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CASE REPORT

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Schwannoma of the brachial plexus with cystic degeneration: A case report schwannoma of the brachial plexus

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Abstract

Schwannoma of the brachial plexus can be present as a painless swelling without an upper limb functional or sensitivity deficiency. Thorough examinations, including MRI, are necessary to identify the nerves of origin and prevent potential harm.

1 | INTRODUCTION

The case of a supraclavicular cystic mass arising from the brachial plexus and histologically confirmed to be a schwannoma is presented. Due to its low accuracy, fine needle aspiration cytology (FNAC) should not be used as an ultimate diagnostic tool. Complete surgical removal while maintaining the nerve trunk was performed.

Schwannomas are benign encapsulated tumors that originate from the Schwann cells of the cranial, peripheral, or autonomic nerve sheaths. These can affect the third to the twelfth cranial nerves and the peripheral and autonomic nerves.[1-3] Discovered by and, thus, named after the German physiologist Theodor Schwann, Schwann cells, or neurolemmocytes are the most important glial cells of the peripheral nervous system, as they provide insulation and promote the saltatory conduction of action potentials along axons.[2,4] Nearly 25%-45% of schwannomas occur in the head and neck region; however, those arising from the brachial plexus are rare, accounting for only 5% of all schwannoma cases.[1-5] Schwannomas localized to the brachial plexus pose diagnostic challenges, as they are often confused with cervical lymphadenopathy.[2,6]

In this study, a lateral supraclavicular cystic mass originating in the brachial plexus histologically confirmed to be a schwannoma is reported in a 32-year-old man.

2 | CASE HISTORY/ EXAMINATION

A 32-year-old male patient experienced painless swelling in the left supraclavicular region over the past six years. The swelling progressively enlarged from the size of an almond to a lime. He had no history of trauma, fever, or systemic illness. The only complaint was the visible swelling; there was no weakness, numbness, or loss of function in the upper limb. Upon examination, a painless, round, firm, mobile in the medio-lateral direction swelling with a diameter of 4.0 cm was found in the left supraclavicular region (Figure 1).

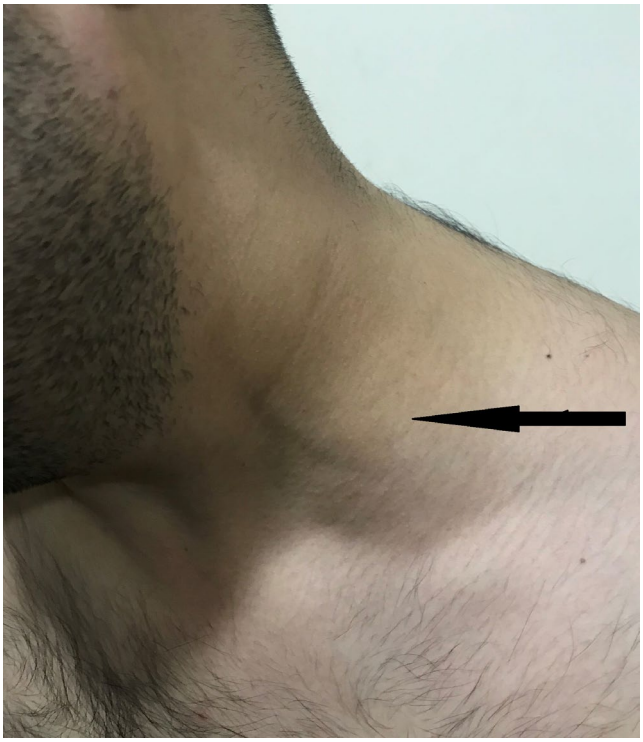

FIGURE 1 External view of patient's neck—the swallow on the left supraclavicular region

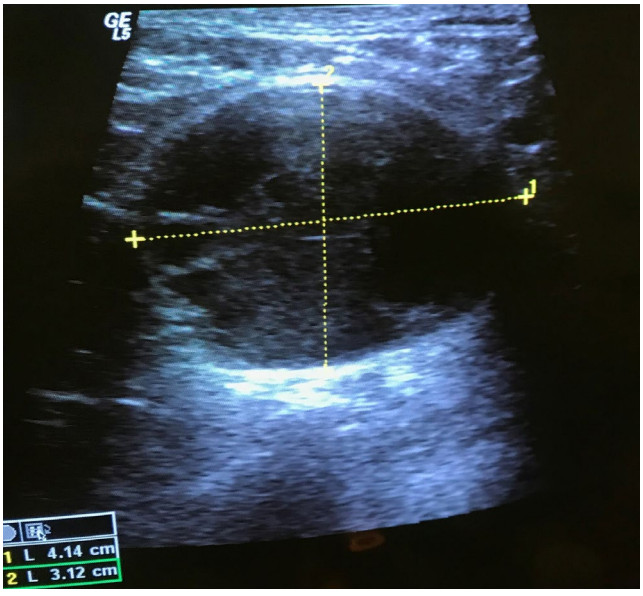

FIGURE 2 Ultrasound imaging of the cystic mass on the neck

3 | DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS, AND TREATMENT

Differential diagnosis of the presented supraclavicular mass included different benign and malignant conditions such as lymphoma, lymph node metastasis, tuberculosis, sarcoidosis, and cystic lymphangioma.

The ultrasound examination was performed as a first-line radiological investigation. A superficial heterogeneous round cystic mass measuring 47 × 33 mm was visible in the left supraclavicular region in ultrasound imaging; this mass was lateral to the sternocleidomastoid muscle (Figure 2). The margins of the mass were well-defined and smooth, and there were many intralesional septa. The lower border of the formation was adjacent to the subclavian artery, while the medial border lined the carotid artery and the external jugular vein. The ultrasound examination diagnosed a branchial cleft cyst with an atypical location.

Fine needle aspiration (FNA) was performed, and the cystogram corroborated the diagnosis. The patient was scheduled for surgery with the initial diagnosis of a lateral branchial cleft cyst of the neck with a rare atypical supraclavicular location.

During the surgery, general anesthesia was used. The anterior supraclavicular approach was followed, and an approximately 4-cm-long incision was made parallel to the external jugular vein to expose the lesion. Perioperatively, a well-encapsulated mass was found lying medially within the C6 and C7 roots and pushing the nerve fascicles toward the periphery (Figure 3). The intralesional fluid aspiration method reduced the size of the lesion and tension in the capsule. The tumor was carefully enucleated, avoiding any damage to the trunk and preserving the nerve fascicles. Macroscopically, the cystic mass was yellowish and well-encapsulated (Figure 4).

Multiple hematoxylin and eosin sections showed a lesion with multiple pseudocystic areas without an apparent

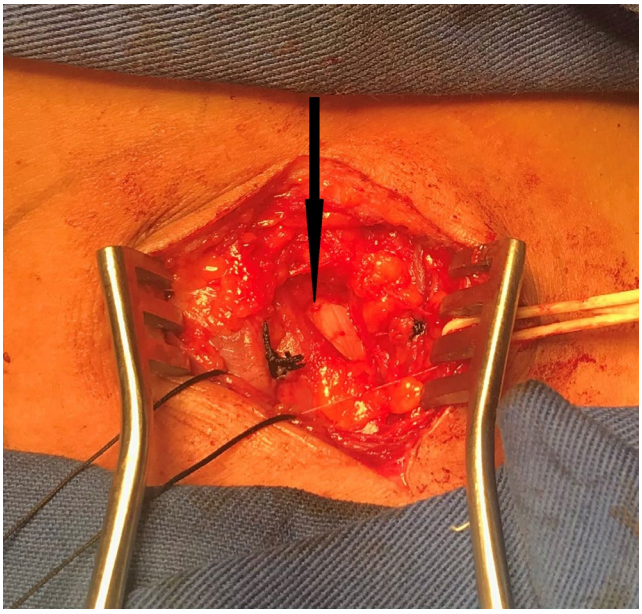

FIGURE 3 Intraoperational view of the lesion: encapsulated mass within the C6 and C7 roots

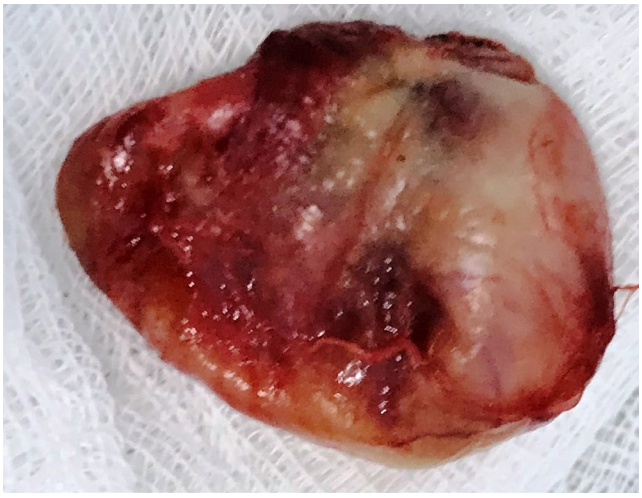

FIGURE 4 Macroscopic view of the enucleated tumor

epithelial lining; in addition, there were cellular regions with interlacing bundles, palisading the eosinophilic regions called Verocay bodies and other hypocellular areas of spindle cells (Figure 5). No nuclear atypia, mitotic activity, or foci of necrosis was observed. Immunohistochemical analysis showed diffused strong expression of the S-100 protein (Figures 6 and 7). "Cystic" spaces in the inner surfaces tested negative for CD34 (Figure 8) and AE1/3 (Figure 9).

The histopathology and immunohistochemical analysis of the tumor supported the diagnosis of schwannoma with cyst degeneration.

4 | OUTCOME AND FOLLOW-UP

The patient had an uneventful recovery postoperatively, with a complete range of active motion and intact sensations but severe discomfort in the shoulder and left arm for 10 days. He was closely followed up for 10 months through monthly visits and remained symptom-free during this period. In a subsequent visit 16 months after the surgery, he had assumed his normal routine with no evidence of recurrence.

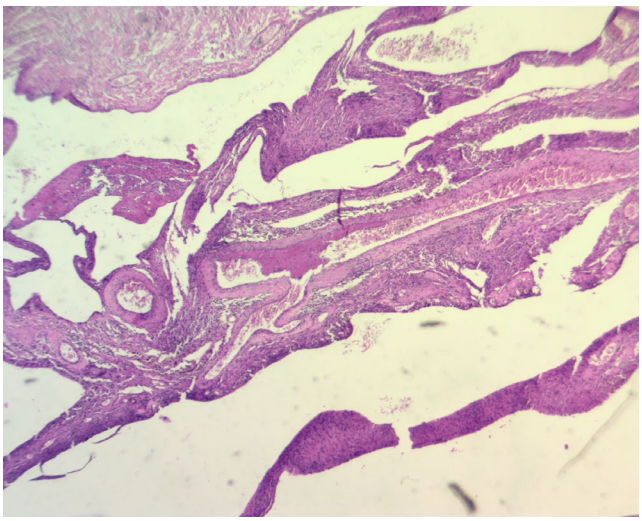

FIGURE 5 Pseudocystic areas without apparent lining, ×40, H@E

5 | DISCUSSION

Brachial plexus tumors are uncommon, accounting for only 5% of tumors in the upper limb.[5] Schwannomas are slow-growing, benign, encapsulated masses originating from Schwann cells.[2-6] They are usually painless, asymptomatic, and secluded masses. However, some patients may exhibit symptoms of nerve compression and, less commonly, radiation pain, and sensory or motor deficiency.[5,7,8] The clinical diagnosis of schwannomas from the brachial plexus of the upper limb depends on the evidence of the following features: firm, non-tender, mobile swelling with symptoms of pain upon palpation, and paresthesia along the nerve involved.[8-10] However, not every patient with brachial plexus schwannoma experiences such typical clinical features; therefore, differential diagnoses with other lesions are essential for these cases.[2,6,8,9] Due to their rarity and complex anatomical location, these cases are a diagnostic and therapeutic challenge for surgeons.[2,6,11] A more common differential diagnosis for a painless, lateral, cystic neck mass is a lymphangiomatous malformation, the histological subtypes of which include cavernous lymphangioma, lymphoma, lymph node metastasis, fibrosarcoma, leiomyosarcoma, sarcoidosis, tuberculosis, capillary lymphangioma, cystic hygroma, and brachial cleft cysts.[1,11,12] Brachial plexus schwannomas are rarely found and uncommonly present as a cystic neck mass.[13] In the present case, the patient had no upper limb weakness, pain, tingling sensations, or numbness in the left upper limb. Kumar and Akhtar[5] also presented a case of brachial plexus schwannoma in a patient who did not have any neurologic deficits in the upper limb. Considering the localization of the mass (lateral neck), there was no suspicion of a lateral cyst of the neck on the clinical examination tool. As noted above, the main differential diagnosis was carried out for lymphoma, lymph node metastasis, tuberculosis, sarcoidosis, cystic lymphangioma, and, therefore, ultrasound investigation was assigned for this patient as a first-line investigation.

For superficial lesions, ultrasound is the first-line imaging method of choice for diagnosis. It is non-invasive, rapid, and low-cost; moreover, there is no involvement of exposure to ionizing radiation, and the method clearly reveals the cystic nature of the mass.[14] However, in the present case, the ultrasound examination was suboptimal and led to a misdiagnosis. The heterogeneous round cystic mass with well-defined and smooth margins and intralesional septa was misdiagnosed as a third branchial cleft cyst with atypical location. Cystic

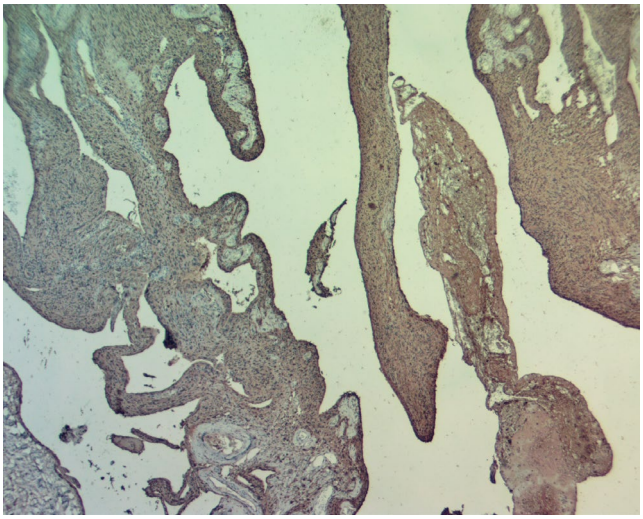

FIGURE 6 Immunohistochemical staining S100, ×40, diffuse strong expression of the S-100 protein

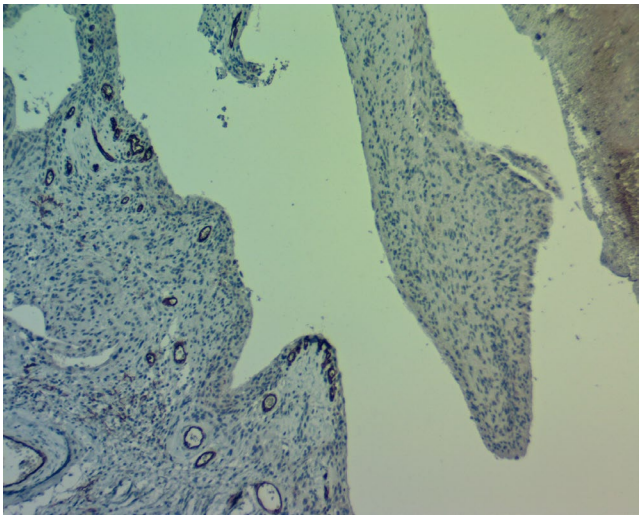

FIGURE 8 Immunohistochemical staining CD34 shows negative expression for lining (arrows), positive in vessels (arrowheads), x200

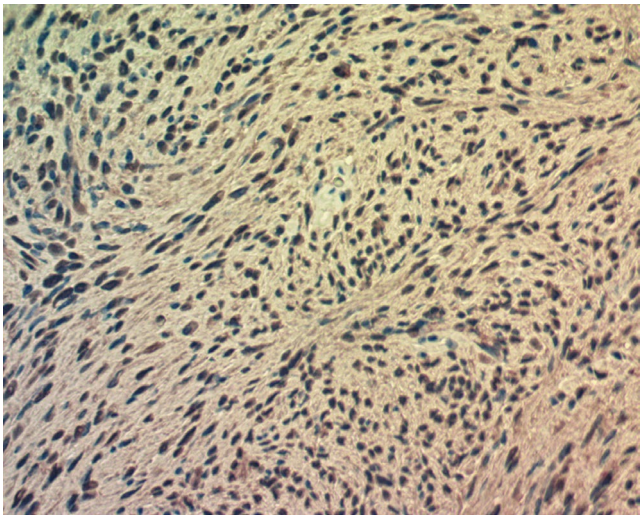

FIGURE 7 Immunohistochemical staining S100, ×200, diffuse strong expression of the S-100 protein

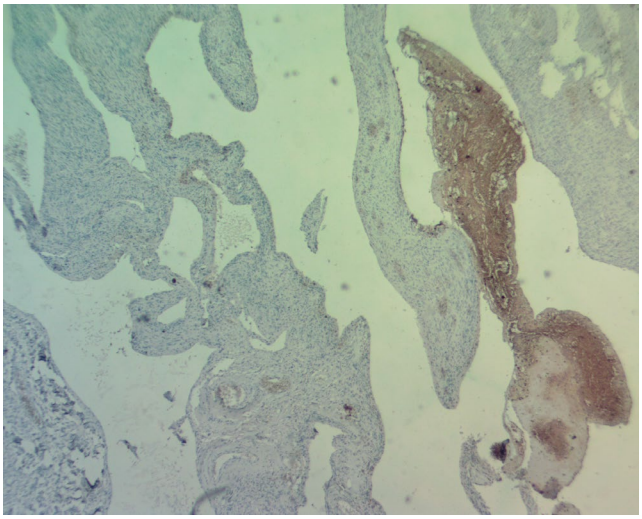

FIGURE 9 Immunohistochemical staining AE1/3-negative reaction for lining (arrows) ×100

masses of neck include a wide range of congenital and acquired lesions and various inflammatory and neoplastic diseases.[15] Third branchial cleft cysts are located in the posterior cervical space posterior to the common or internal carotid artery and the sternocleidomastoid muscle, but not as low as the supraclavicular region.[15] Metastatic nodes from head and neck malignancy, especially papillary carcinoma of the thyroid, are the most common types of nodal metastases, presenting as cystic masses in the neck; however, they are present in the central cystic area with thick irregular walls, or an eccentric solid component may be seen. Occasionally, necrosis within a metastatic lymph node may be florid, mimicking a congenital cyst.[15]

As noted by Yasumatsu et al,[16] in their review of 27 cases of schwannomas, ultrasonography was performed in all cases, but none of the cases were diagnosed with schwannoma. Ultrasound examination images of schwannoma typically show a well-defined, ovoid (or round), hypoechoic, and primarily homogeneous solid mass with or without moderate posterior acoustic enhancement, but rarely show a direct connection to the nerve.[16] Only one in seven cases (14%) was diagnosed as a schwannoma by CT scan, whereas 20 of 25 cases (80%) were diagnosed as schwannomas by MRI scan. Although both MRI or CT scans can be informative and helpful in confirming the diagnosis,[1-5,8,11,12,15,16] the relationship between the schwannoma and its nerve of origin can be better appreciated through an MRI. In addition, an MRI appears to be the investigation of choice for diagnosis and identification of nerve of origin.[2,15,16] An MRI consistently identifies these

lesions on both T1- and T2-weighted imaging. T1-weighted images display low signal intensity, and T2-weighted images show high intensity.[16]

Once the mass is confirmed to be nonvascular in origin, FNAC is recommended.[1,11,16] According to literature, cytomorphological features of schwannomas contain a mixture of two distinctive areas. Antoni A areas are cellular with nuclear palisading and Verocay bodies where two rows of palisading nuclei are separated by pink fibrillary material. Antoni B areas are less cellular and microcystic areas. Degenerative changes such as cyst formation, focal calcifications and hyalinized, thrombosed blood vessels with associated hemorrhage and deposition of fibrin are typically present.[2,11,16] However, there are several challenges in the diagnosis of neural lesions by cytology.[17] Yasumatsu et al[15] noted that only three of 12 cases (25%) were specifically diagnosed as schwannomas by FNAC. In the presented case, FNAC revealed histiocytes and dendritic cells, while the cytogram revealed a cyst.

Surgical excision is recommended as the first choice of treatment for tumors of this type, that is, which cause neurological deficits and discomfort.[1-12,18,19] Since schwannomas are well encapsulated, it is almost always possible to enucleate and separate the tumor from the neural membrane, which is the main goal of the operation.[1] Courvoisier reported the first surgical removal of a schwannoma of the brachial plexus in 1886; however, paralysis of the deltoid and bicep muscles occurred after surgery.[1] The complications reported in most case series are temporary sensory and motor deficits.[2,5,12,18,19] In the present case, we were also able to dissect the nerve fascicles and successfully enucleate the tumor while preserving the nerve fascicles. Thus, we were able to preserve nerve function and, therefore, the patient did not postoperatively exhibit sensory or motor deficits.

6 | CONCLUSION

Schwannoma of the brachial plexus is a rare tumor and can be present as a cystic mass of the neck. A proper diagnosis of the lesion should be established before surgery, as it can be misdiagnosed as a branchial cleft cyst of the neck, as in the present case. MRI diagnostics should be performed if schwannoma is suspected. Moreover, due to its low accuracy, FNAC should not be used as an ultimate diagnostic tool. Further, complete surgical removal while maintaining the nerve trunk is recommended.

ACKNOWLEDGMENTS

Published with written consent of the patient.

CONFLICT OF INTEREST

The authors have no conflicts of interest to declare.

AUTHOR CONTRIBUTIONS

LVA: was accountable for all aspects of the work in ensuring that the questions related to the accuracy or integrity of any part of the work were appropriately investigated and resolved and that the clinical work was implemented. TSA: was responsible for performing an accurate literature review and implementing the clinical work. PSK: made contributions to the design of the histological examinations and was responsible for the histological data, design, and literature review. AYP: drafted the manuscript for important intellectual content and made substantial contributions to the concept and design.

ETHICAL APPROVAL

We confirm that explicit written consent to publish the results has been received from the described patient.

INFORMED CONSENT

Written informed consent was obtained from the patients for the publication of this case report and any accompanying images. A copy of the written consent is available for review to the editor-in-chief of this journal.

DATA AVAILABILITY STATEMENT

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

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