venous anesthetic induction, 30mcg of sufentanil and 200mg of propofol were administered and after the loss of consciousness was verified, they were administered 50mg of rocuronium, the patient received manual ventilation until complete muscle relaxation and then orotracheal intubation was performed with an endotracheal tube in volume-controlled mode, with protective parameters, with fraction of inspired oxygen (FiO2) of 50%. Maintenance of general inhalational anesthesia was established by administration of sevoflurane at a concentration of 2% in low flows of fresh gases 2L/min in the anesthesia machine.

For regional anesthesia, the patient was positioned with the head elevated at 30° and the head lateralized to the right and after asepsis of the region with alcoholic chlorhexidine, an ultrasound-guided puncture was performed with abocath 18g to block the left brachial plexus via interscalene, the path of the needle was completely visible at all times, aspirations were performed before the infusions without blood return, a total of 20ml of an anesthetic solution consisting of 10ml of 2% lidocaine with 1:200,000 epinephrine were infused through the region and 10ml of levobupivacaine 0.5% with epinephrine 1:200,000 totaling 200mg of lidocaine, 50mg of levobupivacaine and 100mcg of epinephrine.

The intraoperative period was uneventful and the patient maintained stable vital signs throughout the period. At the end of the surgery, which lasted two hours, the patient was decurarized with 2mg of neostigmine and 1mg of atropine, and when adequate ventilatory parameters were reached in spontaneous breathing, extubation was performed. Then, the patient was taken to the post-anesthesia care unit (PACU), awake, conscious, without complaints, using a nasal catheter with oxygen at 2L/min, eupneic and with peripheral oxygen saturation of 99% before leaving the operating room.

In the PACU, the patient evolved with acute respiratory failure, with progressive dyspnea and desaturation, verified by pulse oximetry, in the amount of 56% on the monitor. The assistant anesthesia team opted for venous induction in rapid sequence intubation with 200mg of propofol, 100mcg of fentanyl and 100mg of rocuronium. After orotracheal reintubation with a 7.5 tube, mechanical ventilation started in volume-controlled mode, with protective parameters and with an inspired fraction of oxygen of 60%, there was a progressive improvement in peripheral oxygen saturation, returning to the value of 99% indicated on the monitor.

With the stabilization of the clinical picture, the patient was taken to the computed tomography of the hospital, for diagnostic purposes. Chest angiotomography with contrast and chest X-ray were performed, represented by figure 01, below:

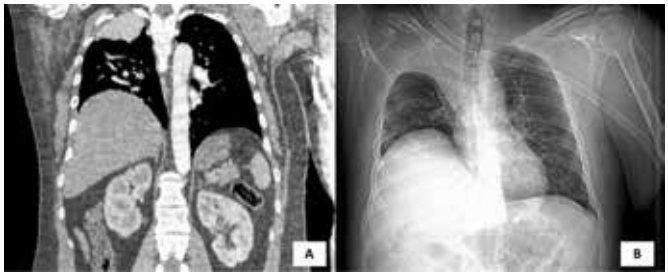

Figure 01. Images of the patient. A. CT angiography of the chest showing elevation of the right diaphragmatic dome and almost complete atelectasis of the upper lobe of the right lung. B. Chest X-ray showing the same CT findings

Chest angiotomography (Figure 01.A) showed elevation of the right diaphragmatic dome and almost complete atelectasis of the upper lobe of the right lung, in addition to atrophy with marked liposubstitution of the muscles of the shoulder girdle and right chest wall, findings compatible with the chronic lesion of ipsilateral brachial plexus. There was also the presence of bilateral pulmonary atelectatic opacities, predominating in its posterior portions; the other pulmonary fields did not present alterations. Signs of pulmonary thromboembolism were ruled out, there were no perfusion failures in the pulmonary trunk, in the main pulmonary arteries or in their lobar and segmental branches.

Thus, the main diagnostic hypothesis for the picture of acute respiratory failure presented by the patient in the PACU was accidental blockade of the left phrenic nerve after regional anesthesia of the brachial plexus via interscalene, leading to ipsilateral diaphragmatic paralysis, which caused respiratory fatigue that was not compensated because of the pre-existing contralateral pulmonary dysfunction and morphofunctional sequelae resulting from complete and chronic injury to the brachial plexus and right phrenic nerve.

The patient was transferred to the intensive care unit, where he remained monitored, under ventilatory support

and clinically stable. In the late afternoon of the same day, the patient recovered respiratory function with the end of the effect of the local anesthetics used in regional anesthesia and was successfully extubated, without presenting new episodes of dyspnea or desaturation, maintaining peripheral oxygen saturation of 95% in use of supplemental oxygen 2L/min through a nasal catheter. During the 48 hours after extubation, treatment was performed with respiratory physiotherapy and intermittent non-invasive ventilation with the aim of lung re-expansion. The patient had a good evolution of respiratory function with progressive improvement in oximetry, without the need for supplemental oxygen, maintaining a peripheral saturation of 99% on room air, and was discharged from the ICU to his home on March 12, 2022.

DISCUSSION

The brachial plexus is a nerve network that supplies the upper extremity of the human body, formed by nerve roots from C5 to T1. It emerges from the cervical spine and travels between the anterior and middle scalene muscles and distally around the axillary artery. Interscalene brachial plexus block is a common practice for procedures on the distal clavicle, shoulder and proximal humerus, as it ensures excellent anesthetic coverage and postoperative analgesia. The space between the scalene muscles is called the interscalene groove. This space is palpable behind the lateral head of the sternocleidomastoid muscle and adjacent to the lateral tubercle of C6, also known as the Chaissaignac's tubercle [9]. In fact, interscalene brachial plexus block is the most used postoperative analgesic technique in shoulder surgery. In addition to postoperative analgesia, it reduces pain scores and opioid consumption. However, it has disadvantages and contraindications, including short duration of analgesia, rebound pain, high incidence of unilateral diaphragmatic paresis, and potential risk of nerve damage when targeting nerve roots in the neck rather than peripheral nerves [10].

The interscalene block covers most of the brachial plexus, sparing the ulnar nerve (C8-T1). The interscalene space is identified by palpation or ultrasound visualization. Under ultrasound visualization, the brachial plexus can typically be seen as 2 or 3 hollow circles ("traffic lights") that correspond to the upper, middle, and lower trunks. The lower trunk can sometimes be difficult to visualize as the muscle thickens. Once visualized, injection of a long-acting local anesthetic can block nerve impulses and cause upper extremity numbness and weakness. Structures immediately distal to nerve block placement consistently block nerve impulses and cause sensory and movement loss [9].

A nerve stimulator can also be used as an adjunct to confirm placement. The nerve stimulator causes muscle contractions in the deltoid muscle, arm or forearm when the corresponding nerve is stimulated. A volume of local anesthetic is injected, usually between 15 and 25mL. Commonly used local anesthetics include bupivacaine and ropivacaine. After injection of the anesthetic, the patient presents pain relief and a feeling of heaviness in the limbs during the action of the local anesthetic [9].

Among the complications of this blockade route are described: postoperative neurological symptoms, vascular complications (bruises, intravascular injections leading to systemic intoxication by local anesthetics), respiratory complications (pneumothorax), epidural or subarachnoid injection, undesirable blocks (Horner syndrome generating miosis, palpebral ptosis and anhidrosis when blocking the cervical sympathetic chain, dysphonia due to vocal fold paralysis when blocking the recurrent laryngeal nerve and alteration of the diaphragmatic function when blocking the phrenic nerve). This block is contraindicated in patients with respiratory failure due to the high probability of ipsilateral phrenic nerve block and diaphragmatic hemiparesis. This can lead to a 25% reduction in lung function. Due to unilateral diaphragmatic paresis, respiratory mechanics can be considerably impaired [9,11]. The phrenic nerve arises from the C3-C5 nerve roots of the deep cervical plexus, passes over the anterior surface of the anterior scalene muscle, and descends into the thoracic cavity, providing motor innervation to the ipsilateral hemidiaphragm. When performing an interscalene block, due to its proximity to the target nerve, the phrenic nerve is typically blocked inadvertently by ventral spread of the local anesthetic. The same may occur, however, with lower incidence, in subclavian perivascular blocks [12].

In healthy adult patients transient unilateral diaphragmatic paralysis is tolerated and generally asymptomatic. It is possible that the accessory muscles compensate for the restriction imposed by the paralysis and the expansion of the contralateral lung manage to produce enough negative pressure to guarantee good ventilation [13]. Based on studies, it can be assumed that patients with ASA II (less than or equal) without pre-existing pulmonary disease are not clinically impaired by a phrenic nerve block induced by an interscalene block, with subsequent hemidiaphragmatic paresis. Despite the paresis, they can be transferred directly from the operating room to the general care unit, as long as they have an Aldrete and Kroulik score of 10 (a scale used to assess the post-anesthetic recovery of patients undergoing anesthesia) when leaving the operating room [11]. However, severe respiratory symptoms may appear in patients with limited pulmonary reserve or previous pulmonary dysfunctions (obese, asthmatic, patients with chronic obstructive pulmonary disease, elderly) and the blockade is contraindicated in the presence of deficit of the contralateral phrenic nerve, since blocking the functioning phrenic nerve may result in severe respiratory distress or respiratory arrest. [14]. In cases of bilateral shoulder surgeries, due to the risk of total diaphragmatic paralysis, bilateral interscalene blocks

are not recommended [15].

Due to the hypothesis that the phrenic nerve block occurs by dispersion of the local anesthetic to the anterior region of the anterior scalene, it is presumed that by reducing the volume and concentration of the anesthetic solution used in the interscalene block, there is a limitation of the amount of local anesthetic capable of reaching the region of the phrenic nerve, thus reducing side effects of respiratory function. Ultrasound guidance allows using a smaller volume of local anesthetic and depositing it more precisely, leading to a reduction in the incidence of hemidiaphragmatic paralysis and other side effects, as the trajectory of the needle can be tracked and controlled in real time, minimizing the risk of trauma to critical structures including nerves, vessels and pleura [16].

Riazi et al compared the efficacy and respiratory consequences of ultrasound-guided interscalene brachial plexus blocks using 5ml and 20ml of 0.5% ropivacaine. The incidence of diaphragmatic paralysis due to phrenic nerve block was 45% in the low volume group and 100% in the 20ml group. There were no significant differences in pain scores and opioid consumption 24 hours after surgery [17]. Renes et al determined that the minimum effective volume of 0.75% ropivacaine for shoulder analgesia in ultrasound-guided C7 root block was 2.9 ml in 50% and 3.6 ml in 95% of the patients studied. The lung function of this sample remained unchanged up to two hours after the completion of the surgery [18]. Gautier et al showed that the minimum effective anesthetic volume of 0.75% ropivacaine for adequate surgical analgesia for shoulder arthroscopy with interscalene brachial plexus block was only 5 ml or 1.7 ml for each of the upper, middle and lower trunks of the brachial plexus [19]. Such evidence suggests that the use of low-volume local anesthetics for the interscalene block provides adequate analgesia for shoulder surgeries and at the same time reduces the incidence of phrenic nerve block and its repercussions on the respiratory system.

Bergmann et al performed a randomized clinical trial of 84 patients scheduled for elective shoulder surgery, divided into two groups to receive an ultrasound-guided approach to interscalene brachial plexus block via the anterior (n = 42) or posterior (n = 42) approaches. Both groups received 15 ml of 1% ropivacaine. Spirometry was performed at baseline and 30 min after blockade [11]. Spirometric results showed a significant decrease in vital capacity, forced expiratory volume in the first second and maximum nasal inspiratory capacity after interscalene brachial plexus block; indicating a phrenic nerve block. Despite changes in spirometry, no case of dyspnea was reported. The authors mention in their discussion that the respiratory/clinical effects of phrenic nerve blocks are usually well compensated. It was not possible to identify a significant difference in the impairment of spirometric parameters between the group that used the anterior route and the group using the posterior route [11].

Since the phrenic nerve block was considered an inevitable consequence of the interscalene block [20], comparative studies have appeared in relation to the effectiveness of the supraclavicular block of the brachial plexus in substitution of the interscalene block, for shoulder surgeries, in order to reduce the incidence of diaphragmatic paralysis, since there is less occurrence of this complication in supraclavicular blocks. Hussain et al showed that interscalene block offers an analgesic advantage limited only to the immediate postoperative period (post-anesthetic recovery room) and that supraclavicular and interscalene blocks did not present statistical differences in pain severity and opioid consumption in the first 24 hours after shoulder surgery. Given the above data, patients with a risk factor for respiratory failure with a phrenic nerve block may benefit from choosing the supraclavicular approach for brachial plexus block [20].

A review carried out by Kang and Ko in 2023 corroborates the benefit of the brachial plexus block via the supraclavicular route, which can be considered an effective and safe alternative to shoulder surgery, especially in patients with preexisting pulmonary impairment. When performing supraclavicular brachial plexus block, the incidence of hemidiaphragmatic paresis was effectively reduced when local anesthetic was injected primarily into the corner pouch (20 ml) and secondarily into the neural cluster (5 ml) during right-sided supraclavicular brachial plexus block. Other alternatives to avoid phrenic nerve block, suggested in this review, were extrafascial injection for interscalene blocks, with the potential to reduce the incidence of hemidiaphragmatic paresis and consequently preserve lung function, while providing analgesia similar to a conventional intrafascial injection. Furthermore, this may reduce the potential for neurological injury inherent to the interscalene brachial plexus block [10].

Another alternative is the injection around the upper trunk of the brachial plexus. The upper trunk is formed by the fusion of the C5 and C6 nerve roots. Therefore, local anesthetic injection around the upper trunk should produce similar analgesia in the shoulder, because the main terminal nerves innervating the shoulder arise distal to the upper trunk. Furthermore, the injection site is farther from the phrenic nerve, which theoretically reduces the risk of hemidiaphragmatic paresis. Studies have shown similar pain scores, duration of analgesia and opioid consumption in 24 hours, and less frequent hemidiaphragmatic paralysis in the upper trunk block group [10]. One more strategy to reduce the risk of hemidiaphragmatic paresis would be to inject local anesthetic into the terminal nerves of the shoulder, more distally to the upper trunk. A shoulder block is an alternative approach that blocks the suprascapular and axillary nerves. These two nerves innervate most of the shoulder, with additional minor contributions from the

subscapular and lateral pectoral nerves. Suprascapular and axillary nerve blocks reduced the incidence of hemidiaphragmatic paresis and pulmonary dysfunction while providing similar postoperative analgesia [10].

Addition of liposomal bupivacaine is a viable option without refining the interscalene brachial plexus block technique. The addition of liposomal bupivacaine to bupivacaine resulted in statistically significant reductions in diaphragmatic excursion and tested lung function 24h after block placement compared with bupivacaine alone. However, this reduction was within the range of normal diaphragmatic function [10].

The respiratory limitation imposed by the phrenic nerve block is a self-limiting event, therefore, the treatment consists of providing ventilatory support until the patient recovers respiratory function with the end of the action of the local anesthetic in the phrenic nerve region. There are reports of cases [21,22] of reversal of the phrenic nerve block with recovery of pulmonary function after the infusion of saline solution in the interscalene region, but the efficacy of this technique needs more consistent studies for its recommendation. The use of saline solution was also studied by Srinivasan et al as a form of prophylaxis for unwanted blockage of the phrenic nerve. Their study demonstrated a 50% reduction in the incidence of hemidiaphragmatic paralysis when filling the region anterior to the anterior scalene muscle with 10ml of saline solution before proceeding with the interscalene blockade [23]. The mechanisms underlying recovery from blockade or prophylaxis for phrenic nerve involvement are still not well understood, saline boluses can cause a dilution effect, a local pH reduction, alteration of the local sodium content or even a placebo effect [24], interfering with the effectiveness of the effect of the local anesthetic in the region, allowing the phrenic nerve to recover more quickly.

CONCLUSION

The interscalene brachial plexus block may have phrenic nerve block as a frequent complication. In populations with risk factors for respiratory failure, the choice of technique and blockade route should be a cautious decision. Given the above, the patient in the case report could have benefited from a supraclavicular block that provides similar analgesia and lower risk of involving the phrenic nerve. He could also, in case the interscalene route was chosen, have received a smaller volume of local anesthetics, in lower concentrations, with ultrasound help in order to precisely determine the injection site. Studies are still needed regarding prophylaxis of phrenic nerve involvement and reversal of unwanted blocks with the use of saline solution.

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HEMODYNAMIC TREATMENT OF SEVERE LESION OF THE LEFT MAIN CORONARY ARTERY: A CASE REPORT

THAÍS BASTOS ROCHA

ABSTRACT

Compared to other cardiac artery stenosis, the left main coronary artery stenosis is associated with a higher risk of mortality and myocardial injury due to the greater amount of subtended myocardium. For treatment, myocardial revascularization surgery and percutaneous coronary intervention are indicated. Therefore, this work has as main objective to present the benefits of the treatment of a lesion of the main coronary artery by hemodynamic approach and to report a case of a severe lesion treatment of the left main coronary artery. This is a case report and an integrative literature review. The search was carried out in the PubMed and Scielo libraries. The works that were included discuss the theme proposed for the study in Portuguese and English, with full texts and available in free versions. In conclusion, for patients with stenosis of the main coronary artery with low and medium complexity of coexisting coronary artery disease, PCI treatment offers a favorable long-term result and, therefore, it constitutes an alternative therapy to MRS. While complex disease is best treated with MRS. For evaluation of the angiographic SYNTAX score, it can be used to indicate the best treatment tool.

KEYWORDS: CORONARY TRUNK LESION; TREATMENT; HEMODYNAMIC WAY; REVASCULARIZATION; ALTERNATIVE THERAPY.

INTRODUCTION

The coronary trunk (CT) originates from the aortic sinus, passing behind the pulmonary trunk. Generally, the path is horizontal or slightly caudocranial, dividing itself into the anterior descending artery (ADA) and the circumflex artery (CXA). Occasionally, the CT ends in a trifurcation, thus initiating the diagonalis branch, which runs laterally to the ADA 1. Compared to other sites, CT stenosis is associated with an increased risk of mortality and myocardial injury due to the greater amount of subtended myocardium2.

For treatment, myocardial revascularization surgery (MRS) was introduced in 1968, becoming the standard for the treatment of symptomatic patients with coronary artery disease. With advances in the area over the years, providing smaller incisions, greater myocardial preservation, use of arterial conduits and better postoperative care, it was possible to reduce morbidity, mortality and graft occlusion rates. Another available technique is percutaneous coronary intervention (PCI), which was introduced in 1977. Through this intervention and with current high technology, it has become possible to treat complex lesions safely and efficiently3.

Thus, efforts are increasingly being made to provide scientific advances in the area so that the patient has access to cutting-edge therapies. Therefore, studies on the subject make this progress possible. Presenting the benefits of the treatment of coronary trunk lesions by hemodynamic way is the objective of this work through the conceptualization of coronary trunk lesions, highlighting the differences between treatments, as well as pointing out the indications for treatment of coronary trunk lesions by hemodynamic way.

METHODS

A qualitative narrative review study was carried out. The search was carried out in the virtual libraries of the United States National Library of Medicine (PubMed) and Scientific Electronic Library Online (Scielo). The keywords used were: coronary trunk lesion; treatment; hemodynamic way, isolated or associated through the Boolean AND operator.

The articles were selected from a previous reading of the abstracts in order to compare the respective points proposed, used and discussed by each author. In cases where reading the abstracts was not enough to understand the context, the full article was accessed. Subsequently, a selective reading of the articles was carried out to organize the information found, an analytical reading to highlight the most relevant themes and topics from a selection of information that will interest the research in general.

The inclusion criteria were: works that discussed the theme proposed for the study in Portuguese and English, with complete texts and available in free versions.

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The exclusion criteria were: works that did not contemplate the proposed objective of the research; that did not adhere to the research area and that were unavailable at the time of collection and that, therefore, would not be relevant for this study.

CASE REPORT

Patient N.P.S.A., female, 65 years old, previously suffering from hypertension, insulin-dependent type 2 diabetes, chronic stable angina (functional class 2) and previous history of breast cancer with right mastectomy 27 years ago. Admitted in November 2021 due to an episode of stabbing retrosternal chest pain radiating to the left upper limb, prolonged at rest, intensity 8/10, associated with dysautonomia. Upon admission, a twelve-lead electrocardiogram and myocardial necrosis markers were evaluated, both negative for ischemia. A diagnosis of unstable angina was made and an invasive coronary risk assessment was performed through cardiac catheterization.

Previous use medications: Sustrate 20 mg/day, Clopidogrel 75 mg/day, Atenolol 100 mg/day; Nifedipine Retard 20 mg/day, Pitavastatin 2 mg/day, Gligafe XR 2000 mg/day and NPH Insulin 30 U/day. Despite the previous use of antianginal drugs, the patient reported having recurrent angina.

On November 19, 2021, cardiac catheterization was performed, which showed: Right Coronary Artery: Dominant, without obstructive lesions (Figure 1); Posterior Ventricular and Posterior Descending: without obstructive lesions. Left main coronary artery: 90% severe ostial lesion followed by aneurysm in the distal third (Figure 2). Anterior Descending Artery: without obstructive lesions; Circumflex artery: ostial occlusion. On ventriculography: Left ventricle with slightly increased volumes, diffuse hypokinesis, no left ventricle-aorta gradient, mitral valve didn't not allow reflux into the left atrium. Moderate left ventricular systolic dysfunction.

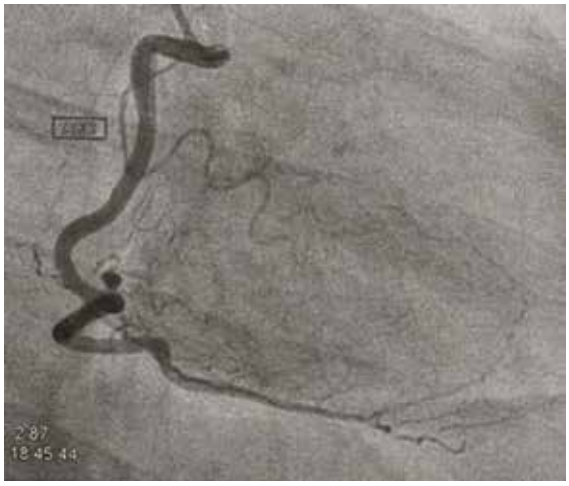

Figure 1. Videography Right Coronary Artery: Dominant, without obstructive lesions. Rocha, T.B., 11/19/2021;

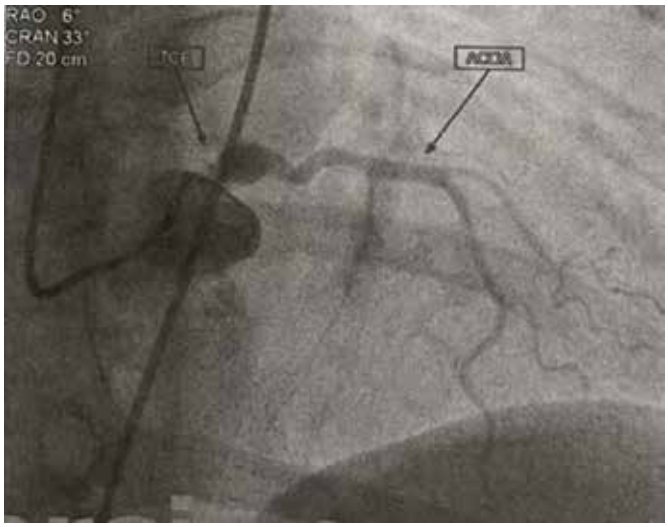

Figure 2. Cardiac catheterization image showing left main coronary artery: severe ostial lesion of 90% followed by aneurysm in the distal third. Rocha, T.B., 11/19/2021;

After evaluating the cardiac catheterization, the SYNTAX score was assessed, which presented an intermediate score, and on November 23, the intervention was performed by hemodynamic way, with angioplasty and implantation of a drug-eluting stent in the left main coronary artery, showing TIMI 3 final flow (Figure 3).

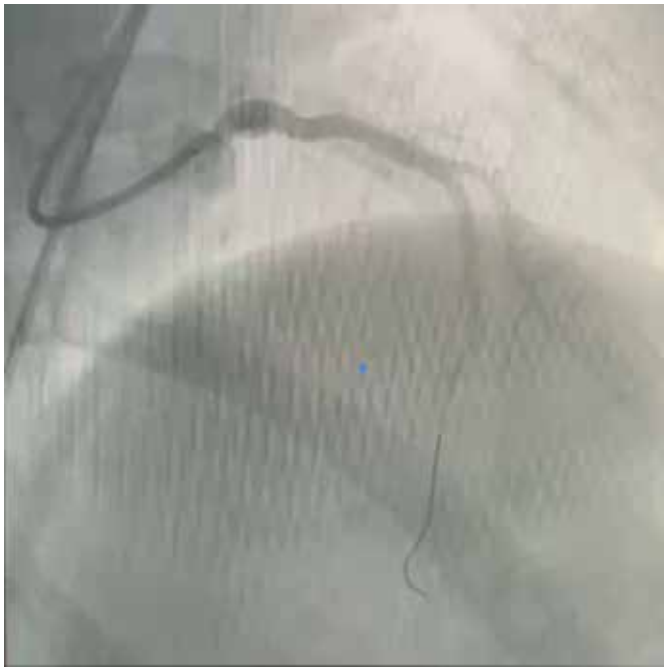

Figure 3. Imaging catheterization, Patient N.P.S.A., demonstrating final TIMI 3 flow after a procedure by hemodynamic way with angioplasty and implantation of a drug-eluting stent in the left main coronary artery. Rocha, T.B., 11/23/2021;

DISCUSSION

A prospective, randomized, open-label, non-inferiority NOBLE study was performed by Holm et al. 4 in 36 hospitals in nine northern European countries. Patients with left coronary artery disease requiring revascularization were randomly assigned to receive either percutaneous coronary intervention (PCI) or myocardial revascularization surgery (MRS). Patients were followed up for an average of 3.1 years, with this, the authors found that, in revascularization of left main coronary artery disease (LMCA), PCI was associated with a lower clinical outcome at 5 years compared to MRS. Mortality was similar after both procedures, but patients treated with PCI had higher rates of nonsurgical myocardial infarction and the need for revascularization. These results were also pointed out by Giacoppo et al. 2 the authors emphasized that PCI and MRS show comparable safety in patients with LMCA stenosis and low-to-intermediate coronary artery disease. However, repeated revascularization is more common after PCI.

In the research by Serruys et al.3, the authors evaluated 1,800 patients with disease of three vessels or of the left main coronary artery to be submitted to MRS or PCI. Most preoperative characteristics were similar in both groups. The 12-month rates of major adverse cardiac or cerebrovascular events were significantly higher in the PCI group (17.8%, vs. 12.4% for MRS), largely because of an increased rate of repeat revascularization (13.5 % vs. 5.9%). As a result, the non inferiority criterion was not met. At 12 months, death and myocardial infarction rates were similar between the two groups; stroke was significantly more likely to occur with MRS (2.2% vs. 0.6% with PCI). It was concluded that MRS remains the standard of care for patients with three-vessel or left main coronary artery disease, as compared with PCI, it resulted in lower rates of major adverse cardiac or cerebrovascular events at 1 year.

For Head et al. [5] the SYNTAX score has emerged as a valuable tool for classifying the complexity of patients with coronary disease. Although there is inter- and intra-observer variability in the calculation of the SYNTAX score, this appears to no longer be a clinically relevant issue after appropriate training. The SYNTAX score is now advocated in clinical guidelines and is increasingly being used worldwide in daily clinical practice. Integrating the SYNTAX score in multivessel coronary patients and decision making seems inevitable as current studies and clinical guidelines continue to expand its use. The method evaluates the difficulty and the chance of success of the percutaneous intervention, through the evaluation of several items related to the coronary plaque - such as its location in the vessel, length, calcification, proximity to the bifurcation, chronic occlusions, etc., and the larger, the more technically difficult to perform PCI.

In the study by Jahangiri et al. 6 the authors reviewed the methodology, results, caveats and statements about the EXCEL and SYNTAX study. It was concluded that for patients with less complex lesions, SYNTAX found PCI to be an acceptable alternative, although it was not designed to assess the overall effectiveness of PCI versus MRS. It was in this context that the EXCEL trial was designed to investigate new-generation PCI versus MRS in low-risk or intermediate-risk patients. The EXCEL authors' initial conclusion that there was no significant difference between PCI and MRS with regard to the composite endpoints of death, stroke, or myocardial infarction at 5 years, these results have subsequently been questioned. This is related to controversies over study methodology, disagreements over which definition of periprocedural myocardial infarction was used, and alleged investigator conflicts of interest. The EXCEL disaster has, to some extent, undermined public confidence in medical research in general and clinical trials in particular.

The 2021 guidelines of the Brazilian Society of Cardiology on unstable angina and acute myocardial infarction without ST-segment elevation indicate that to choose the treatment, the SYNTAX score tool should be used, and patients with SYNTAX > 22 points (intermediate or high) have greater long-term benefit from surgical revascularization. In case of urgency or emergency, the use of venous to arterial grafts should be recommended. Cardiopulmonary bypass aid may or may not be used, taking into account the conditions of each individual. For patients with cardiogenic shock, complete revascularization through angioplasty should be the initial option; however, due to its limitation, surgery may be indicated according to a multidisciplinary evaluation[7].

European guidelines highlight that predicted surgical mortality, anatomical complexity of coronary artery disease and early completion of revascularization are important criteria for decision making regarding the type of treatment. Whether conservative therapy, PCI, or MRS is preferred should depend on the risk-benefit ratio of these treatment strategies, assessing the risks of periprocedural complications (e.g., cerebrovascular events, blood transfusions, renal failure, new-onset arrhythmias or surgical wound infections) against improvements in health-related quality of life, as well as prevention of death, myocardial infarction or repeated revascularization in the long term8.

Buszman et al.[9] reported the 10-year clinical follow-up of 105 patients with stenosis of the unprotected left main coronary artery with low and medium complexity of coexisting coronary artery disease according to the SYNTAX score. Patients were treated with PCI with stent (n = 52) or MRS (n = 53). Drug-eluting stents were implanted in 35%, while arterial grafts for the anterior descending artery were used in 81%. At 10 years, there was a trend towards higher ejection fraction on stents compared to surgery. There was no statistical difference in mortality between the groups, however, numerically the difference was in favor of the

stent. Likewise, there was no difference in the occurrence of myocardial infarction, stroke and rates of repeated revascularizations.

Ruel et al. 10 pointed out that when the stenosis is in the initial part of the coronary tree and is relatively large in caliber and short in length, the stenosis seems to be an anatomically attractive target for PCI. However, as the tissue is more elastic than coronary vessels, balloon angioplasty has been associated with immediate procedural unpredictability as well as unacceptable rates of restenosis and early mortality. The adoption of bare metal stents has rejuvenated interest in PCI for coronary disease, with a reduction in acute procedural complications (eg, recoil, abrupt closure, or dissection). Along with the non-negligible risks of operative mortality and morbidity associated with MRS, as well as the high rate of saphenous vein graft attrition, many interventional cardiologists have sought to explore less invasive procedures. Among elective, low-risk patients, procedural and short-term results are acceptable. However, the stent repetition rate still remains excessive.

In this sense, for Lee et al.11 patients with stable ischemic heart disease, anatomical conditions associated with a low risk of complications from the PCI procedure and a high probability of good long-term results (for example, a low SYNTAX score, ostial stenosis or of the main coronary artery), in addition to clinical characteristics that predict a significantly increased risk of adverse surgical results, conservative treatment with PCI should be performed.

CONCLUSION

In patients with stenosis of the main coronary artery with low and medium complexity of coexisting coronary artery disease, PCI treatment offers a favorable long-term outcome, therefore, it constitutes an alternative therapy to MRS, as it is a less invasive and effective treatment.

Percutaneous coronary intervention and coronary artery bypass graft surgery are treatment options for coronary artery bypass grafting in selected patients with stable coronary artery disease and ischemia. Current revascularization guidelines indicate that treatment selection depends on patient preferences, comorbidity, and the complexity of the coronary artery disease. Less complex one- or two-vessel coronary artery disease is preferentially treated with PCI, where the level of acceptance is higher for PCI, while complex three-vessel disease is better treated with MRS.

DATA AVAILABILITY

Data used to support the findings of this study are available from the corresponding author upon request.

CONFLICTS OF INTEREST

The authors declare that there are no conflicts of interest in the publication of this manuscript.

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OSLER WEBER RENDU SYNDROME: CASE REPORT

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ABSTRACT

Osler Weber Rendu syndrome or Hereditary Hemorrhagic Telangiectasia is a rare autosomal dominant genetic disorder. The most common clinical manifestation is epistaxis, but it can also present with melena and dyspnea. The diagnosis can be made with the criteria of Curaçao, which are four, namely: recurrent noseblreds, telagiectasias, arteriovenous malformations and family history; three positive criteria confirm the diagnosis. Objective: To present a case of a female patient, diagnosed at age 53 with Osler Weber Rendu syndrome. Methodology: Collection of data from the medical records of the selected patient diagnosed with Osler Weber Rendu syndrome. Information was collected, clinical, exams performed, the therapy of choice and outcome. Articles were searched for bibliographic review in the Virtual Health Library (VHL), LILACS, PubMed. Conclusion: The case report contributed to the discussion and warning about the clinical manifestations of a rare syndrome, but which can be present in patients from different medical specialties, since its main signs and symptoms affect several systems of the human body.

KEYWORDS: RENDU-OSLER-WEBER SYNDROME, HEREDITARY HEMORRHAGIC TELANGIECTASIA, ARTERIOVENOUS MALFORMATIONS, EPISTAXIS, AUTOSOMA

INTRODUCTION

Osler Weber Rendu Syndrome or Hereditary Hemorrhagic Telangiectasia (HHT) is a rare systemic fibromuscular dysplasia, whose basic defect is an alteration of the elastic lamina and muscular layer of the blood vessel wall, which makes them more vulnerable to trauma and spontaneous ruptures[1,2].

The disease has an autosomal dominant transmission, although in about 20% of cases there is no family history. Its incidence in the population is 1-2/100,000 and it has a homogeneous distribution between race and sex.[3]

The diagnosis is made according to the Curaçao criteria: telangiectasias on the face, hands and oral cavity; recurrent epistaxis; arteriovenous malformations with visceral involvement; family history. The diagnosis is confirmed in the presence of at least 3 of these manifestations.[4]

Otorhinolaryngological manifestations are the most frequent, with recurrent epistaxis being the main one. Blood vessels in other regions may also be affected, especially the lungs, brain, skin and gastrointestinal tract1,[4-5]. The most common bleeding from this pathology is non-traumatic epistaxis, which affects about 50% of patients before twenty years of age and approximately all of them throughout their lives (78% to 96%)6; gastrointestinal bleeding, recurrent in 33% of patients, mainly after the fourth decade of life7; and pulmonary and cerebral bleeding, with a 0.5% chance of bleeding per year[8,9].

The present study presents a case of a 56-year-old patient who was diagnosed with Osler Weber Rendu syndrome at the age of 53, but who, since the age of 42, had presented signs and symptoms suggestive of the syndrome, but had not been diagnosed. Thus, this case report aims to alert physicians from different specialties about the importance of diagnosing this syndrome to contribute to the quality of life of patients by reducing and treating their decompensations.

METHODOLOGY

It consists of a case report, with data collection from the medical records of the selected patient diagnosed with Osler Weber Rendu syndrome. Information was collected, clinical examinations performed, the therapy of choice and outcome.

Articles were searched for bibliographic review in the Virtual Health Library (VHL), LILACS, PubMed and Scopus.

The study followed the ethical guidelines for the development of research with human beings, in particular what is recommended in resolution 466/2012 of the National Health Council. Obtaining the patient's informed consent form.

CASE REPORT

Female patient, 56 years old was admitted to the gastroenterology service of the Hospital Geral de Goiânia (HGG) in May 2023 to undergo an argon colonoscopy due to chronic melena and anemia with frequent need to re-

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ceive blood transfusions.

The patient reports that at the age of 42 she had a stroke that evolved with mild motor sequelae. At the age of 53, she presented dyspnea, orthopnea and edema of the lower limbs that had lasted for 6 months, accompanied by anemia and urinary tract infection.

The patient was diagnosed with Osler Weber Rendu syndrome, as she had telangiectasias in the gastrointestinal tract seen by upper digestive endoscopy and colonoscopy. In addition to this, the pulmonary arteriovenous malformations evidenced by angiotomography corroborated the diagnosis of the syndrome. The telangiectasias justified the episodes of melena and, consequently, the anemia. Arteriovenous malformations (AVM) justified the picture of decompensated heart failure secondary to pulmonary hypertension.

Chest angiotomography performed in 2021 showed: Arteriovenous malformation located predominantly in the upper left lobe, measuring approximately 63 x 38 mm, nourished by the left lower lobe arterial branch and drained by the ipsilateral superior pulmonary vein. Note also another lesion with similar characteristics in the periphery of the left upper lobe, measuring 10 x 9 mm, nourished by a segmental arterial branch and draining into the interior of the malformation described above. Pulmonary arterial trunk with preserved caliber. Heart dimensions and anatomical configuration. There were no signs of pleural or pericardial effusion. Lung parenchyma with usual attenuation, without evidence of consolidations. Trachea, main and lobar bronchi patent, without significant parietal thickening. Absence of mediastinal lymph node enlargement. Musculoskeletal structures of the rib cage with a preserved appearance. Since then, the patient has been followed up by the thoracic surgery team to perform a possible lobectomy or pneumonectomy to correct the arteriovenous malformation.

In May 2023, the patient was admitted to the Emergency Care Unit with syncope, melena and a hemoglobin of 3.5, at the time a blood transfusion was performed and the patient was referred to the HGG for an argon colonoscopy by the gastroenterology team.

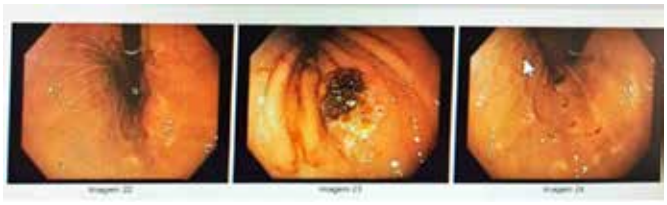
Colonoscopy and endoscopy were performed, both with argon application. The exams carried out during this hospitalization showed, colonoscopy: angiectasia in the cecum without signs of active bleeding, opting for ablative therapy with application of argon plasma. No active or recent bleeding was observed in the studied path. Bloody secretion with loose clots in the terminal ileum was visualized, suggestive of upper digestive bleeding.

At upper digestive endoscopy (figures 1 and 2) gastric and bulboduodenal vascular ectasias were seen. Hemostasis was performed by electrocoagulation with argon plasma, visualization of previous electrocoagulation scars in the gastric body

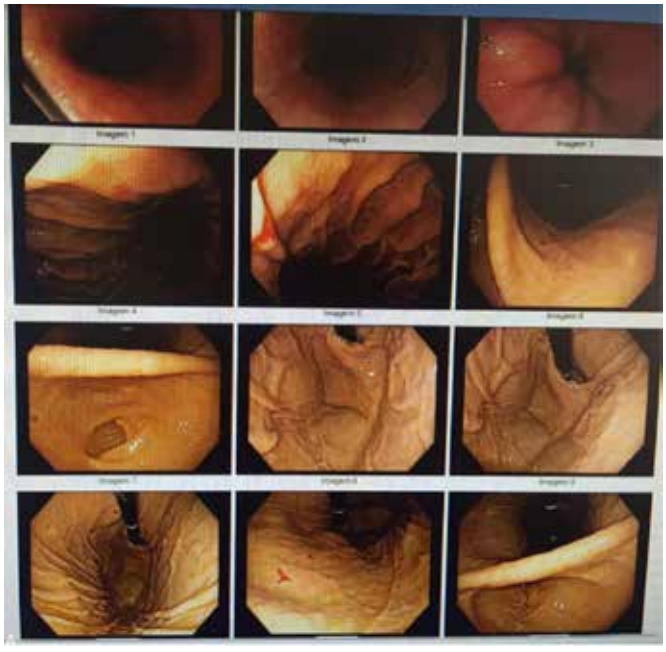
Cervicothoracic arteriography showed a patent left pulmonary artery and descending branch, with the formation of a large mass with rapid emptying of contrast, suggesting arteriovenous malformation. Malformation communication diameter ranging from 13 to 18 mm. Arteriovenous malformation in the left lung with pulmonary hypertension.

The chest angiotomography (figures 4 and 5) showed an arteriovenous malformation involving the apicoposterior and superior lingular segments of the left upper lobe, the arterial supply coming mainly from the left interlobar artery but also on a smaller scale from branches of the superior lingular segment. Venous drainage is through the left superior pulmonary vein. Lesion measuring approximately 70 x 49 mm. In the chest X-ray (figure 3), a radiopaque image can be seen in the left hemithorax, shown by the arrow.

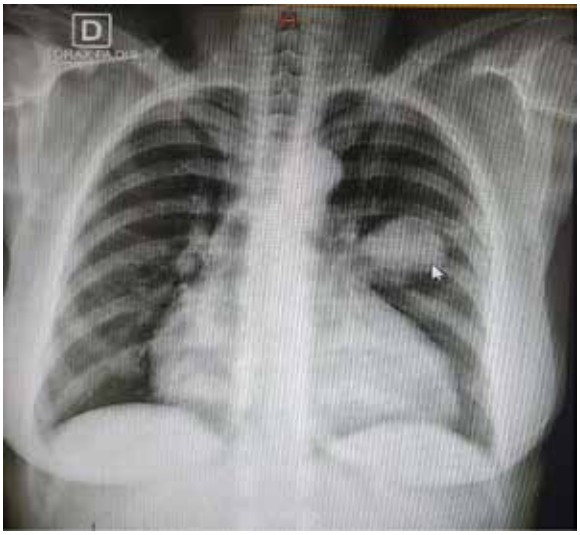
The patient evolved well and did not need new transfusions. During discussions with the multidisciplinary team, the conclusion was reached that pulmonary AVM should be evaluated during hospitalization by the vascular surgery and thoracic surgery team that opted for an elective approach to the arteriovenous malformation.



MORAES. FIGURE 1



MORAES. FIGURE 2



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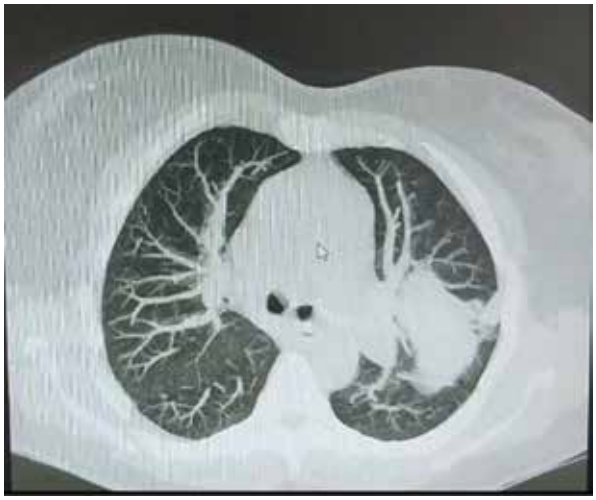

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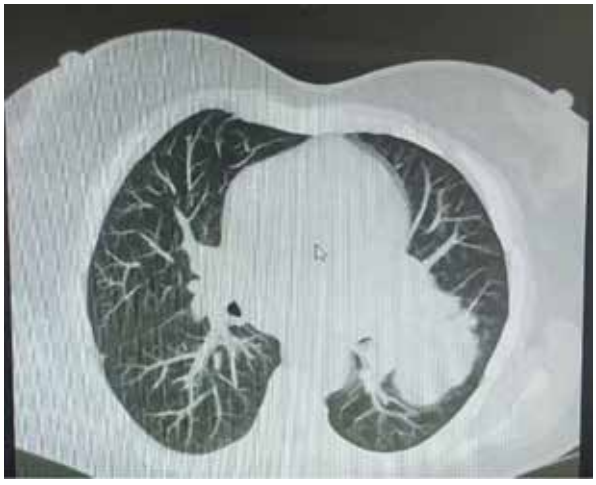

MORAES. FIGURE 5

DISCUSSION

Osler Weber Rendu syndrome, or hereditary hemorrhagic telangiectasia, can be diagnosed using a probability score defined and presented in 2000 by the Scientific Advisory Board of the THH Foundation International, called the Curation criteria.[10] These criteria facilitate the recognition of clinical findings that are less common than epistaxis, which is the main manifestation of the disease in affected individuals,[11] and allow early recognition in individuals with less classic but potentially serious manifestations, such as pulmonary AVMs.

Based on these criteria,[10] the diagnosis can be definitive (when three criteria are present); possible, (if two criteria are present); or suspected (if less than two criteria are present). The criteria are: 1) presence of epistaxis (spontaneous and on more than one occasion); 2) presence of visceral lesions (gastrointestinal telangiectasia or pulmonary, hepatic, cerebral or spinal vascular malformation); 3) presence of mucocutaneous telangiectasia in a typical location; and 4) first-degree family history (or presence of the genetic mutation). In families with individuals with HHT, the diagnosis can be made from the findings of two sites with visceral lesions.[10]

The presence of chest X-ray findings compatible with pulmonary AVMs is the gold standard for diagnosing these malformations. The most common radiological presentation is the presence of well-defined peripheral nodules. The use of intravenous contrast is not mandatory, but it may allow a better definition of the angioarchitecture of the pulmonary AVM to plan endovascular therapy.[12]

Upper digestive endoscopic therapy is indicated in every patient with suspected upper digestive hemorrhage or in cases of patients with HHT when iron replacement is not enough to contain the anemia[13]. Endoscopy has potential for definitive treatment. In cases of intestinal angiectasia, therapy with argon plasma is indicated as the gold standard [13].

Embolization is the standard of care for pulmonary AVMs, 14,15 with substantial improvement in oxygenation and reduced risk of embolic events. [16.17]

Performing a lobectomy or segmentectomy is restricted to cases of complex or multiple pulmonary AVMs, when catheter embolization is not possible. [18]

FINAL CONSIDERATIONS

Osler Weber Rendu syndrome or hereditary hemorrhagic telangiectasia is rare, but must be part of the differential diagnosis of several signs and symptoms, including: epistaxis, melena, dyspnea, orthoodexia, platypnea, syncope and telangiectasia.

The patient in the present case already presented complications of the syndrome at the age of 42, however, she received the diagnosis only at the age of 53. Upon admission to the HGG service, the digestive hemorrhage was

approached through endoscopy and colonoscopy, both with argon injection. The pulmonary arteriovenous malformation was discussed by the vascular and thoracic surgery teams and the teams decided to study the best approach and schedule the procedure on an elective basis.

Thus, this case report alerts physicians from different specialties, whether clinical or surgical, about the importance of diagnosing this rare but serious syndrome that can lead to early mortality and morbidity.

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AIRWAY CARE DURING INTUBATION OF A SUPER-OBESE PATIENT: CASE REPORT

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ABSTRACT

The definition of difficult airway is based on the difficulty of orotracheal intubation (OTI) and/or mask ventilation by a trained professional. Planning for airway management is of paramount importance in the daily lives of anesthesiologists and is sometimes underestimated. Obesity is a worldwide public health problem, with multifactorial causes, and this is a condition that can hinder both mask ventilation and OTI. It is essential to know how to identify predictors and prepare for unexpected and expected scenarios. This report deals with the management of the airway of a 21-year-old male patient with morbid obesity (weight 254 kg, height 1.76 cm, body mass index (BMI) 81 kg/m2), who would be maintained at passage of a gastric balloon for weight loss and subsequent bariatric surgery. Although the procedure is often performed with sedation, it was decided to obtain a definitive airway and balanced general anesthesia, avoiding possible unhealthy complications in the intraoperative period. In assessing the airway, the patient had a beard, BMI> 26, good inter-incisor distance (>5cm), large tongue, Mallampati two, with good mandibular protrusion (upper lip bite test - ULBT class 1), sternum- mentalis > 12cm, with good extension and cervical mobility, but with cervical inclusion >40cm. OTIs were considered with a flexible bronchoscope or videolaryngoscope, with the second option being preferred after assessing the patient's airway. OTI uneventful surgery was also performed without setbacks, the patient being decurarized and extubated in the room, sent to the post-anesthesia recovery room. Thinking about a safe anesthesia, it is not recommended to perform an airway procedure without careful planning. In this sense, a pre-anesthetic evaluation is of paramount importance and should not be overlooked due to care time.

KEYWORDS: MANAGEMENT, AIRWAY; INTUBATION, INTRATRACHEAL; OBESITY; OBESITY, MORBID; BARIATRIC MEDICINE.

INTRODUCTION

Planning for airway management in anesthesiology is very important in the daily practice of anesthesiologists, although it is sometimes overlooked. The definition of difficult airway (DA) according to the American Society of Anesthesiologists (ASA) would be the difficulty of orotracheal intubation (OTI) and/or mask ventilation by a trained professional [4]. Simple physical examination can help to assess possible predictors, which increases the importance of pre-anesthetic evaluation even before the patient arrives at the operating room. Planning is something that helps prepare for the possibility of difficult mask ventilation, difficult intubation, and/or obtaining an advanced airway [1]. Knowing this, several algorithms were created to standardize the management of DA in order to avoid behaviors that put the patient's life at risk [4].

Obesity is a worldwide public health problem, with multifactorial causes [3]. The alterations resulting from obesity can cause difficulties in mask ventilation, OTI and mechanical ventilation, with important peculiarities that must be taken into account already in the pre-anesthetic evaluation [3].

The main purpose of this report is to show the evaluation of the airway in the pre-anesthetic consultation and the plans for OTI of this patient.

CASE REPORT

Male patient, 21 years old, morbidly obese (weight 254 kg, height 1.76 cm, body mass index (BMI) 81 kg/m2) denied other cardiovascular, respiratory, endocrine comorbidities, among others. He denied addictions, known drug allergies, prior surgical history. Accompanied by family members at the appointment, he would undergo an intragastric balloon for weight loss and future gastroplasty in a second stage.

Regarding the assessment of the airway, the patient had a beard, BMI> 26, good inter-incisor distance (>5cm), large tongue, Mallampati two, with good mandibular pro-

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trusion (upper lip bite test - ULBT class 1), sternum- mentalis >12cm, with good cervical extension and mobility, but with cervical circumference >40cm. During the pre-anesthetic consultation, he was instructed to remove the beard, which would be a possible modifiable difficulty agent for mask attachment (Figure 01).

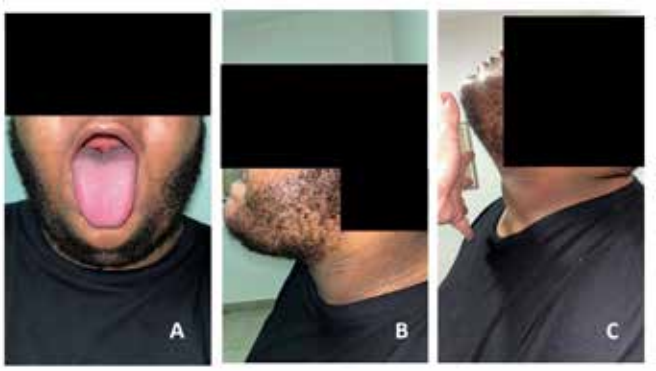

Figure 1. Images from the primary assessment of the airway in a pre-anesthetic consultation. We observed the presence of a beard, cervical circumference, Mallampati (A), upper lip bite test - ULBT (B), sternomental distance, cervical mobility (C).

Generally, the procedure of passing an intragastric balloon is performed with sedation. Knowing the predictors, general anesthesia balanced with OTI was previously chosen to obtain a definitive airway, thus avoiding some possible complications that could endanger the patient's airway during the procedure. Two possibilities for OTI were suggested, the first being via flexible fiberoptic bronchoscopy, and the second under direct visualization with videolaryngoscopy. After discussion among the anesthesiologists and considering the predictors, they chose the second option, but keeping the flexible fiberoptic bronchoscope in the room. So, when the day of the procedure arrived, the patient was fasting properly, he was positioned in dorsal decubitus under a trapeze, joint care was observed, monitored with oximetry, cardioscopy, non-invasive blood pressure (NIBP) in the right forearm, CONOX monitor, punctured 20G Jelco in left upper limb. Dexmedetomidine 50mcg was administered, followed by pre-oxygenation with an appropriately sized mask for the patient, FiO2 100% for 5 minutes. Venous induction was performed with fentanyl 50mcg, propofol 200mg, lidocaine 100mg and rocuronium 50mg. Ventilation was performed under a mask for 03 minutes and laryngoscopy was performed with direct videolaryngoscopy, observing Comarck-Lehane 1, periglottic anesthesia with 1% ropivacaine 5ml, Bougie passage and subsequent passage of an orotracheal tube (OTT) No. 8.0, with cuff, which was inflated with direct videolaryngoscopy and capnography was detected. There was no hemodynamic instability or other setbacks during induction. Surgical procedure was performed uneventfully (Figure 02). For extubation, he was decurarized with sodium sugammadex 200mg. Woke up peacefully, without agitation and extubation while still in the operating room. Kept in anesthetic recovery room (PACU) for another hour and released to the ward with an Aldrette-Kroullik (AK) scale of 10.

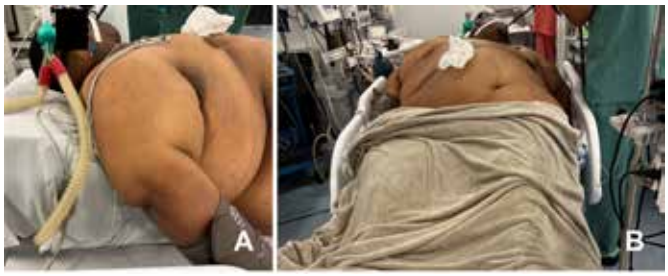

Figure 2. Images of positioning on ramp for induction, mask-assisted ventilation, orotracheal intubation during surgical procedure.

DISCUSSION

At first, in the pre-anesthetic evaluation, we see some worrying factors in relation to this patient's airway. Factors such as male gender, neck circumference, tongue size, BMI and beard are some important predictors of difficulty with mask ventilation and difficult OTI. On the other hand, the interincisor distance, ULBT, young age and Mallampati are factors of probable non-difficulty in OTI. Knowing this, we have two scenarios, the expected and unexpected factors [1]. The main idea is to adapt the conduct so that the expected factors could be controlled and be better prepared for the unexpected. One of them, modifiable, was the removal of the beard. All safe surgery checklists previously checked. Ramp positioning with the use of the trapeze also influences, being important in improving denitrogenization, reducing possible formations of atelectasis and aligning the airway axes (oral, pharyngeal and laryngeal) [1,2]. Regarding pre-oxygenation, a criticism could be the use of a high-flow nasal catheter during the apnea phase, where there is evidence of improvement in the safe apnea time.

From the point of view of the surgical procedure, usually performed under sedation, ensuring a definitive airway was imperative for anesthesiologists in order to avoid complications and unexpected intraoperative airway scenarios, which is why general anesthesia with OTI was chosen.

Regarding mechanical ventilation, obesity can cause restrictive disorders mainly due to the weight of the rib cage, reducing functional residual capacity (FRC) and increasing airway pressure due to the reduction in airway caliber [3]. Knowing this, all the necessary precautions were taken to maintain protective ventilatory parameters, with positioning being important both for OTI and for mechanical ventilation.

During extubation, a peaceful awakening and topical airway anesthesia were designed to avoid significant

bronchospasm and/or laryngospasm, which are also unexpected factors, but which can be avoided with the measures taken, as well as serial evaluation in the PACU in order to quickly identify complications [1].

CONCLUSION

In the case presented here, considering morbid obesity and several DA criteria, the pre-anesthetic evaluation was extremely important for the surgical procedure to be carried out safely, and should not be overlooked by the anesthesiologist and the care team.

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CHRONIC MASTITIS

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ABSTRACT

Non-puerperal mastitis is a chronic inflammation of the breast that may or may not be associated with an acute infection. It is a benign and low-recurrence breast condition. There are different types of mastitis, classified as infectious or non-infectious. When infectious, it may have a viral, fungal or bacterial etiology. When non-infectious, it can often be associated with another underlying disease, or still have an unknown etiology. The diagnosis of these mastitis can be made through imaging tests, such as mammography and ultrasonography, in addition to biopsies and other laboratory tests. Despite being a rare process, its diagnosis is important to reduce prolonged morbidity and alleviate the suffering of affected patients. Treatment varies depending on the type of mastitis and may include the use of anti-inflammatories, antibiotics, surgery and even immunosuppressive drugs.

KEYWORDS: MASTITIS, NON-PUERPERAL MASTITIS, CHRONIC INFLAMMATION, BREAST CONDITION

INTRODUCTION

Non-puerperal mastitis is a chronic inflammation, with a considerably slow evolution, which may or may not be preceded by an acute infection. It is identifiable through the appearance of connective tissue rich in macrophages and fibroblasts in the breast parenchyma, in addition to vascular neoformation and exudative phenomena. It is a benign breast entity with low recurrence rate, representing about 1-2% of all symptomatic breast processes [1]. However, it can be a source of prolonged morbidity, and it is important to pay attention to imaging, physical and clinical findings , in order to advance the diagnosis and the most appropriate treatment [1].

Mastitis can be classified into infectious and non-infectious, as described in table 1:

Mastitis	
Infectious	Non-infectious
Recurrent chronic subareolar abscess	Ductal ectasia or periductal mastitis
Bacterial infections	Granulomatous mastitis
• Tuberculosis	Steatonecrosis
• Leprosy	Mondor's disease or superficial phlebitis
• Syphilis	Sarcoidosis
• Atypical mycobacteria	Lupus mastitis
• Gonococcal	Actinic mastitis
• Actinomycosis	Oleogranulomatous mastitis.
• Luetic mastitis	Lymphocytic mastitis
Fungal lesions	Diabetic mastopathy
• Paracoccidioidomycosis	
• Pityriasis	
• Versicolor	
Viral infections	
• Herpes	
Parasitic infections	
• Cysticercosis	
• Filariasis	
Infected sebaceous and epidermal cysts.	

Table 1 - Difference between infectious and non-infectious types of mastitis

MAIN MASTITISES FOR CLINICAL PRACTICE

RECURRENT CHRONIC SUBAREOLAR ABSCESS

Also known as Zuskas disease, it is a chronic inflamma-

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tory process that affects the central portion of the breast, outside the pregnancy-puerperal period, and may lead to the formation of fistulas. It is more common in people between 30 and 40 years old, with smoking and vitamin A deficiency being important risk factors.

The disease begins with a localized inflammation in a subareolar area, evolving to the formation of small abscesses that drain spontaneously, forming fistulas that heal over time.

This process can be repeated several times, with intervals of months to years. Furthermore, younger patients are more prone to mastalgia, which may precede the development of inflammatory masses, while older patients tend to have less pain associated with palpable masses, possibly due to less acute inflammation and greater presence of fibrosis.

The consistency of secretions also varies between ages, with younger patients having less viscous discharge and older patients having increased viscosity.

Non-puerperal subareolar abscesses often require multiple drainage or surgical procedures, and approximately one third of patients develop fistulas [1].

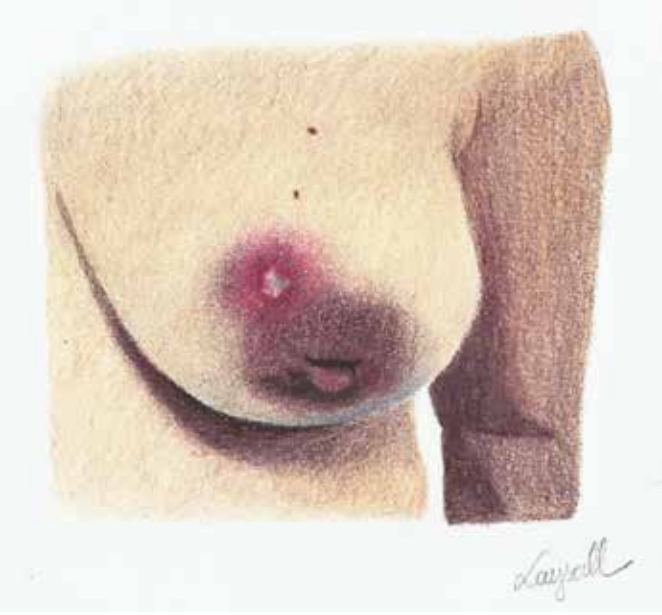

Figure 1 – Recurrent chronic subareolar abscess. The distended canals can then rupture into the periductal mesenchymal tissue, leading to the formation of a periareolar fistula, with the outflow of secretory material.

Clinical condition

Inflammation with abscess formation in the breast is probably caused by an obstruction of the distal subareolar main ducts due to squamous metaplasia. This obstruction leads to distention of the ducts due to the accumulation of secretions, keratin, and sloughing necrotic cells. Thus, rupture of the ducts in the periductal tissue results in the formation of a fistula, with drainage of cell debris and secretory material. In addition, the presence of a bacterial superinfection in the inflamed and necrotic region can lead to the formation of an abscess, which can communicate with the skin through a fistula (Figures 2 and 3). Subareolar abscess is associated with infections caused by Staphylococcus aureus, S. albus and Streptococcus [1].

Figure 2 – Photograph of woman presenting chronic recurrent subareolar abscess with periareolar fistula.

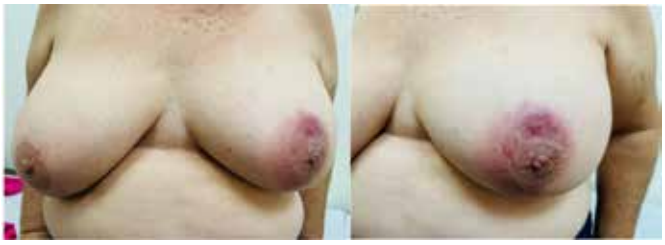

Figure 3 – Photograph of a woman with a recurrent chronic subareolar abscess with periareolar fistula and reactivation of the infectious process.

Diagnosis

The most frequently described mammographic findings include a mass, either focal or diffuse asymmetry. Lesions range from 1.0 to 5.0 cm (mean 2.0 cm). Ultrasound findings include complex cystic lesions (about 50% of cases) and non-specific heterogeneous hypoechoic masses. In cases of breast abscess, imaging is recommended, possibly followed by a biopsy. The diagnosis of fistula is clinical and is identified by a persistent periareolar flow [1].

Treatment

The most effective treatment for primary or secondary fistulas is surgical excision of the fistula, along with removal of adjacent retroareolar tissue and inflammatory tissues. In turn, pharmacological treatment can also be used, with antibiotics that include anaerobes, such as metronidazole and doxycycline. Therefore, when inflammation is in remission, surgery is recommended to prevent recurrences, including resection of the fistulous tract or, in more severe cases, radical surgery, such as resection of the terminal ductal system (Urban's surgery) and removal of the inflammatory tissue and the fistulous tract, accompanied by antibiotic therapy. Particularly in patients who do not wish to breastfeed, retroareolar cone excision of the main ducts can be performed [1].

Granulomatous mastitis

Chronic granulomatous mastitis is a rare inflammatory condition of unknown cause, characterized by the presence of noncaseating granulomas and microabscesses confined to the mammary lobe. Clinical symptoms and radiological findings can be confused with a neoplasm or acute breast infection, which can lead to a delay in the definitive diagnosis. This condition occurs mainly in young women, aged between 17 and 42 years, who have had a recent history of lactation, and it is even rarer in nulliparous women [2–4].

The etiology is unknown, but there is evidence of factors such as microbiological agents, hormonal effects and immunological changes. Corynebacteria, Gram-positive bacteria found in the epithelial flora, can invade deep into the breast tissue through the ducts, for example. Thus, the condition involves autoimmune reactions, resulting in extravasation of secretions and persistent inflammation of stromal cells [2–4].

Premenopausal women often have clinical symptoms such as a breast mass that is hard and adherent to the skin, in addition to axillary nodules and nipple retraction, which can resemble breast cancer (Figure 4). These symptoms are usually associated with abscesses, inflammation and fistula formation in the mammary duct, affecting the skin of the areola itself [2–4].

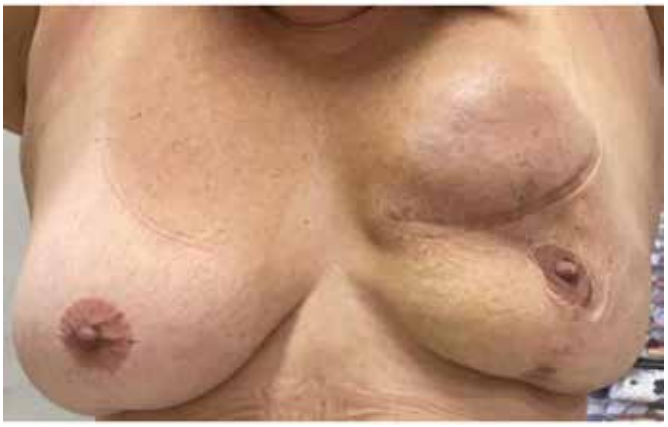

Figure 4 - Granulomatous mastitis in the left breast

Diagnosis

Granulomatous lobular mastitis is diagnosed by excisional biopsy or percutaneous core needle biopsy. Pathological findings reveal the presence of non-caseating granulomas in the lobules, with Langhans giant cells, epithelioid and polymorphic histiocytes.

Imaging tests, such as mammography and ultrasound, can show variable characteristics and sometimes suggest malignancy. These tests can detect skin thickening, calcifications, asymmetrical density, single or multiple masses, area of architectural distortion, and hypoechoic nodules.

However, in many cases, these tests do not show specific abnormalities. Furthermore, the presence of diffuse abscess and fistula formation can also be observed [2–4].

Treatment

There is no standardized treatment for granulomatous lobular mastitis. The main options include surgery, drug treatment, or a combination of both. Preferred treatment consists of taking Prednisolone 40 mg daily for four weeks, with doses progressively reduced over the weeks, plus Doxycycline 100 mg every 12 hours for 10 days or Tetracycline 500 mg every 6 hours for two to four weeks.

In cases of persistent large tumors and breast deformities, wide surgical resections may be considered. In some cases, when there is a contraindication for the use of corticosteroids or relapse after discontinuing the corticosteroid, immunosuppressive agents such as methotrexate can be used [2–4].

Ductal ectasia mastitis

Ductal ectasia mastitis, also known as mastitis obliterans, is a benign condition characterized by dilation of the mammary ducts, periductal inflammation, and fibrosis. Its exact cause is still unclear, but studies indicate that stimulation of the squamous epithelium, infections and smoking may be related. It presents symptoms similar to granulomatous lobular mastitis and can be observed through imaging exams. However, mastitis from ductal ectasia is often accompanied by discharge and nipple retraction, and breast masses are usually found in the subareolar region. Symptoms include serous or hemorrhagic papillary effusion and a retroareolar tumor. Some patients may have episodes of acute infection with swelling, redness and fever, in addition to increased local sensitivity and masses or palpable dilated ducts [5,6].

Diagnosis

The pathological characteristics revealed in the biopsy are dilation of the main ducts, diffuse infiltration by plasmocytes, sometimes even foreign body granulomas, which can be found around the ducts and lobules [5,6].

Mammography may show increased bilateral retroareolar density and ultrasonography identifies dilated ducts close to the papilla, with dense content and eventually abscesses (Figures 5-7) [5,6].

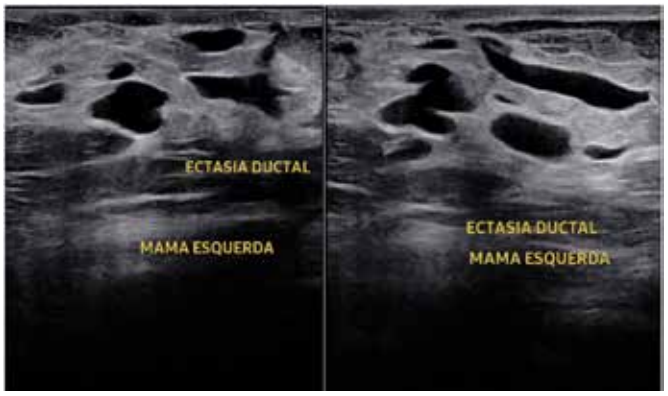

Figure 5 – Ductal ectasia – Left breast showing, on ultrasound, dilated ducts close to the papilla.

Figure 6 – Ductal ectasia. Ultrasonography identifies dilated ducts close to the papilla and dense content.

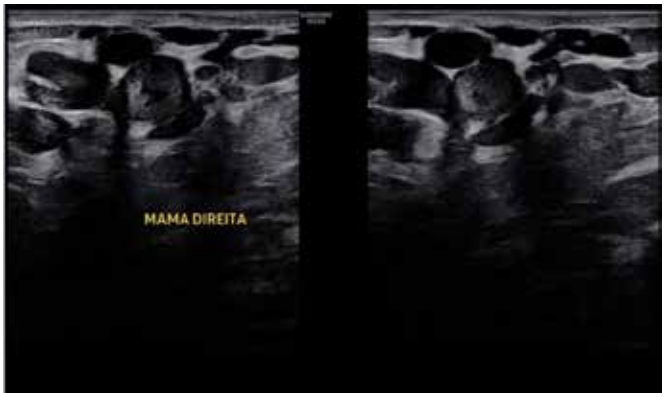

Figure 7 – Ductal ectasia. Ultrasonography identifies dilated ducts close to the papilla and dense content.

Treatment

In mild cases without significant clinical impact, a hands-free observational approach can be adopted. However, in specific cases, the surgical option can be considered, involving the removal of the affected ductal tree (Urban surgery) [5,6].

Luetic Mastitis

Specific infectious disease caused by Treponema pallidum, which manifests itself through primary, secondary and/or tertiary lesions in the breast.

The symptoms of mammary syphilis can vary according to the stage of the disease and can include a variety of skin manifestations, specific lymphadenitis, impairment of the general condition and lesions in the internal organs. In the primary form, it is common to observe a hard chancre located in the nipple-areola complex, due to the inoculation of Treponema through contact with the mouth of an infant with congenital syphilis. In the secondary form, the skin lesions may initially appear as spots and progress to papular and papulosquamous lesions typical of secondary syphilis. In the tertiary form, syphilitic mastitis goes through the three stages of syphilitic gum, with slow-growing hardened nodules that can ulcerate or form fistulas [7].

Diagnosis

The diagnosis is confirmed by the results of serological tests and cytological smears with ulcer borders [7].

Treatment

The treatment is performed with Penicillin G benzathine 2.4 million IU intramuscularly, the dose being repeated in a week [7].

Specific mastitis

The breast can be affected by several specific infections, including tuberculosis, leprosy, syphilis, gonorrhea, atypical mycobacterioses, mycobacterial infection in prostheses (Figure 8), actinomycosis, nocardiosis, cat scratch disease, candidiasis, cryptococcosis, aspergillosis, chromomycosis, blastomycosis and sporotrichosis (Figure 9). Viral infections such as herpes simplex and herpes zoster (Figure 10), in addition to parasitic infections such as myiasis (Figure 11), helminthiasis, filariasis and schistosomiasis mansoni can also occur [7,8].

The most common clinical manifestation is the presence of one or more slowly evolving and painless hardened nodules. Acute and recurrent abscesses with caseous necrosis can be seen in more than 90% of cases and multiple fistulous pathways to the skin. Diffuse thickening and sclerosis of the affected mammary parenchyma can also be observed. Many patients with tuberculous mastitis do not have symptoms such as fever, weight loss, and cytological aspiration, in addition to the fact that the tuberculin skin test and pathological examination can be negative [7,8].

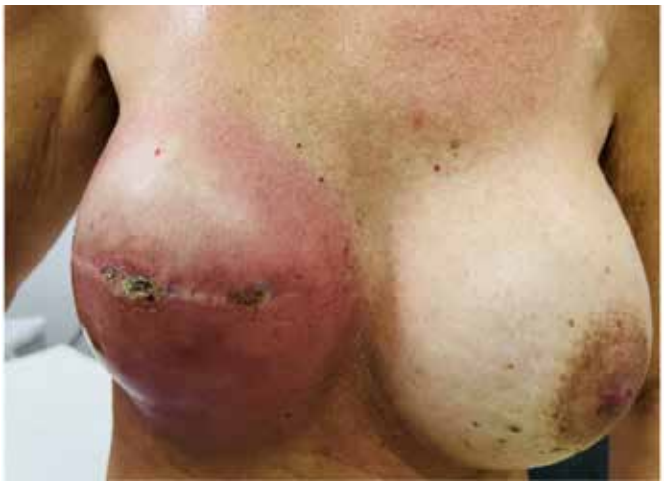

Figure 8 – Mycobacterial infection in a patient undergoing reconstruction with a silicone prosthesis after right mastectomy for the treatment of breast cancer.

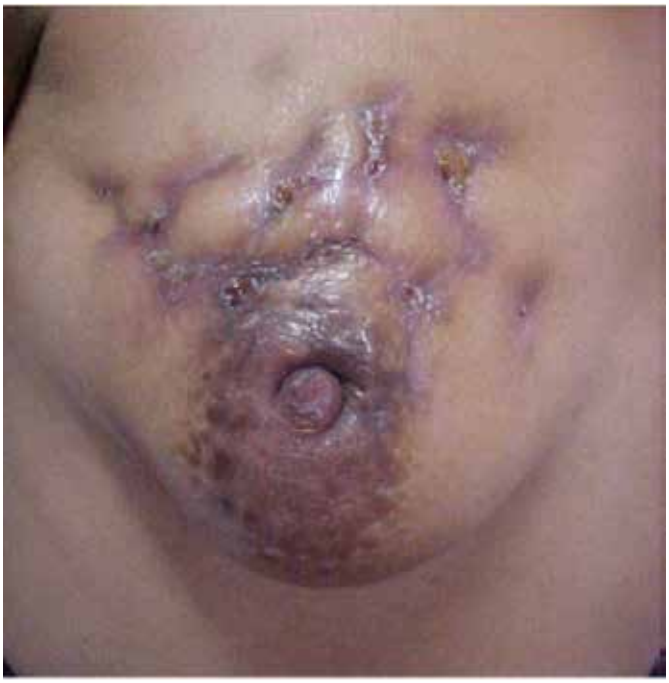

Figure 9 – Mammary sporotrichosis in a young woman from a rural area.

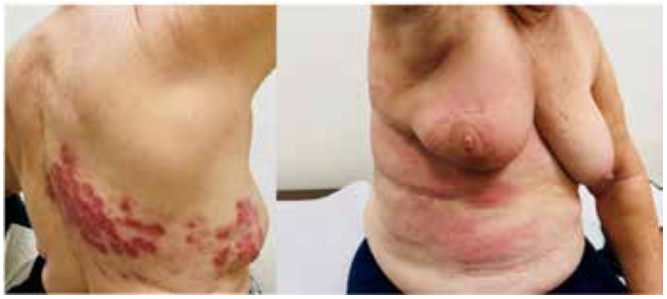

Figure 10 - Herpes Zoster in women undergoing treatment for breast cancer.

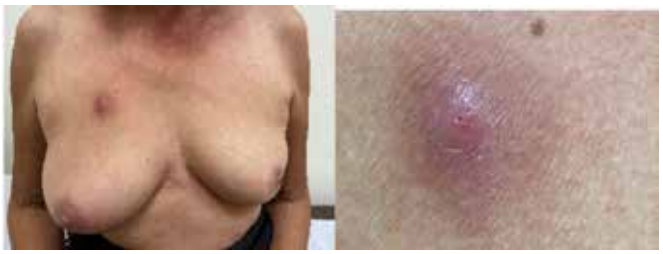

Figure 11 – Myiasis in the region of the upper medial quadrant of the right breast.

Diagnosis

The diagnosis of tuberculous mastitis, for example, involves the identification of Koch's bacillus through biopsy, Ziehl-Nielsen staining and culture. The molecular PCR test can also be used to detect the bacillus in specific cases. On imaging studies, mammography may show asymmetrical density, fibrogranular tissue, and enlarged axillary lymph nodes. Ultrasound shows solid masses with heterogeneous characteristics, presence of cystic areas and multifocal abscess cavities, in addition to enlarged axillary lymph nodes [7,8].

Treatment

The treatment for each disease must be individualized and specific. In the case of tuberculous mastitis, for example, treatment usually consists of using antituberculosis drugs for six to nine months, and surgical removal of the lesion may be necessary in more severe cases [7,8].

OTHER ENTITIES

Mondor's disease

Characterized by Henri Mondor, the disease is a superficial thrombophlebitis of subcutaneous veins, which can affect other regions such as the penis, neck and cubital fossa. It is rare, self-limiting and benign. The clinical picture presents with acute local mastalgia associated with a palpable cord or linear cutaneous depression, whose skin remains mobile. It may be asymptomatic and not show inflammatory signs [9].

Diagnosis is primarily based on clinical examination, but imaging tests support ruling out other etiologies, such as ultrasonography. In this one, the superficial vein can be found with or without the intraluminal thrombus and without flow on Doppler. The origin of the disease is still unclear, but it may be associated with Virchow's triad factors. In addition, it can be caused by muscle strain, use of tight clothing or even be related to malignant neoplasm [9].

Treatment is essentially symptomatic, given its self-limited character, with the use of anti-inflammatory drugs. In exceptional cases, resection of the affected vein may be considered if the disease does not spontaneously regress [9].

Diabetic mastitis

Associated with poorly controlled diabetes mellitus, di-

abetic mastitis is likely to have an autoimmune character related to the development of antigens in the mammary gland due to hyperglycemia, accumulating lymphocytes and proliferating epithelioid myofibroblasts. It is rare, representing less than 1% of benign breast diseases, but it can affect up to 13% of diabetic patients during the course of their illness. It usually affects premenopausal women, but it can affect men with gynecomastia [10].

It presents as a palpable tumor, with inflammatory symptoms, with recurrent and painless hyperemia, but it can become painful. Histologically there is proliferation of lymphocytes and epithelioid myofibroblasts. Diagnostic images are usually nonspecific and may look like breast carcinoma. At mammography, it appears as an area with radiodense and homogeneous tissue. On the other hand, ultrasonography may show an irregular, hypoechoic nodule with a posterior acoustic shadow. Due to their similarity, neoplasms must be discarded. The therapeutic approach consists of controlling glycemic levels and treating symptoms. The use of antibiotic therapy may be necessary in case of infection [10].

Sarcoidosis

Sarcoidosis is a systemic inflammation that mainly affects people of European or African-American descent, whose etiology is still largely unknown. Characterized by the presence of non-caseating granulomas, it mainly affects the lung parenchyma and lymph nodes, with breast cases being very rare. It is usually self-limited and in most cases resolves within a few months [11].

When it affects the breast, it manifests as a mobile nodule with a hardened consistency, similar to cancer. Mammography and ultrasound findings are nonspecific. Chest radiography may show bilateral hilar adenopathy and pulmonary nodules. The treatment is symptomatic, since in most cases it resolves spontaneously. Anti-inflammatories can be used to control the inflammatory process [11].

Actinic mastitis

Resulting from radiotherapy lesions in the treatment of breast cancer, actinic mastitis is characterized by inflammatory lesions that last for several years. Treatment with radiation progressively affects the intima of the vessels, leading to inflammation that manifests itself with redness, skin edema, pain, hyperemia and increased redness. The approach to actinic mastitis includes the use of nonsteroidal anti-inflammatory drugs for 3 to 5 days and hydration of the affected skin.

Mastitis due to disseminated lupus erythematosus

Systemic lupus erythematosus is an autoimmune disease that affects several organs. Among its multiple manifestations, it can also affect the breast. When this happens, breast inflammation can mimic an infection. It usually affects women in menacme with a previous diagnosis of SLE [12].

As in other cases of chronic mastitis, the presence of firm nodules can mimic breast neoplasms and these should be ruled out by diagnostic imaging and histopathological methods, if necessary. Histologically, there is lymphocytic infiltrate, hyaline adiponecrosis, microcalcifications and lymphoid nodules. On mammography, one can see nodules associated with calcifications, related to steatonecrosis and generally with lymph node enlargement. Ultrasonography demonstrates architectural distortion associated with a hypoechoic nodule with posterior acoustic shadowing [12].

Lupus mastitis is usually accompanied by other characteristic manifestations of the disease. The treatment is clinical related to the control of inflammatory reactions and therapeutic approach of Systemic Lupus Erythematosus [12].

Oleogranulomatous mastitis

Oleogranulomatous mastitis results from the ingestion of a foreign body in the breast in order to increase breast volume, and may occur as a result of the use of liquid paraffin, industrial silicone, gel, beeswax and others.

The foreign body undergoes typical reactions, such as encapsulation and fibrosis. There is an intense inflammatory reaction with pain, hyperemia, edema and breast swelling. Mastitis can evolve with fistulas, abscesses and even necrosis.

Mammography may show high-density fibrous capsule nodules. Ultrasound can identify calcified oil cysts and nodules with parenchymal distortion.

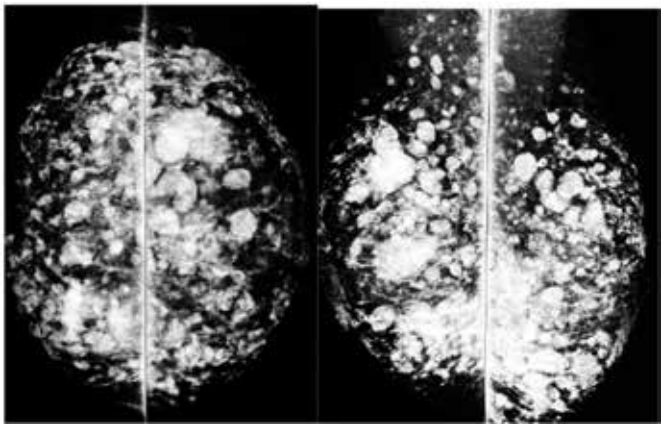

Figure 12 – Mammography. Oleogranulomatous mastitis. Presence of multiple bilateral calcified cysts in a woman who underwent injection of industrial silicone.

CONCLUSION

In conclusion, non-puerperal chronic mastitis can have a very varied etiology and be classified as infectious or non-infectious. In general, it has a benign character and low recurrence. In any case, diagnosis through careful physical examination and complementary exams become even more important to approach the disease properly, due to its rare nature and since mastitis can often mimic a more serious disease, such as neoplasms. In addition, the varied etiologies determine good knowledge so that the case can be approached properly.

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ANATOMICAL ANOMALIES OF THE BREAST

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ABSTRACT

Anatomic breast anomalies are alterations in the structural composition of the breast.
Objectives: The following work aims to list the main deformities, highlighting the importance of knowledge about the anatomy of this organ.
Results: The analysis of the literature makes it possible to divide, didactically, breast anomalies into numerical or structural, being generally originated during embryogenesis or pubertal development. Diagnosis and treatments are based on individual history, taking into account female anxieties and concerns.
Conclusion: anatomical abnormalities of the breasts are conditions that directly affect the physical and emotional aspects of women and should therefore be thoroughly investigated.

KEYWORDS: ANATOMICAL ANOMALIES; BREAST; NUMERICAL ANOMALIES; STRUCTURAL ANOMALIES; DIAGNOSIS.

INTRODUCTION

The breasts are external organs of the female reproductive system, whose glandular structure is responsible for the secretion of breast milk, intended for the nutrition of the offspring. Anatomically, they have a strong symbolism linked to self-image, the construction of identity and sexuality, especially for women, representing a symbol of femininity[1]. In this context, it is clear that anatomical anomalies of the breasts, which may be numerical or structural in nature, and associated body distortions have a major impact on health and quality of life, especially for females, both in terms of physical as well as psychosocial aspects.

Based on this assumption, knowledge about the main anatomical abnormalities of the breasts is of paramount importance for targeted attention and comprehensive care for women, understanding the main complaints and insecurities that are often associated.

Breast Structure and Development:

The breasts are arranged bilaterally on the anterior wall of the thorax, in the adipose tissue of the subcutaneous layer underlying the skin. On its outer portion, each breast has a conical projection called the papilla and a thick, reddish-brown circular region, the areola. Internally, it is composed of a glandular parenchyma and a fibrous stroma, the first being a set of numerous secretory lobules united in lobes, which have lactiferous ducts that carry secretion to the papillary surface. The stroma, on the other hand, consists of the suspensory ligaments of the breasts (bands of dense connective tissue originating from the dermis that surround and intersect the lobes), in addition to layers of loose connective tissue that separate the mammary complex from the underlying pectoral muscles[2].

From this, it should be noted that most anatomical changes in the breasts are correlated with an anomalous development at the embryonic or pubertal level:

As far as embryogenesis is concerned, in the fourth week of intrauterine development, a pair of epidermal thickenings called mammary ridges form along each side of the body of the embryo from the area of the future axilla. Although much of the mammary line disappears shortly after its formation, a small portion in the thoracic region persists and penetrates the mesenchyme. Thus, in view of the structures involved in this stage, this involution is essential for breast development, so that, if altered, it can lead to the appearance of some numerical anomalies, such as polymastia and politelia. By the seventh week, the remnant of the mammary crest produces a well-defined primary bud of the mammary gland, which grows towards the dermis and the adipose tissue. In the tenth week, the bud starts the branching process, forming between 16 and 24 smaller units. These resulting epithelial sprouts elongate and multiply throughout pregnancy, and at the end of prenatal life, they are channeled and form the lactiferous ducts, which open into a small fossa. Shortly after birth, this pit transforms into the papilla by proliferation of the

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underlying mesenchyme. The surrounding skin also proliferates and thickens to form the areola. Thus, developmental dysfunctions at this stage can also generate anatomical anomalies of the organ, such as atelia and inverted nipple[3].

At puberty, increasing concentrations of estrogen and progesterone stimulate the development of the glandular parenchyma through budding and elongation of the ducts, forming alveoli and inactive secretory cells. In addition, there is intense deposition of adipose tissue, which essentially reflects the volume of the breasts[4]. Thus, attention is needed for possible anatomical anomalies of the breasts that arise during this period, such as hypomastia, gigantomastia and breast asymmetry.

Diagnosis

The diagnostic investigation of anatomical breast anomalies must have a special concern with the anguishes and apprehensions expressed by the patient. The physician must pay attention not only to the physiological impact generated by the disease, but also to the social and emotional impacts caused.

During the anamnesis, symptoms such as pain, changes in size, presence of masses and secretions, asymmetries in shape, changes in the nipple and inflammatory signs must be well characterized and defined. Its investigation should focus on the time of emergence and the mode of evolution; its relationship to menstrual periods, pregnancy, or surgeries; menstrual history; the use of drugs, especially contraceptives; and its relationship with other symptoms or findings[5].

The physical examination should be thorough, with breast inspection and palpation. Anatomical anomalies, such as politelia, can be easily confused with nevi and other skin lesions, while gigantomastia and breast asymmetries can be mistakenly associated with neoplastic masses[6].

Imaging exams provide great help, especially in diagnostic elucidation and malignancy exclusion. Often, mammography is the method of choice for screening, however, it has certain limitations, such as the approach to dense breasts and the young age group of patients. In addition to this exam, investigation via ultrasound is highly indicated, with several advantages for breast evaluation. It is a methodology that does not use ionizing radiation, and has better applicability in these morphological changes, considering the target audience of young patients, in the pubertal period, with parenchyma rich in fibroglandular tissue[7].

Main Numerical Anatomical Anomalies:

Polythelia

Polythelia is defined as the presence of supernumerary nipples without the presence of associated parenchymal tissue, that is, without mammary glands. This anatomical anomaly is caused by the non-regression of the mammary line during embryogenesis[3]. Commonly confused with dermatological spots or nevi, supernumerary nipples occupy, as a rule, the anterior region of the thorax, lying between the axillary and inguinal regions (Figure 1). Generally, politelia may be related to organic dysfunctions of the urogenital tract6.

Polythelia can be treated surgically by excision of the supernumerary nipple, especially when its presence causes aesthetic discomfort to the patient, impairing his emotional health.

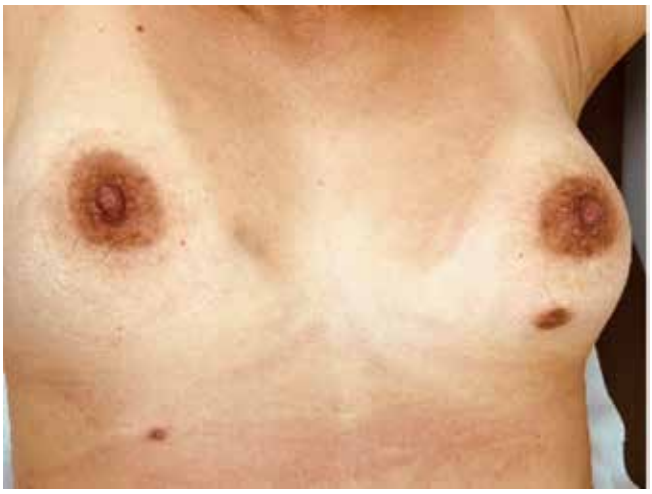

Figure 1 – Polythelia. Supernumerary nipples in the right inframammary region and in the lower medial quadrant of the left breast.

Polymastia

Polymastia, also called supernumerary breast tissue, is a condition in which there is the presence of glandular parenchymal tissue in ectopic regions. Mostly found in the armpit, chest wall, inguinal region and, more rarely, in the vulva, their cause is the non-regression of the milk lines during embryogenesis[3] (Figure 2).

During childhood, there are no major clinical manifestations caused by this anatomical anomaly. However, in the pubertal phase and also in the pregnancy-puerperium cycle, polymastia often begins to present itself as a region of edema with a feeling of heaviness and, sometimes, pain. Hormonal stimulation, especially due to the production of gonadotropins and sex hormones, leads to proliferation of both the stroma and the supernumerary glandular parenchyma, leading to the development of the accessory breast.

The accessory breast is made up of the same structures that make up the physiological breast tissue, therefore, it can be affected by the same pathological processes, benign and malignant, that compromise the normal breast. Thus, the diagnosis of fibroadenoma, cyst, infection and carcinoma may be present in the supernumerary breast, so that imaging investigation is essential for the correct diagnosis and treatment[7].

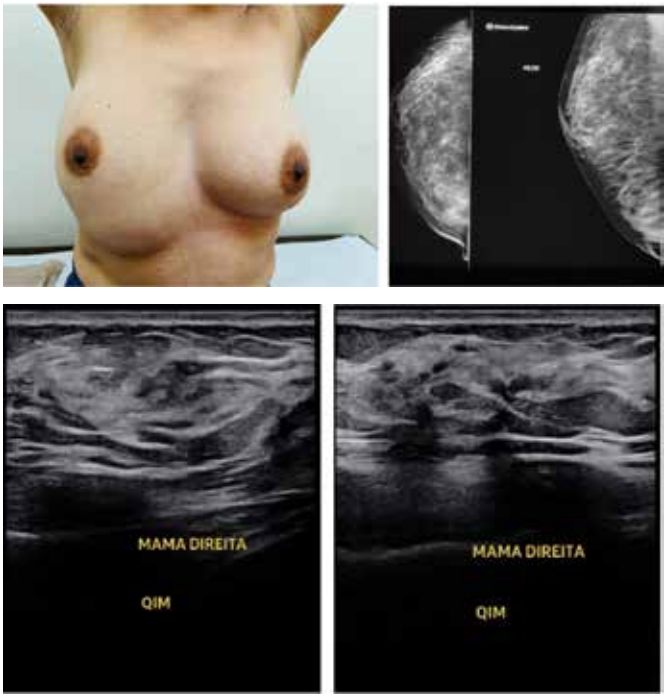

Figure 2 – Polymastia. A. Photograph of a woman with polymastia in the lower medial region (LIQ) of the right breast. B. Mammography in CC and MLO views showing accessory fibroglandular tissue located in the LIQ of the right breast. C and D. Ultrasonography showing accessory fibroglandular tissue (polymastia) in the LIQ of the right breast.

Amastia

Amastia is defined as the complete absence of the glandular structure that constitutes the breasts. This condition can be present unilaterally or bilaterally (Figure 3) and have its etiology in iatrogenic or congenital causes[6]. Caused by a dysfunction during the process of breast formation in embryogenesis, such as the failure of the ectoderm to invaginate towards the mesoderm, this anatomical anomaly can be accompanied by a series of malformations of other structures originating from these leaflets, such as skin, hair and the muscles, especially the pectoralis major[3].

In these cases, the use of imaging tests is fundamental for the correct diagnosis and clinical management. Magnetic resonance imaging is an interesting exam for analyzing the integrity of structures, such as the ribs and pectoral muscles; while ultrasonography is an important test for confirming cases of breast mass, attesting to the absence of underlying breast tissue and ruling out other diagnoses, such as breast hypoplasia[7].

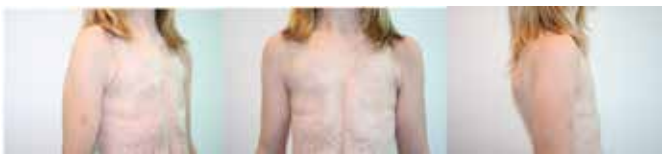

Figure 3- Amastia in a young patient.

Amazia:

Amazia is characterized by the absence of parenchymal tissue in the breast, with the presence of a nipple-areolar complex (Figure 4). The picture arises as a result of a defect in the formation of the mammary parenchyma and stroma during embryogenesis[3]. Due to the non-commitment of the nipple, the diagnosis may be impaired during childhood, only becoming noticeable with the entry into the pubertal period.

Ultrasonography reveals retroareolar tissue with a fibrotic appearance, local adipomastia, and absence of mammary glandular parenchyma.

This lack of breast growth and development, associated with the common development of the nipple and areola, is usually correlated with other congenital anomalies, such as the presence of cleft palate, saddle nose and piriform hypoplasia[8].

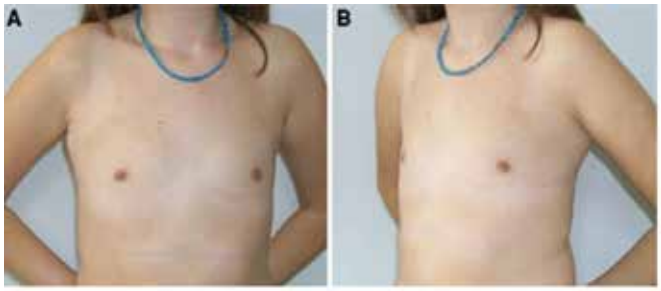

Figure 4- Amazia. Characterized by the absence of glandular parenchyma in the mammary structure.

Atelia:

Atelia is characterized by the absence of the nipple-areolar complex in the presence of mammary parenchyma and stroma (Figure 5). Although normally this anatomical anomaly is associated with trauma or iatrogenic events, its congenital form can also appear, especially when associated with amastia.

Currently, there are specific surgical techniques for the construction of a new structure of the areola and papillary structure, through the implantation of dermal flaps. Furthermore, alternative reconstitution through tattooing or micropigmentation is not uncommon[6].

Figure 5- A) Atelia. Represented by the absence of nipple-areolar structure, with stroma and glandular parenchyma present. B)

Main Structural Anatomical Anomalies:

Hypomastia:

Hypomastia, also called mammary hypoplasia, is an anatomical breast anomaly noticed after the onset of puberty and characterized by poor development of the glandular parenchyma and stroma that make up the organ[6]. (Figure 6)

Hypomastia is caused by a decrease in estrogen production by the body or the insensitivity of the breast tissue to recognize and respond to the produced and circulating hormones. Thus, it is, as a rule, an anomaly secondary to other dysfunctions such as gonadal dysgenesis or pituitary hypogonadism[4].

Ultrasonography is the preferred method for evaluating hypomastia, especially due to the reduced size of the breasts, making it difficult to use mammography. This anomaly is diagnosed, as a rule, under the age of 30 years. The examination reveals breasts with poorly developed glandular parenchyma and little support tissue[7].

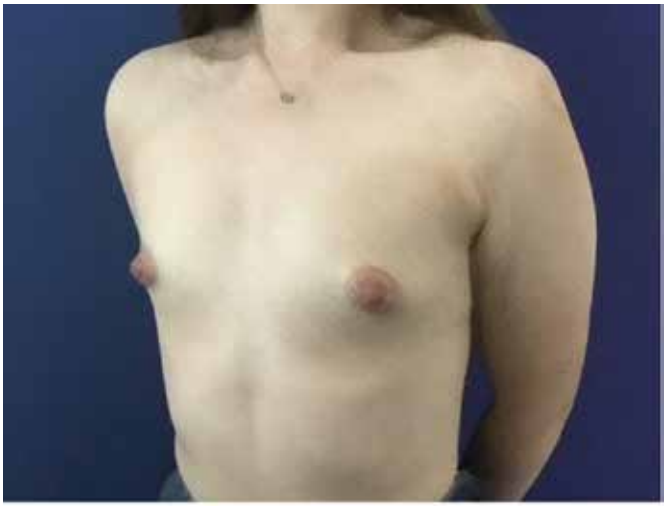

Figure 6- Hypomastia in a young patient. Areopillary development considered normal, with small parenchymal and stromal proportions.

Breast hypertrophy:

Mammary hypertrophy is an abnormal increase in the size of the breasts (Figure 7). Its extreme form, gigantomastia, is defined as dysfunctional and disabling breast enlargement that requires an average reduction of 1500 grams per organ for correction. Its occurrence is more common in pregnant women and in women in the pubertal period, and may even have an idiopathic or pharmacological character[6].

The pathophysiology of this anomaly is generally associated with an increase in circulating levels of steroid hormones, such as occurs in pregnancy, puberty and the use of medications; or to an increased sensitivity of breast tissue to physiological levels of these substances. However, regardless of the etiopathogenesis, the condition must have a clinical investigation based on differential diagnoses, such as lipoma, fibroadenoma, phyllodes tumor and cancer. In this context, the use of imaging tests is fundamental for the search for tissue alterations that help to rule out the hypothesis of malignancy. Breast pain, nipple-areola complex ulcerations, low back pain, inflammatory abscesses and psychological disorders associated with self-image are common[9].

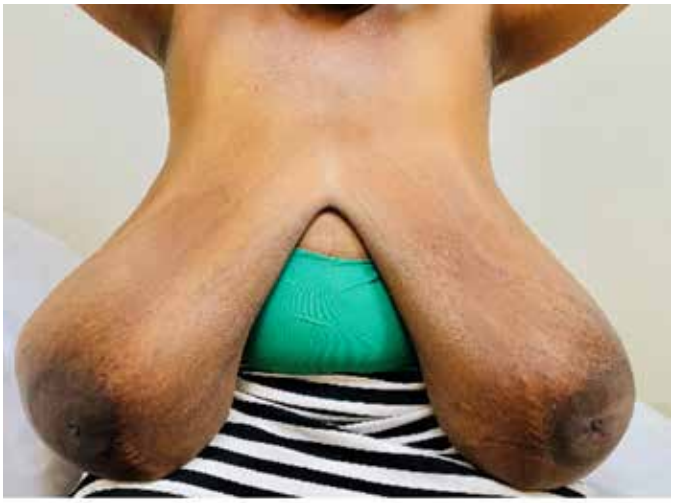

Figure 7 – Bilateral breast hypertrophy in a postpartum woman.

Asymmetry:

Breast asymmetry must be differentiated from a purely physiological condition that is common to women, after all, the majority of the female population has some degree of variation in the size of the breasts. However, in some cases, it can be configured as pathological[6]. (Figure 8)

When the discrepancy in the size of the breasts becomes large, it can become an aesthetic problem that impacts the quality of life of the woman, requiring a medical approach. Yet another reason that leads to the need for better clinical investigation is the fact that voluminous breast asymmetries may be a manifestation of other diagnoses that are causing the observed size variation, such as fibroadenomas, cysts and carcinomas.

The expected increase in breast volume at puberty can be affected by a series of factors that lead to asymmetry, such as genetic influences on the higher concentration of hormone receptors in one of the breasts, trauma, infections and iatrogenic or growing nodules and masses[10].

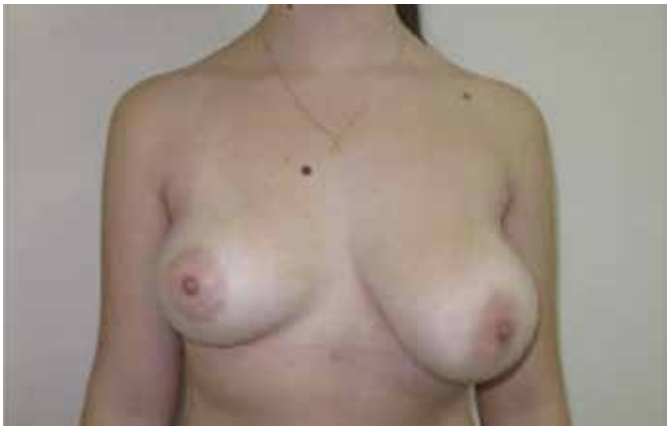

Figure 8- Physiological breast asymmetry.

Inverted Nipple:

Nipple inversion is an anatomical anomaly, congenital or acquired, characterized by the transposition of the entire nipple towards the subareolar region, inside the breast. Developed due to a proliferation of fibrous tissue between the nipple and the subareolar parenchyma, this inversion can have different etiologies.

Although congenital presentations (Figure 9) of inverted nipple do not have major clinical consequences beyond the aesthetic deformity, this anatomical anomaly, when acquired, must be thoroughly investigated and detailed, seeking to identify the benign or malignant nature of the lesion. Other manifestations such as eczema, pain, secretions, loss of sensitivity and nipple stiffness may be associated with the condition.

It is relevant to analyze the presence of bilaterality of nipple inversion: bilateral inversion is a finding suggestive of congenital anomaly and benignity, whereas unilateral inversion is an indication that there may be an associated malignant tumor lesion. In this same analysis, the time elapsed by the inversion indicates that acute anatomical alterations generally occur with benign processes, with an infectious aspect, while chronic variations are more suggestive of neoplasms[11].

Figure 9– A and B. Right inverted nipple. Benign congenital anomaly.

Tuberous breast:

The tuberous breast is a congenital anatomical anomaly that becomes noticeable at puberty, through the neuroendocrine stimulus on the breasts. This deformity is characterized by hypoplasia and tissue narrowing at the base of the breast, constricting the site and leading to vertical and horizontal underdevelopment of the entire structure. Thus, the breast acquires a tubular cylindrical format anteriorly, at the same time that the glandular parenchyma undergoes herniation in the area of the nipple-areolar complex, leading to areolar dilation6 (Figure 10).

As for its pathophysiology, the exact mechanism is still uncertain. It is believed that the deep fascia of the breast, located between the dermis and the pectoral musculature, has greater thickening and a more resistant adherence to the regions at the base of the breast, inducing the observed narrowing and hypoplasia. Additionally, the fibrous ring formed restricts the radial growth induced during puberty, favoring development towards the nipple, where the absence of resistance and adequate support on the part of the superficial fascia causes an areola enlargement, typical of the tuberous breast[12].

Present, as a rule, in young women, although it may also be associated with cases of gynecomastia in the male population, the main negative impact of tuberous breasts is on aesthetic discomfort. Due to the narrowing of the local parenchyma and the alteration of the nipple, this condition can still impair the breastfeeding process. The proposed treatment for this anatomical anomaly is surgical correction[12].

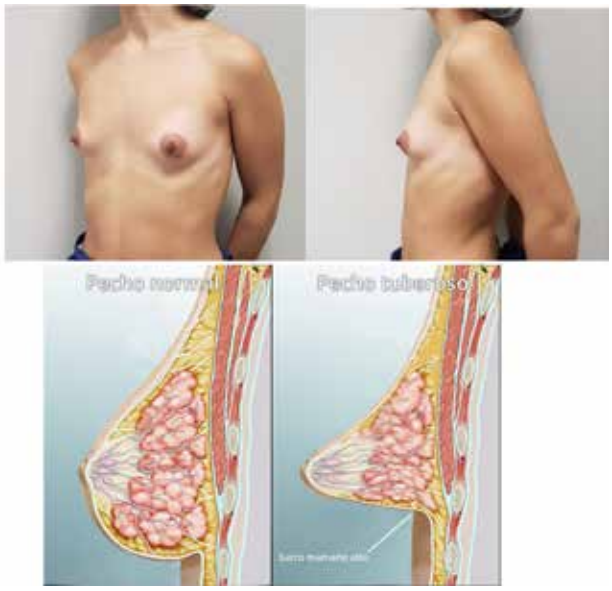

Figure 10- A) Tuberous breast. B) Schematic representation of the typical mammary structure of the tuberous breast.

Symastia:

Symastia is an anatomical anomaly characterized by the medial confluence of the breasts. It is a condition of congenital or acquired origin, in which a medial connec-

tion of the breasts appears, with the presence of an intermammary membrane on the mid-sternal line, uniting both structures[6].

The most common form of this anomaly is iatrogenic (Figure 11). It is not uncommon for patients to develop symptoms of symmastia after breast augmentation, with the placement of breast implants, or reduction, with the removal of local tissue excess. In this scenario, the dysfunction is linked to the dissection of the midsternal fascia, responsible for joining the skin to the bone, allowing the healing process to penetrate the created space, uniting the breasts.

In the background, with rare cases recorded, simastia can also be configured as a congenital anomaly. In this condition, a malfunction in mammary embryogenesis leads to the formation of a web of supporting glandular parenchymal and connective tissue, uniting the breasts medially over the sternum region.

The therapeutic approach is surgical and is based on the removal of the tissue that composes the intermammary web, with the fixation of the dermis to the sternal periosteum and with the postoperative use of compressive pads in the region of the intermammary fold created on the sternum[13].

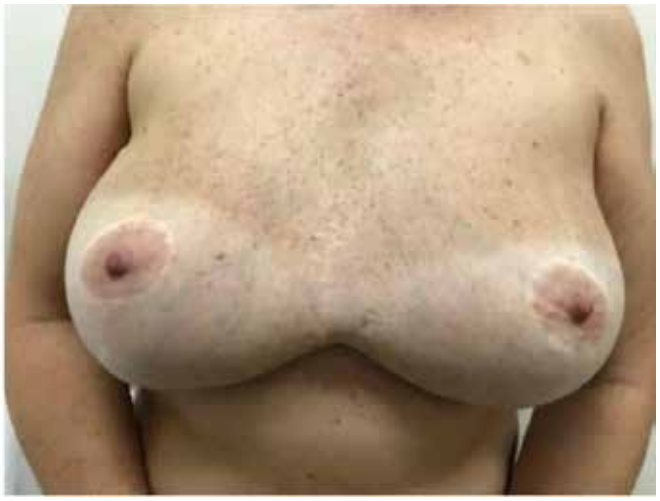

Figure 11 – Iatrogenic symmastia resulting from mammoplasty and exeresis of cutaneous melanoma in the region of the intermammary sulcus.

CONCLUSION

The anatomical integrity of the breasts is an aspect of great physical and emotional importance for women, considering the strong biological and feminine symbology that these organs have. The main breast anomalies, which may be numerical or structural in nature, commonly derive from embryological inconsistencies or hormonal dysregulations, especially during puberty. Thus, a thorough investigation of cases of breast deformities is relevant in order to determine an accurate diagnosis, rule out possible malignancies and ensure adequate treatment. Furthermore, active and comprehensive listening to women's insecurities and fears about such anomalies are essential behaviors for effective care.

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PAGET'S DISEASE OF THE BREAST

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ABSTRACT

Paget's Disease of the Breast (PDB) is a type of adenocarcinoma that affects the areola-mammary complex. Its unusual presentation can be confused with inflammatory or dermatological conditions. PBD is associated with breast neoplasms and is characterized by the presence of Paget cells. There are theories about its origin, such as trophoepidermal and intraepidermal transformation. PDB is more common in women between 50 and 60 years old, usually associated with carcinoma in situ or invasive lesions. Diagnosis is difficult and can take years, requiring a biopsy for confirmation. Treatment involves surgery and radiotherapy, especially in advanced cases. Late diagnosis can impact the prognosis and quality of life of patients.

KEYWORDS: DIAGNOSIS; PAGET'S DISEASE MAMMARY; NEOPLASMS.

INTRODUCTION

Diseases with an unusual presentation are quite worrying in clinical practice and tend to cause concern in affected individuals, such as Paget's disease of the breast (PDB). This condition is characterized as a malignant entity with benign characteristics that affect the areola-mammary complex, being considered a type of adenocarcinoma, which may or may not be associated with cases of breast cancer (Figure 2)[1]. Its benign behavior can make early diagnosis difficult, as its presentation mimics inflammatory conditions or even dermatological disorders[2].

The name of the condition came from the discovery of James Paget, in 1874, who was a British surgeon and pathologist, who first described Paget's Disease with presentations in different places, which could be bone, mammary and extra mammary. Since the beginning of the discovery, there has been a correlation between the entity and neoplasms, such as the mammary gland[3]. Currently, PDB is known as a rare malignant skin condition of intraepithelial origin, characterized by the presence of Paget cells, which are large epidermal cells of adenocarcinoma, which lead to desquamation of the nipple epithelium[4].

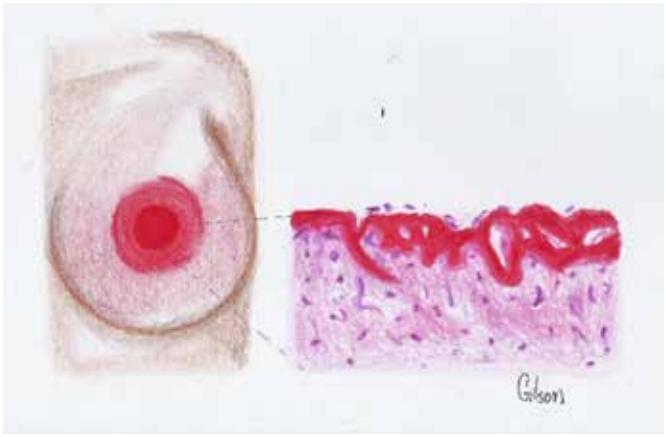

Figure 1 – Schematic representation of a breast with normal skin showing the layers of keratin, epidermis and dermis. Source: The author (2023)

Figura 2 - Desenho de mama esquerda apresentando Doença de Paget de aréola e mamilo. Fonte: O autor (2023)

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The presentation, usually confined to the nipple, is described as eczematous lesions in the periareolar region, which progress to bleeding or even growth of masses in the underlying tissues5. Thus, when faced with this pathological profile, many physicians mistakenly end up diagnosing the lesion as benign dermatitis, which can delay treatment and worsen the prognosis of young women[6]. The extra mammary or mammary description refers only to the site of appearance of the lesions, however, the histopathological examination reveals vacuolated squamous cells with bluish cytoplasm, which are highly infiltrated and diffuse[7].

Thus, the present study aimed to review some concepts related to Paget's disease, such as pathophysiology, epidemiological profile, tools for diagnosis and treatment.

PATHOPHYSIOLOGY

There are currently theories to explain the appearance of Paget's Disease, however, there are still factors that still do not make clear what the real explanation for the appearance of this condition is.

a) TROPHOEPIDERMAL THEORY

The trophoepidermal or epidermotropic theory concerns the transformation of Paget cells, which originate in the duct of apocrine glands, into underlying ductal carcinoma cells that migrate to the epithelial tissue of the nipple through the basement membrane[8]. This theory is supported by the high incidence of PDB in people with ductal carcinoma in situ and by the high level of expression of the HER2/neu oncogene in Paget[4] cells, suggesting that keratinocytes synthesize heregulin-alpha, a motility factor that attracts Paget for nipple[3].

b) INTRAEPIDERMAL ORIGIN OR TRANSFORMATION THEORY

It is suggested that the Paget cell is a pluripotent keratinocyte that has gone through the process of malignancy. This suspicion arises from the rare cases in which there is no malignancy adjacent to the nipple tissue[8]. This theory is based on the morphological similarity of Paget cells and Toker cells (benign epithelial cells of sebaceous glands present in the areolar skin in 10% of women)[4].

Figure 3 – Paget's disease in the left breast, showing erythematous squamous plaque affecting the entire areola and nipple, exceeding the areolar limits.

EPIDEMIOLOGY

The epidemiological profile of PDB seems to be related to female patients, aged between 50 and 60 years, with a worse prognosis when the condition occurs in males9. PDB can appear in isolation, without association with other conditions, however, in most cases, in about 32 to 41% of cases, there is a correlation with carcinoma in situ or more invasive lesions10. It is present in 1 to 3% of cases of primary breast cancer, with 93 to 100% of these associated with underlying breast cancer, commonly in central and multifocal tumors, predominantly located near the areola[4].

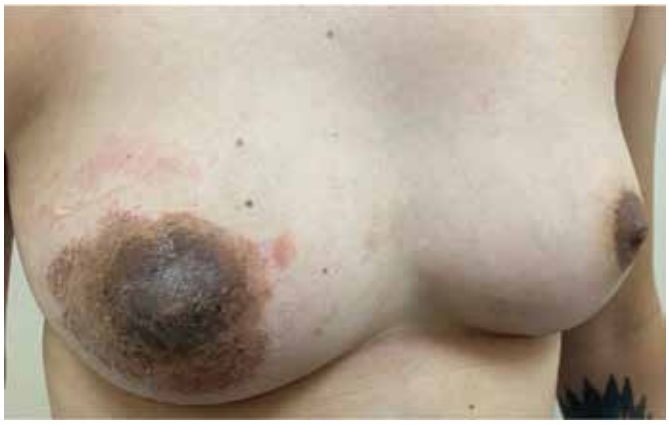

Figure 4 – Right breast eczema showing desquamative lichenified plaque partially affecting the nipple-areolar complex. Treated with topical corticosteroid therapy. Source: The author (2023).

DIAGNOSIS

Because it is a rare and poorly studied condition, PDB is difficult to diagnose, and it can take years for a professional to identify and treat the disease correctly. In the literature, there are reports of cases that had a delay between 10 and 15 years in diagnosis[7]. Initially, PDB appears as an irritation, rash or crack in the nipple, which may take months or years to reach the areola and periareolar region, evolving as an erythematous and scaly disease11. In more advanced cases, there may be skin ulceration and nipple retraction, the latter being a pathognomonic sign of PDB[11]. There may also be cases of hyperpigmented lesions that can be confused with melanomas[8].

Therefore, PDB is commonly identified with eczema, dermatitis or psoriasis, leading to an ineffective topical treatment, which delays the diagnosis of the disease. Therefore, to avoid misdiagnosis, when faced with eczematous, pigmented, crusted lesions or with signs of chronic inflammation in the nipple, the most appropriate conduct is to perform a biopsy[4].

The nonspecific presentation, associated with the epidemiology of the disease, makes diagnosis difficult in most cases. For this reason, the protocol for diagnosis should begin with a good physical examination, performing inspection maneuvers and palpation of the breast tissue

and, in this case, the areolas and nipples. Inspection begins with the assessment of lesions that may or may not be clearly present. For this, the use of a dermoscope is indicated for enlargement of the area and careful investigation. However, the final diagnosis must be made by collecting material for histopathological analysis. As previously described, in the case of PDB, vacuolated squamous cells with diffuse tissue invasion and bluish cytoplasm will be seen, thus characterizing the presence of Paget cells[4,7].

TREATMENT

The surgeries are mainly indicated when there is an association of PDB with in situ carcinomas, and breast segmentation may be performed in case of early diagnosis. However, due to epidemiological characteristics and late diagnosis, in many cases radical treatments are required, such as mastectomy, followed by radiotherapy for better prognosis of patients[11].

Segundo a revisão sistemática de Lin et al (2022), mastectomia ou cirurgia conservadoras da mama associado ao tratamento radioterápico obtiveram os melhores prognósticos em relação à reincidência, metástase e taxa de mortalidade[12]. Porém, a abordagem conservadora sem radioterapia obteve uma taxa significativa de recidiva para DMP, sendo recomendado principalmente a associação dos tratamentos quando há presença de lesões de carcinomas in situ ou de perfil invasivo[12].

According to the systematic review by Lin et al (2022), mastectomy or breast-conserving surgery associated with radiotherapy treatment obtained the best prognosis in terms of recurrence, metastasis and mortality rate12. However, the conservative approach without radiotherapy obtained a significant rate of relapse for PDB, and the combination of treatments is mainly recommended when there are carcinoma lesions in situ or with an invasive profile[12].

CONCLUSION

It can be concluded, therefore, that the difficulties in diagnosing PDB are mainly related to its clinical presentation, as well as to the epidemiological profile of the condition. These factors may hinder treatment with more conservative approaches and directly impact the quality of life of patients. Thus, when diagnosed late, the therapeutic association between surgical and radiotherapeutic methods is strongly recommended.

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CEREM-GOIÁS

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