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CASE REPORT

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LEFT AXILLARY SCHWANNOMA WITH GANGLIONAL INVOLVEMENT OF MALIGNANT APPEARANCE: A RARE CASE REPORT

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ARTICLE INFO	ABSTRACT
Article History: Received 14 th March, 2024 Received in revised form 20 th April, 2024 Accepted 17 th May, 2024 Published online 28 th June, 2024	Schwannomas are tumors, generally benign, originating in the Schwann cells of peripheral nerves. These tumors are usually located in the head, neck, arms, legs, body and chest, while axillary involvement is uncommon. The etiology of schwannomas is uncertain, but it is believed to be related to metastatic diseases, radiotherapy and advanced age. Symptoms are often absent or present as a tumor with paresthesia and/or radiating local pain, making early diagnosis difficult. That said, the case reported below is unique and describes a 43-year-old female patient, with the presence of an expanding left axillary nodule, with no history of previous illnesses and previous surgical or radiotherapy procedures, making the case unusual. Diagnosis was only possible with immunohistochemical analysis after tumor excision, confirming the diagnosis of malignant axillary schwannoma with lymph node involvement, requiring adjuvant chemotherapy and radiotherapy. In the literature there are only four reports of malignant schwannomas in the breast, highlighting the importance of an early diagnosis and effective therapeutic method, given the difficult diagnosis and rarity of the case.
Key Words:	
Schwannomas are Tumors, Unique and Describe, Diagnosis.	
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INTRODUCTION

Schwannoma was first described in 1908 by Verocay, being characterized as a neoplasm developed from the nerve sheath on the periphery of Schwann cells. Collins, in 1972, reported the first case of schwannoma of breast location, this tumor of non-epithelial tumoral origin of the breast is extremely uncommon, representing 2-3% of all schwannomas, with only 0.2% of all breast tumors being schwannomas (Hassouna et al., 2006). The axillary location represents 5% of all schwannomas, malignancy is even rarer, corresponding to between 5-10% of all sarcomas malignant, mainly in males (Hassouna et al., 2006) (Lucca et al., 2021). The risk factors for developing the schwannoma are not well established, however it is known that a history of previous radiotherapy, neurofibromatosis and advanced age predispose to the disease (Fritsche et al., 2023) (Sordillo et al., 1981). Our report describes a female patient, 43 years old, with axillary schwannoma and ganglionic involvement with a malignant aspect, without any risk factor for the development of the neoplasia, making the report extraordinary. They can occur at any age, but the age of greatest incidence is in people over 40 years of age, with no predilection for sex or ethnicity (Sengul et al., 2019). Diagnosis is generally late, as signs and symptoms are confused with a series of lesions benign and malignant, such as fibroadenomas, phyllodes tumor, mesenchymal neoplasms or breast cancer (Uchida et al., 2005). Although rare and difficult to diagnose, the possibility of malignancy and possible metastasis demonstrates the importance of

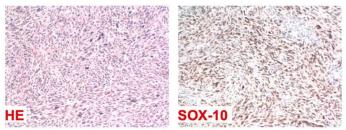
studying this pathology, proving the need for an early diagnosis, rapid and aggressive therapy. The literature records approximately 37 cases of breast and armpit neurilemmoma, most of them with benign evolution, with only 4 reports of malignant schwannomas in the breast. To our knowledge, this is the only report of an axillary schwannoma with malignant lymph node involvement, which demonstrates satisfactory clinical and laboratory evolution (Roncatti *et al.*, 2019).

CASE REPORT

Female patient, 43 years old, two pregnancies with two vaginal births, denies previous comorbidities, denies previous surgeries and continuous medication use, has a family history of a sister with breast cancer. A patient was admitted to the hospital in March 2018, complaining of a palpable nodule in the left armpit. An investigation was carried out with mammography revealing BI-RADS 2 with benign changes, an ultrasound that showed a nodular, heterogeneous image, suggestive of an atypical lymph node, with a displaced hyperechogenic center with an irregular appearance, measuring approximately 4.2 cm in largest diameter, and other lymph nodes of atypical appearance up to axillary level. In addition, an MRI was performed, showing confluent nodular lesions in the axillary region, with regular contours, presenting an intermediate signal on T1, high on T2, with contrast enhancement, involving the vascular-nervous bundle, but without signs of compression, which must correspond to

lymph node enlargement. FNAC did not show signs of malignancy, however, as the nodule continued to expand, axillary tumorectomy was chosen. The procedure was uneventful and she patient was discharged from hospital on the second postoperative day, need for immunohistochemistry. After surgery, she patient continued outpatient follow-up with a clinical oncologist, who requested laboratory tests, serology and immunohistochemistry, which were normal: Cytomegalovirus IgG >250 and IgM 0.37; Hepatitis B o.21; Hepatitis C 0.04; Epstein barr virus IgG 62.71 and IgM 0.20; HIV negative. However, the immunohistochemical study confirmed the diagnosis of axillary Schwannoma, with findings compatible with malignant neoplasia (Image 1).

Figure 1. Immunohistochemistry study



Subtitle: metastatic high-grade spindle cell/epithelioid malignancy, expressing S-100 protein positivity, focally positive smooth muscle actin, positive SOX10 and positive Histone.H3.3K27me.

Therefore, adjuvant chemotherapy and local radiotherapy were chosen. The patient maintained follow-up with the multidisciplinary team, receiving care from a psychologist, nutritionist, oncologist and breast specialist. Postoperatively and during treatment, she patient reported pain and edema in the left arm, and was prescribed gabapentin, diosmin 450/50 and advised lymphatic drainage and the use of a compressive glove for edema prophylaxis. In addition, she was patient underwent computed tomography scans for reevaluation after chemotherapy treatment, after the end of treatment the patient underwent CT control every 4 months, then every 6 months and finally annually, associated with laboratory, mammography and axillary ultrasound as per control with tomography, transthoracic echocardiogram, electrocardiogram and laryngoscopy were also requested for monitoring. Currently, the patient is in good general condition, with no complaints or sequelae related to the schwannoma, denies taking continuous medications and denies comorbidities linked to the neoplasm. The patient undergoes annual follow-up at the mastology outpatient clinic and undergoes an annual screening mammogram.

DISCUSSION

Schwannoma or Neurilemoma is an oval, slow-growing, firm, solitary, well-circumscribed and encapsulated tumor, generally benign, measuring on average 2.25 cm in diameter, with the presence of large nodules, as well as multiple nodules, being rare (Hassouna et al., 2006). They originate in Schwann cells and frequently affect intracranial or peripheral nerves, commonly affecting the head, neck, trunk and extensor surfaces of the extremities, with axillary involvement being rare. It can happen at any age, but it frequently occurs during the third decade of life, with an average of 48.6 years, with no predilection for sex or ethnicity. (Grupta et al., 2000) (Hassouna et al., 2006). The risk factors for developing schwannoma are not well established, however it is known that a history of previous radiotherapy, neurofibromatosis and advanced age predispose to the disease (Fritsche et al., 2023). Most patients are asymptomatic, however dysesthesia caused by palpation, sensory loss, weakness and radicular pain may occur. Late-diagnosed schwannomas can generate degenerative conditions, with nuclear pleomorphism, stromal edema, fibrosis and xanthomatous changes (Sengul et al., 2019). Tinel's sign, characterized by radiating pain caused by palpation of the affected nerve, is a typical sign of axillary schwannoma (Duehrokoop et al., 2021) (Kumagai et al., 2022). Schwannoma in the axillary region is very similar to swollen lymph

nodes on computed tomography, presenting a low signal on T1 and high signal on T2 and being able to observe, in some cases, the target signal on magnetic resonance imaging. On mammography, most schwannomas are described as a well-circumscribed area of round or oval opacification, which may have a density similar to soft tissue. On ultrasound, schwannomas present as solid lesions with a benign appearance, with a round or oval shape, well-defined margins and a hypoechoic pattern and may include the target sign. Fluorodeoxyglucose (FDG) positron emission tomography is a powerful test for detecting malignant tumors, but schwannomas also have an accumulation of fluordeoxyglucose, limiting the use of the test. Therefore, distinguishing between lymph node metastasis and axillary schwannoma using imaging studies alone is unfeasible (Fujiuchi et al., 2009) (Meira et al., 2023). Cytology by FNA fine needle puncture or CNB thick needle puncture for neoplasms in the axillary region is technically difficult and has low diagnostic accuracy, due to the similarity of cytology between the mesenchymal cells of the neoplasia and the smooth and fibromatous cells, in addition to providing risk of vascular or nervous injuries at the time of puncture (Kumagai et al., 2022) (Tan et al., 2014) (Uchida et al., 2005). Patients with axillary schwannoma present with severe radiating pain at the time of biopsy, which is a valuable finding and determines that the tumor originates in the nervous system (Kumagai et al., 2022)

The diagnosis is defined with an anatomopathological study after surgical excision of the nodule, revealing two types of tumor growth patterns, the Antoni A pattern with elongated cells with cytoplasmic processes arranged in fascicles in areas with moderate to high cellularity and scarce stromal matrix. The nucleus-free zones of the processes that extend between the nuclear palisade regions are called Verocay corpuscles. In the Antoni B growth pattern, the tumor is less densely cellular and consists of a loose meshwork of cells, microcysts, and myxoid stroma. In both areas, individual cells have an elongated shape and a regular ovoid nucleus (Meira et al., 2023) (Tan et al., 2014). Immunohistochemistry reveals the positivity of the S100 protein, a strong predictor for neoplasia (Sengul et al., 2019). Furthermore, through electron microscopy it is possible to visualize the deposit of the basement membrane involving single cells and collagen fibers, as the lesion displaces the nerve of origin to as it grows, allowing immunohistochemical staining for neurofilament proteins, showing the separation of the axons and the tumor, although they may be united by the capsule. Malignancy is histologically represented by peri- and intraneural amplification, with herniation into the vessel lumen (Gupta et al., 2000) (Parikh et al., 2016). The high incidence and lethality of breast cancer in the current population questions the importance of addressing the differential diagnoses of axillary neoplasms, since schwannomas are commonly considered a series of benign and malignant lesions, such as fibroadenomas, phyllodes tumors, mesenchymal neoplasms or even breast cancer (Uchida et al., 2005). Therefore, the consensus treatment is surgical excision of the tumor with preservation of the nerve trunk when possible, following some criteria such as: Size, tumor location, surgical access, growth rate, patient age, suspicion of malignancy and clinical evaluation of symptoms. Furthermore, possible surgical complications must be taken into account, such as neurological deficits due to nerve damage, seromas and hematomas (Hassouna et al., 2006) (Parikh et al., 2016) (Tan et al., 2014). In the case of the presence of malignancy, complete excision of the tumor must be ensured, in order to avoid local recurrences and metastases. In addition, adjuvant therapy after surgical resection has proven inconclusive, but has shown promising survival and low recurrence.

CONCLUSION

In conclusion, axillary schwannomas are rare, but should be considered in the differential diagnosis of axillary nodules. Surgical excision of the nodule is the most recommended treatment, however it is necessary to consider the patient's clinical condition, age, tumor growth, location, malignancy and the possibility of surgical and postoperative complications. Therefore, clinical monitoring is essential, both for an early approach and for biopsy of suspicious lesions, corroborating a favorable outcome. Finally, studies must be carried out to optimize early diagnosis and safe therapy, aiming for a lower risk of complications and better quality of life for patients suffering from axillary schwannomas.

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