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Cystic Schwannoma of Plexus Brachialis

Marco Mele[1*], Katrine Rye Hauerslev[2], Soren Redsted[3], Pernille Vahl[4] and Anne Kristine Larsen[1]

[1]Breast Surgery Department, Gentofte and Herlev Hospital, Copenhagen, Denmark.
[2]Department of Plastic and Breast Surgery, Aarhus University Hospital, Denmark.
[3]Department of Radiology, Aarhus University Hospital, Denmark.
[4]Department of Pathology, Aarhus University Hospital, Denmark.

Authors' contributions

This work was carried out in collaboration among all authors. Author MM designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors KRH and PV managed the analyses of the study. Author PV managed the literature searches. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Schwannoma of the Plexus Brachialis is a benign nerve sheath tumor and is a rare entity.
We report a case of a Cystic schwannoma (CS) from the left brachial plexus in the axilla, our surgical management of the case and a literature review.

Keywords: Plexus brachialis; cystic schwannoma; sheath tumors.

1. INTRODUCTION

Schwannomas are benign nerve sheath tumors (Table 1). They are rare and about 25% of them are localized in the head and neck region, involving cranial nerves and the sympathetic chain with the possible sparing of the brachial plexus.

A pure CS is relatively rare and localization in the brachial plexus is uncommon [1,2].

**Corresponding author: E-mail: mele_marco@hotmail.com;*

Sonnez et al. report that 8% of intracranial tumors are schwannomas and they represent about 30% of primary spinal tumors [3].

These tumors are also known as neurofibroma, neurinoma, neurilemoma or schwann cell tumor [2].

In the peripheral nerves they can show as sporadic tumors or associated with neurofibromatosis type 1 (NF1) and neurofibromatosis type 2 (NF2) [4].

In literature several cases of cervical or sacral nerve plexi schwannomas have been described but there is only a few cases upon brachial nerve plexus.
This type of tumor is mostly solid and malignant transformation is extremely rare [5,6].

They are composed of Schwann cells and are neuroectodermal in origin.

The patients affected by Schwannomas can refer to pain, loss of function, numbness or a progressively growing mass in the supraclavicular region. Sometimes the tumor is asymptomatic and can be misdiagnosed.

We report a rare case of a cystic schwannoma from the left brachial plexus in the axilla and our management. This localization is rare and in most cases asymptomatic.

2. CASE REPORT

A 77-year-old woman was referred to our Breast Surgical Department, Aarhus University Hospital, Denmark, by her general practitioner with the clinical diagnosis of left axillary nodule.

The only symptom reported by the patient was a lump in the left axilla which she felt a few days before seeing her general practitioner.

She had a history of medically treated subclinical thyrotoxicosis, psoriasis arthritis, hypercholesterolemia, and a former trauma to the right shoulder region.
The clinical examination showed normal findings in the breasts. In the top of the left axilla there was a palpable lump.

The visit was proceeded by a clinical mammogram including ultrasound of both breasts and axillary region.

The mammogram was normal. Ultrasound examination suggested a diagnosis of an oval, cystic, well-defined, and mobile lesion-measuring 25 mm in the largest diameter without doppler flow, in immediate proximity to the axillary artery. It was not possible to determine the nature of the tumor (Fig. 1).

Table 1. WHO classification of soft tissue tumors group 10 – peripheral nerve sheath tumors

Benign
Schwannoma (including variants)
Melanotic schwannoma
Neurofibroma (including variants)
Perineurioma
Malignant intermediate perineurioma
Granular cell tumor
Dermal nerve sheath myxoma
Solitary circumscribed neuroma
Ectopic meningioma
Nasal glial heterotopia
Benign triton tumor
Hybrid nerve sheath tumors
Malignant
Malignant peripheral nerve sheath tumor
Epithelioid malignant peripheral nerve sheath tumor
Malignant triton tumor
Malignant granular cell tumor
Ectomesenchymom

The presurgical diagnosis was cystic tumor in immediate proximity to the axillary artery.

We decided to perform an additional MRI of the left arm and left axilla.

The MRI examination was performed with intravenous contrast, “Dotarem®”, with T1 and T2 sequencies (Dixon method), and with axial and coronal reconstruction. A cystic structure, 27x27x23 mm, in close proximity with the axillary artery, was revealed by the MRI. The wall of the cyst was inhomogeneous in thickness, up to 5 mm, with contrast enhancement and it was recommended to abstain from fine needle biopsy. The MRI-diagnosis was a cystic structure in the left axilla suspected to be malign (Figs. 2-3).

The MRI and the findings were discussed in our Multidisciplinary Conference with the participation of our Sarcoma-specialist group, suggesting excision of the cystic tumor.

The patient underwent an excision of the mass in general anesthesia.

During the operation the macroscopically pathological findings revealed a cystic nodule located caudally to the axillary vein at the axillary level 2, firmly adherent to a nerve. The process was carefully dissected and released from the adherent nerve without any macroscopically signs of lesion. The brachial plexus was anatomically identified and distant from the cystic tumor.

In the postoperative period the patient complained about a loss of sensation and function of the left hand, wrist, and the first and second finger as well. An UL of the axilla was performed and excluded that the symptoms was caused by a compressing hematoma or seroma in the cavity.

On the 4th postoperative day, the patient was examined by a neurological specialist. There was no improvement of the first and second finger, but a minor improvement of the motor and sensory functions, and full recovery of the wrist.

Therapy from physiotherapist and occupational therapist was initiated and during the following months the patient showed continuous improvement of movement but no improvement regarding the sensation of the first and second finger. Follow up by a neurophysiologist including EMG, revealed partial affection of the left radial nerve with loss of continuation in the sensoric fibers but preserved motor fibers.

Six months postoperatively the patient was diagnosed with a mild residual neurological deficit.

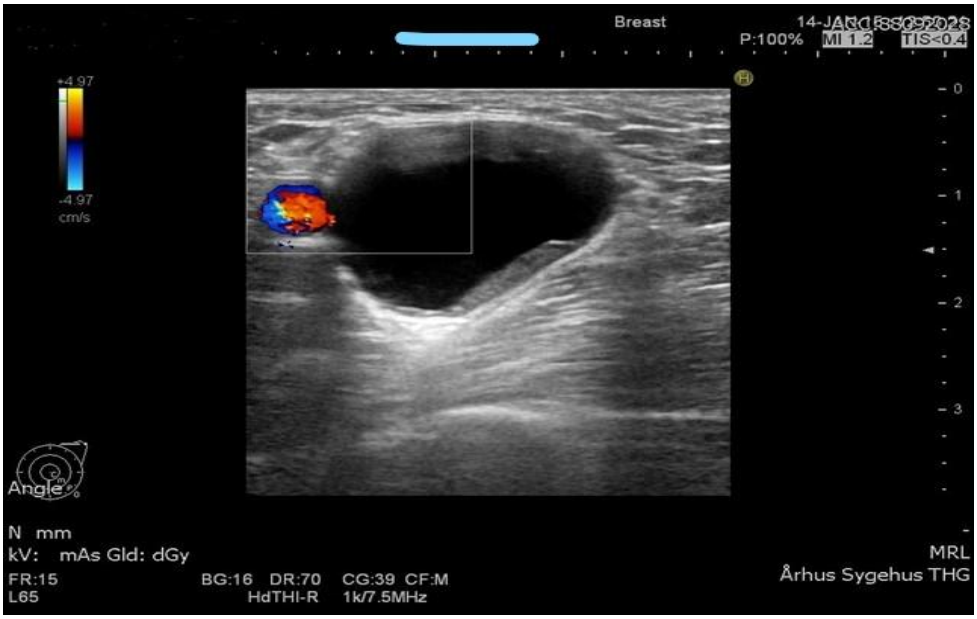

Fig. 1. Ultrasound of the left axilla: Oval, cystic lesion, mobile, well defined, measuring 25 mm

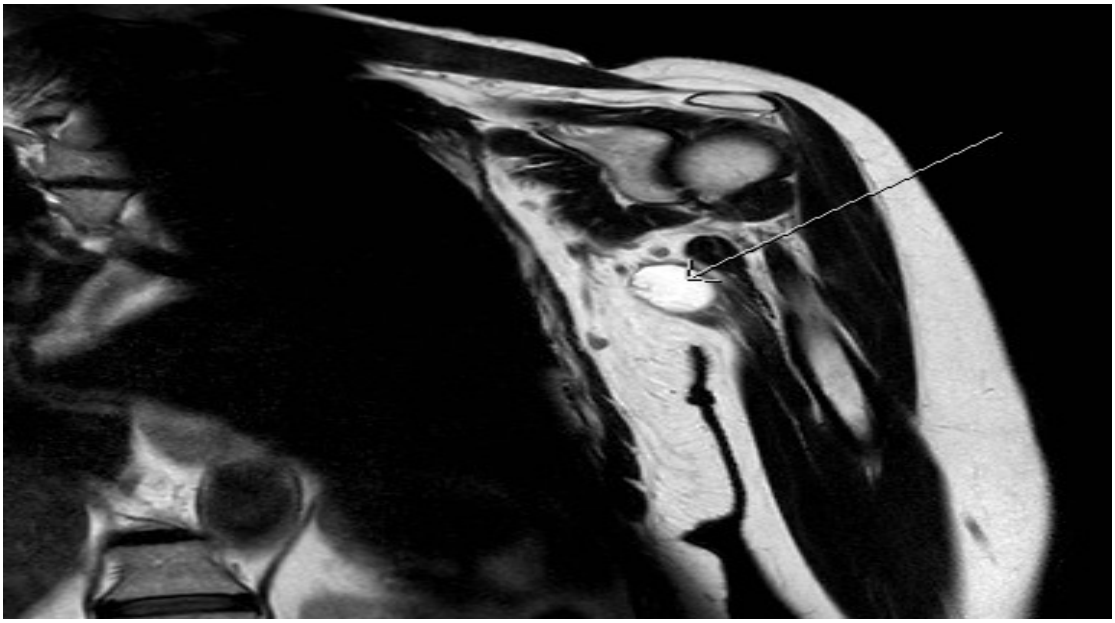

Fig. 2. MRI with Contrast enhanced T1 weighted (Dixon method) axial (A) and coronal (B), with a cystic lesion with uneven wall thickness with contrast enhancement, close to the axillary artery. There is no definitive relation to brachial plexus

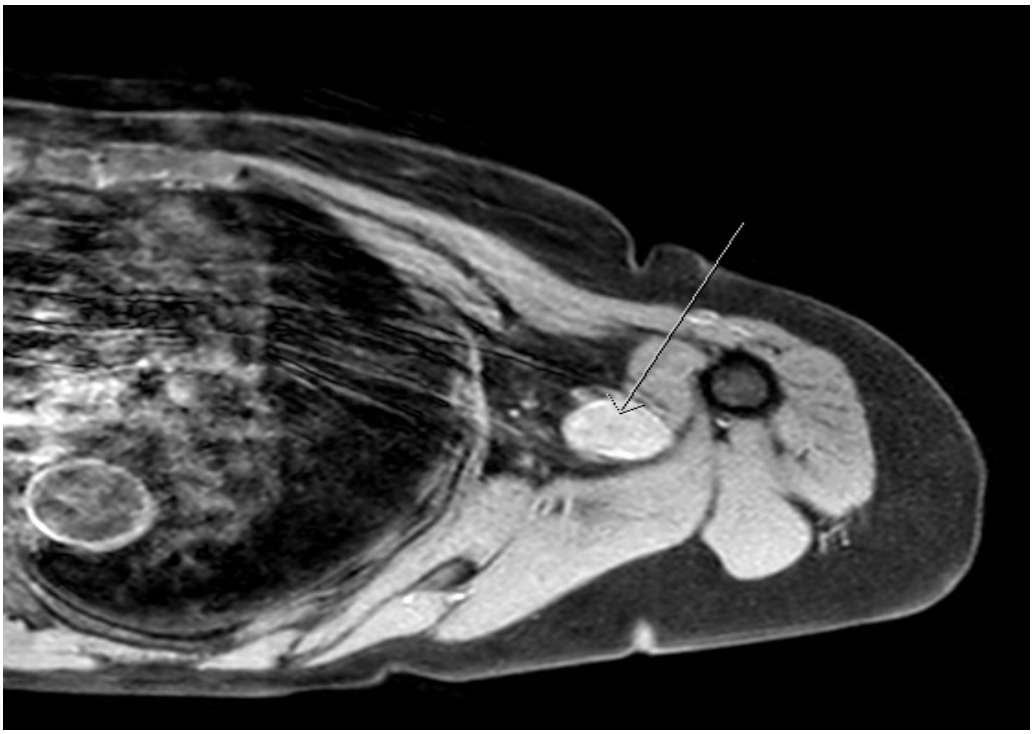

Fig. 3. Contrast enhanced T1 weighted (Dixon method) axial (A) and coronal (B), with a cystic lesion with uneven wall thickness with contrast enhancement, close to the axillary artery. There is no definitive relation to brachial plexus

2.1 Histopathologic Features

2.1.1 Gross pathology

A light-colored tissue with cystic appearance measured 30 x 16 x 5 mm. The cyst had a partially septal inner surface and mucoid contents. No lymph nodes were detected. The whole specimen was sectioned and embedded in paraffin.

2.1.2 Microscopic pathology

The microscopic appearance of the lesion was partially cystic and partially surrounded by a fibrous capsule, which formed septal extensions into the cavity of the cyst. In a septal formation of fibrous connective tissue, a larger peripheral nerve was seen (Fig. 4). The lesion itself, growing close to the nerve, consisted of spindle cells in bundles with focal areas of palisading nuclei. In addition to these features, some areas showed spindle cells with degenerated, enlarged nuclei (Fig. 5). A mild chronic inflammatory perivascular infiltrate was seen together with hemosiderin deposits, and finally slit-like hyalinized vascular structures were observed. Neither ectopic glandular mammary tissue nor lymph nodes were seen in the sections.

Immunohistochemical stains showed the tumor cells with a homogenous positive reaction for S-100 protein (Fig. 6), while the reaction for the vascular marker CD34 was negative in the tumor cells. There was a Schwannoma cells proliferation rate (ki-67) of 1-10%, largely from lymphocytes. This supports the benign diagnosis of ancient schwannoma. The lesion was removed marginally, including the capsule, pointing at an almost non-existing risk of recurrence.

Differential diagnosis of a schwannoma is fibroadenoma, phylloides tumor, fibromatosis and metaplastic carcinoma, all of which lesions usually can be distinguished in H&E histologic sections. Supportive of the diagnosis of a schwannoma is a positive immunohistochemical stain for S-100 protein, a negative reaction for actin and absence of mitoses [7].

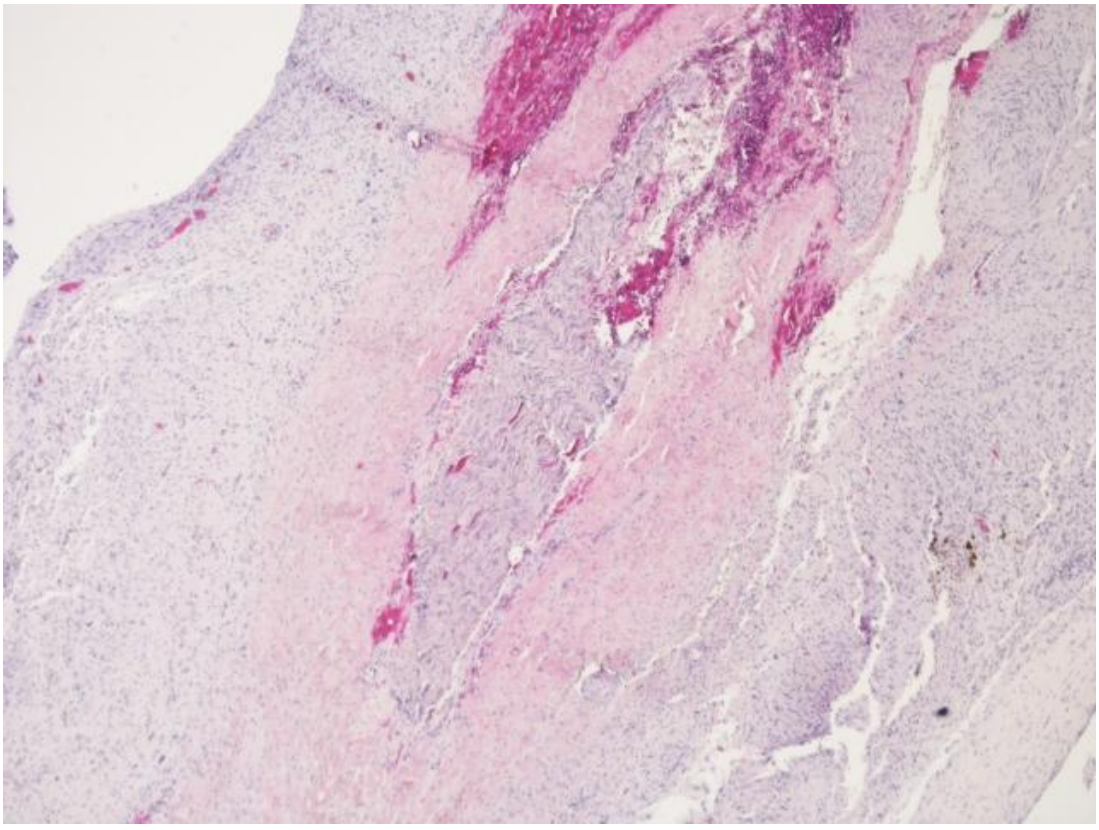

Fig. 4. The schwannoma is growing very close to a peripheral nerve

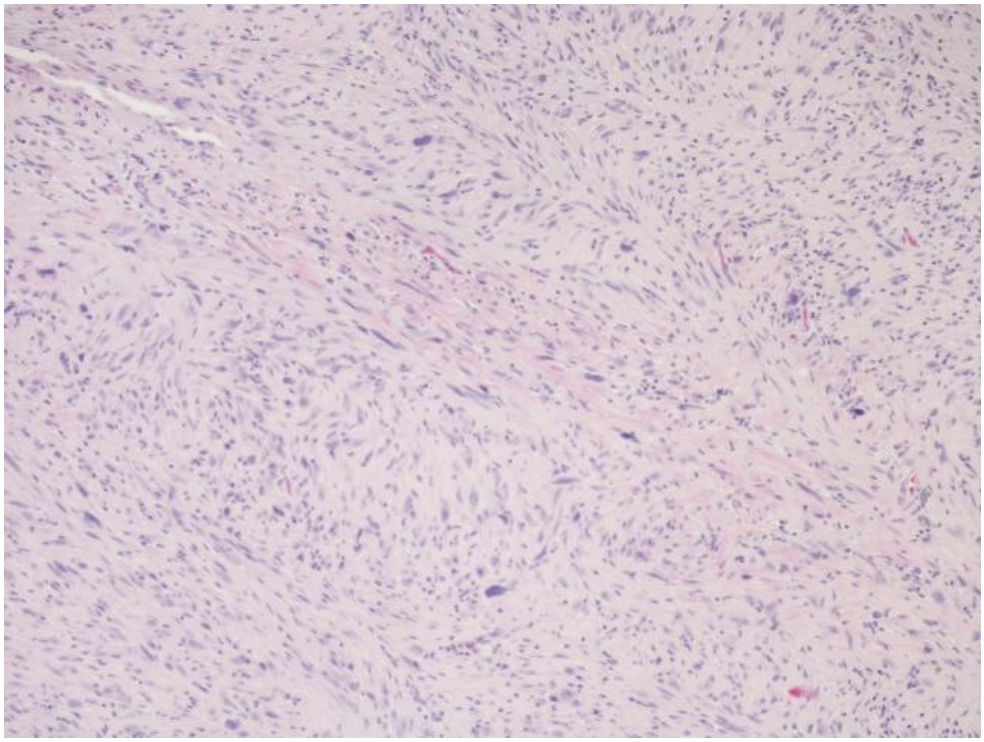

Fig. 5. In this close-up photo, degenerated enlarged nuclei are seen in some tumor cells warranting the diagnosis "ancient" schwannoma

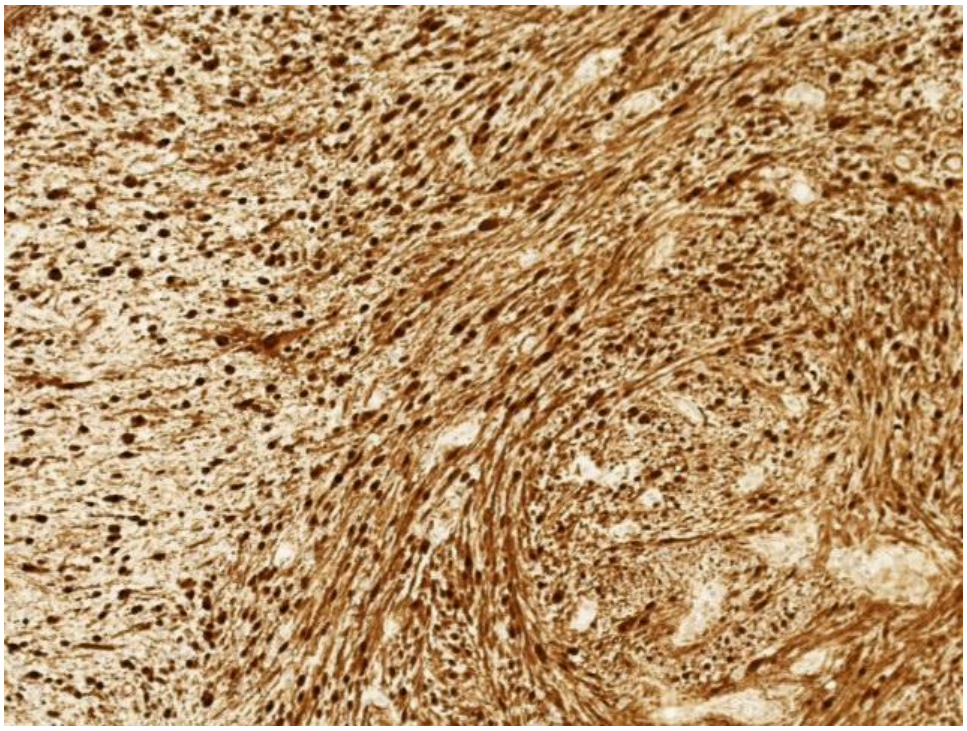

Fig. 6. Immunohistochemical stain shows a homogenous positive reaction for S-100 protein

3. DISCUSSION

Brachial plexus tumors are rare and uncommon. 25% are located on the head and neck [8]. Only 5% of the tumors are located in the upper limb, in the axillary region [9].

Schwannomas (neurilemmomas) are the most frequent and largest category of benign nerve tumors originating from schwann cells in the fascicles of the cranial autonomic or peripheral nerves [2-4,10,11]. Schwannomas are well encapsulated tumors and present a local slow growing mass. They are round, oval or plexiform, mostly solid or heterogeneous, ranging from 1.5 to 3 cm and they usually appear yellow or grey.

Some authors report schwannoma of the brachial plexus as an enlarged cystic mass [10] and it is possible to detect degenerative changes in the schwannoma (hemorrhage, calcification, and fibrosis) whereas cystic changes are very uncommon [11].

In literature they show a low tendency of transformation to malignancy [12].

They usually have a peak of incidence in the second to fourth decade of life. But may occur at any age [9,13,14].

Schwannomas of the plexus brachialis are usually asymptomatic or occurring insidiously, resulting in incorrect or delayed diagnosis. However, the most common symptoms are palpable mass and pain related to local growth of the primary brachial plexus tumor with compression of the vascular and neural structures. Numbness or paresthesia, Tinel's sign, and weakness of the arm are symptoms more frequently related to neurofibromas and malignant nerve sheath tumors [15].

Sönmez et al. report a case with schwannoma of the right brachial plexus causing thoracic outlet syndrome with numbness, pain, and progressive, lingering paralysis in the arm.

This symptomatology is very rare and described in only a few studies. (McAllister et al., 1989; Hornick et al., 1991; Atasoy, 1997; Nakazawa et al. 2005) [3,9,12,16].

Therefore, in the presence of a palpable asymptomatic formation in the axillary cavity it is absolutely necessary not to exclude for a correct differential diagnosis as the schwannoma of the secondary trunks of the brachial plexus, is also rarely accompanied by neurological signs and symptoms (Tinel sign, local pain, radiated pain, paraesthesia), as mentioned above.

The differential diagnosis of these tumors must include cervical lymphadenopathy, lipoma, paraganglioma, angioma, cleft branchial cysts, and vascular tumors [2].

Physical examination show a painless, mobile swelling with symptoms of pain and percussion that produce paresthesia along the involved nerve [9].

Fine needle aspiration has been recommended as an initial testing procedure although in some cases it can be misleading, as it presents inadequacies in diagnosing these tumors and misdiagnosis can lead to unnecessary interventions with serious post-operative consequences [2,17].

Therefore, to avoid such occurrences and to optimize diagnostic confirmation, it becomes essential to perform MRI with which it is possible to highlight a well encapsulated solitary lesion that causes the displacement of the fascicles [9].

Even CT, as for MRI, is considered necessary for preoperative diagnosis: The first highlights cystic degeneration, while the second visualizes the tumor, with its capsule and also the course of the nerve.
In some cases of cystic schwannomas presented in the literature, both ultrasound and CT were falsely suggesting the diagnosis to be complex cyst, lymphadenopathy or hydatid cyst [8].

Obviously, the sensitivity of ultrasound is in many cases able to define the location, morphology and nature (traumatic, neoplastic, inflammatory) of the brachial plexus lesions and therefore remains the main method for the approach in the preoperative evaluation of the pathological processes of the brachial plexus [18].

To exclude any neoplastic lesions, a total body spiral CT and MRI must be performed. The CT is considered in association to neurofibromatosis, while the MRI is considered the “technique of choice” in the diagnosis of peripheral nerve schwannomas.

Given the rarity and diagnostic difficulties of peripheral nerve tumors located in the brachial

plexus at the level of the axilla, a correct preoperative diagnostic procedure has not yet been defined in the literature.

Surgery is especially indicated in cases where the growth of cystic schwannoma causes neurological deficit, discomfort, progressively growing lesions with suspicion of malignancy and to prevent or minimize neural damage.

Both the intracapsular enucleation technique and the extracapsular excision technique are described in the literature, with or without the use of a dissecting microscope. The neural fascicles surrounding cystic Schwannoma are usually separable and tumor enucleation is almost always possible.

The surgical technique with the use of the operating microscope is the treatment of choice used and allows, in most cases, the radical removal of the neoplasm, but presents risk of recurrence if the resection is incomplete. However, functional recovery is faster with the microsurgical technique.

As highlighted in the literature, most patients have transient neurological symptoms, mainly due to nerve manipulation during the operation.

Of course, the complete removal of the tumor with the preservation of the surrounding nerves is the main goal we attempt to achieve.

Malignant transformation of cystic schwannoma of the brachial plexus is very rare, although it has been described by some authors [19,20,21, 22,23].

4. CONCLUSION

Cystic Schwannoma of the region of the brachial plexus, as we reported in our case, is a rare occurrence and only rarely affects the secondary trunks of the brachial plexus.

It can present as a neoformation of the axillary cavity, sometimes totally asymptomatic and is difficult to distinguish from the more frequent axillary lymphadenopathies.

A correct diagnosis of the lesion preoperatively is very important as excision may cause transient and/or permanent neurological damage [9].

Schwannomas of the brachial plexus should be included in the differential diagnosis of lateral neck masses, especially in the presence of neurological symptoms.

In our opinion, CT or MRI should be performed for careful preoperative evaluation, preceded by an initial ultrasound study [2].

As described by other authors, needle aspiration can be misleading.

During the operation, full attention is required to preserve nerve function and therefore use of the microscope is highly recommended.

CONSENT AND ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s). We hereby state that written consent has been obtained from the patient and will be provided upon request.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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