Case Report

A rare presentation of supraclavicular schwannoma

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ABSTRACT

Schwannomas or neurilemmomas are benign tumors that arise from Schwann cells that surround the peripheral, cranial, or autonomic nerve sheaths. These are slow-growing, benign, encapsulated tumors that arise from the proliferating Schwann cells, encompassing the nerve sheath. About 25–45% of the schwannomas are found in the head-and-neck region. These tumors can arise from any of the cranial, peripheral, or autonomic nerves and constitute <1% of the tumors seen in the head-and-neck region. In the present case, the tumor was in the lateral group, localized to the right-sided supraclavicular region, and originating from the cervical or brachial plexus. We present the case of a 44-year-old gentleman who presented with a right-sided supraclavicular swelling for 2 months, which initially was thought to be a lymph node mass but which turned out to be a schwannoma on the pathological and radiological diagnosis.

Key words: Schwannoma, Subclavian vessels, Supraclavicular

Schwannomas or neurilemmomas are benign tumors that arise from Schwann cells that surround the peripheral, cranial, or autonomic nerve sheaths. These are slow-growing, benign, encapsulated tumors that arise when proliferating Schwann cells form a tumor encompassing the nerve sheath. About 25–45% of the schwannomas are found in the head-and-neck region. These tumors can arise from any of the cranial, peripheral, or autonomic nerves. These constitute <1% of the tumors seen in the head-and-neck region [1].

We present the case of a 44-year-old gentleman who presented with a right-sided supraclavicular swelling for 2 months, which initially was thought to be a lymph nodal mass but turned out to be a schwannoma on the pathological and radiological diagnosis. In the present case, the tumor was in the lateral group, localized to the right-sided supraclavicular region, and originating from the cervical or brachial plexus. As schwannomas pose a diagnostic dilemma, particularly when they are in the supraclavicular region, a thorough understanding of its anatomy, clinical, and systematic evaluation is necessary.

CASE REPORT

A 44-year-old gentleman presented to the hospital with a swelling in the right side of the neck in the supraclavicular region for 2 months. The swelling had grown in the course of these 2 months. There was no history of trauma, fever, and systemic illness. The patient had no complaints of pain, weakness, dysphagia, and hoarseness of voice.

In physical examination, his vital parameters were stable with a heart rate of 88/min and blood pressure of 130/90 mmHg. The systemic examination was within normal limits. On local examination, a firm, mobile, and painless mass of about 3 cm × 2 cm was identified in the right supraclavicular region. There was no loss of power in muscles, no signs of wasting, or neurological deficits. An initial clinical diagnosis of supraclavicular neck swelling, probably a lymph node or a lipoma, was made.

Ultrasound (USG) scan of the neck revealed 2.4 cm × 1.6 cm solid lesion in the right supraclavicular region (Fig. 1). It showed moderate vascularity on color Doppler and was in close relation to the subclavian vessels. The lesion was reported as supraclavicular lymph node/swelling. The patient underwent fine-needle aspiration cytology (FNAC) of swelling which revealed hemorrhagic aspirate and no further opinion was made; hence, excision biopsy was suggested. After relevant investigations, the patient was optimized for surgery.

The patient underwent surgery under general anesthesia. The neck was turned toward the left side and a curvilinear skin crease incision was made. The incision was deepened and the fascia was opened which revealed a 3 cm × 2 cm firm, oval swelling deeper to the subclavian veins. The veins were gently retracted and the swelling dissected all around. It was arising from a yellow structure probably a nerve, peripheral nerve. The swelling was sent for excision biopsy.

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On gross examination, the swelling was 2.5 cm × 2.0 cm × 1.2 cm single, nodular, gray-brown tissue weighing 10 g with glistening and hemorrhagic areas on the cut section (Fig. 2). Histopathological examination revealed hypercellular (Antoni A) and hypocellular (Antoni B) pattern of spindle cells with elongated nuclei, Verocay bodies with peripheral lymphoid aggregates, and nerve fibers, suggestive of schwannoma (Fig. 3). The post-operative stay was uneventful. He was discharged on the same day of surgery on a daycare basis. He is on regular follow-up.

DISCUSSION

Schwannomas are benign tumors that arise from Schwann cells in peripheral, cranial, and autonomic nerve sheaths [1]. These are rare, slow-growing, encapsulated, benign tumors with regular margins that originate from the Schwann cells of the nerve sheath. Schwannomas do not arise from the optic and olfactory nerve as they lack Schwann cells. The probability of malignant transformation of schwannomas is around 8–10% [2]. They are the most common tumors of peripheral nerves, but <5% of all schwannomas arise from the brachial or cervical plexus [3].

Although the etiology is unknown, few causative factors such as spontaneous growth, external injury, chronic irritation, or exposure to radiation have been hypothesized. Typically, these are solitary lesions without genetic or gender predisposition, although they can occur rarely in multiples when associated with neurofibromatosis. Head and neck are the most commonly affected regions (25–45%) with the lateral neck being the frequently involved site [4].

Primary tumors of the brachial plexus are rare and are divided into two groups, peripheral neural sheath tumors and peripheral non-neural sheath tumors [5]. Schwannomas, together with neurofibromas, are peripheral nerve sheath tumors and rarely affect brachial plexus. Schwannomas in the neck region most commonly arise from the vagus nerve. Schwannomas are always a diagnostic dilemma as they are asymptomatic for a long time and histopathology is the gold standard for diagnosis. Head-and-neck schwannoma although very rare should be considered as a differential diagnosis when we come across a patient with a unilateral slow-growing mass in the head-and-neck region, particularly in an adult.

In a study, Kehoe *et al*. reported that 36% of the tumors originating from the brachial plexus are localized to the supraclavicular and 64% to the infraclavicular region [6]. Langner *et al*. reported that among 21 schwannoma cases, 5 (23.8%) originated from the brachial plexus, 3 (14.2%) from the 10th cranial nerve, 4 (19%) from the sympathetic plexus, and 3 (14.2%) from other nerves. The authors reported that in six cases (28.4%) of the same series, the nerve from which the tumor originated could not be found [7]. In an unusual presentation, Patel *et al*. described a case of a patient with adenocarcinoma of the descending colon with a right supraclavicular swelling which was hard in consistency, immobile, non-tender, non-fluctuant which revealed to be metastasis to the right clavicle resulting in a pathological fracture [8].

Carotid body tumors, lymph nodal enlargement, thyroid nodules or thyroid cysts, branchial cysts, teratoma, dermoid cysts, lipomas, metastatic masses, and neurofibromas should be considered in the differential diagnosis of schwannomas in the supraclavicular region [9].

As a rule, the treatment is surgical excision and dictated by the location of the tumor and nerve of origin. Complete

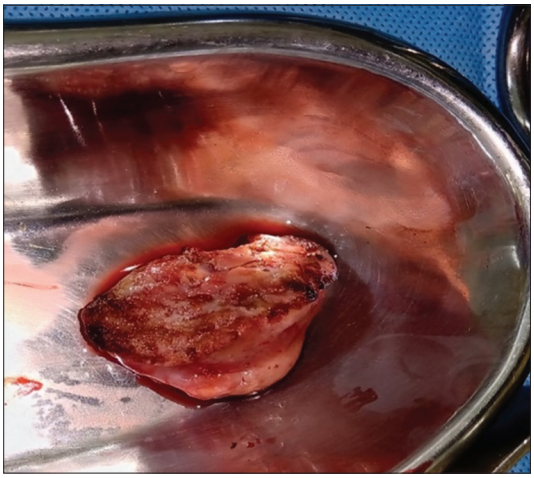

Figure 2: Gross specimen of schwannoma

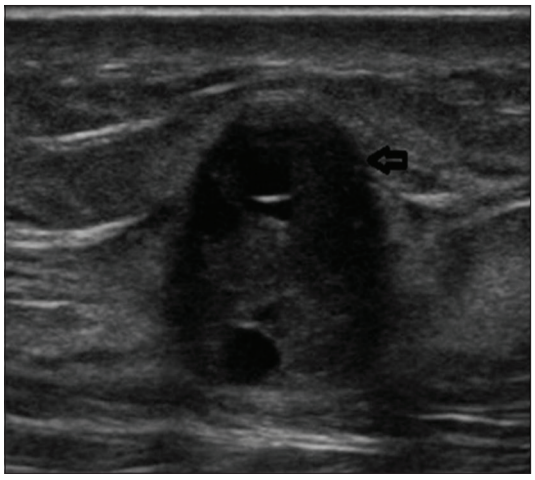

Figure 1: Ultrasonogram of schwannoma

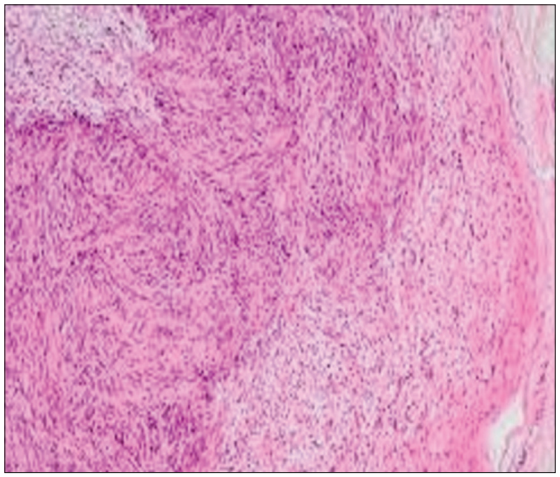

Figure 3: Histopathological examination showing Antoni A and Antoni B pattern

extracapsular excision preserving the nerve of origin should be attempted when feasible but for extensive schwannomas, nerve sacrifice with reconstruction and rehabilitation are important considerations [10]. Due to its rare presentation, complex anatomical location, and morbidity risk post-excision, they always pose a formidable challenge to surgeons and hence must be dealt with utmost patience and meticulous dissection.

CONCLUSION

Supraclavicular schwannoma is a rare entity, often due to a diagnostic dilemma, and is mistakenly diagnosed as a lymph nodal swelling or lipoma of the neck region. In our case, USG of the neck showed the swelling to have moderate vascularity suggestive of a vascular extension of the swelling. FNAC revealed a hemorrhagic aspirate, pointing toward a vascular swelling, complicating the diagnosis. But during the excision of swelling, it was found that swelling was beneath the subclavian vessels, a rare presentation and on histopathological investigations turned out to be a schwannoma.

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