

Schwannoma in the Lateral Neck, A Rare Case and Review of the Literature

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ABSTRACT

Background: Schwannomas are rare tumours which arise from the sheaths of Schwann cells either from the peripheral, autonomic or central nervous system. It is rare and very few have been reported locally. We report a case of schwannoma, in a 40-year-old woman who presented with a ten-month history of right lateral neck mass which was slow-growing and painless. There were no associated obstructive or toxic symptoms. Examination revealed a well-demarcated right-sided lateral neck mass in zone II of the neck measuring 13 X 6cm, firm, non-tender, not attached to the overlying skin and mobile in the horizontal plane. She had an examination under anaesthesia and total excision of lateral neck mass. Histopathological examination of the mass revealed features in keeping with schwannoma. The patient did well postoperatively.

Keywords: schwannoma, lateral neck mass, histopathology

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Introduction

Schwannomas are tumours that originate from the sheath of Schwann cells and could arise from either the peripheral, autonomic or central nervous system.^{1,2,3} The term schwannoma was coined in 1908 by Verocay.^{3,4} It is also known as neurilemmoma, peripheral fibroblast or nerve sheath tumour.^{5,6} They account for 25-45% of head and neck tumours.^{2,4,7}

They are commonly seen in the 4th- 6th decades of life and have no sex predilection.^{3,4,5} The commonest site in the head and neck region is the parapharyngeal space and the commonest nerve of origin is the vagus nerve.^{1,3,5} They present as a painless slow-growing well-circumscribed mass without neurological features.^{1,2} It usually has an enlargement rate of 2.5-3 mm per year.⁸ Malignant transformation is rare and is considered when there is pain or neurological deficits.^{3,4,7} Patients with cervical schwannoma may present with hoarseness due to vocal cord palsy, whereas the pathognomonic sign for vagal schwannoma is a paroxysmal cough during palpation of the mass.^{9,10} Radiologic imaging plays an important role in diagnosing vagal nerve schwannoma. Commonly performed radiologic investigations include ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI). However, MRI is considered the gold standard to assess vagal nerve schwannomas and to evaluate their extent.^{1,11} There are very few reported cases of cervical schwannoma worldwide and even fewer if any reported from this part of the world. We herein report a very rare case of cervical schwannoma presenting as a lateral neck mass.

Case Report

RM is a 42-year-old female who presented to the Ear, Nose and Throat, Head and Neck Surgery out-patient clinic of UMTH Maiduguri on 18/04/2023 with a ten-month history of painless, progressive right lateral



neck mass which was small initially and slowly increased to the size of her clenched fist at presentation. There were no compressive symptoms and no other ear, nose and throat symptoms. There was no history of cough, weight loss, fever or night sweats, and no history of contact with a patient with tuberculosis. Examination revealed a well-demarcated right-sided lateral neck mass along the medial border of the sternocleidomastoid muscle in zone II of the neck measuring 13 X 6cm, firm in consistency and non-tender, not attached to the overlying skin and mobile in the horizontal plane.

The neck ultrasound scan (USS) revealed a well-defined rounded wider-than taller hypoechoic mass measuring 4.4cm X 3.7cm X 5.2 cm noted posterior to the right lobe of the thyroid gland separated from it by a thin echogenic rim displacing it superior-medially. No calcifications were seen within it and it did not show flow on colour Doppler interrogation.

Fine needle aspiration for cytology (FNAC) revealed moderately cellular clusters of spindle-shaped cells. The individual cells were fairly uniform having oval to elongated nuclei with moderate cytoplasm, numerous lymphocytes were present in an amorphous eosinophilic background. The report suggested a spindle cell tumour in keeping with schwannoma.

She had an examination under anaesthesia and excision of the mass. The intraoperative finding was that of a right-sided neck mass measuring 10 x 5cm which was lobulated and yellowish, Figure 1A. The index case had a lateral cervical approach via a skin crease incision (Figure 1B) for total excision of the tumour. Macroscopically, the mass was an irregular fibrofatty tissue which measured 6x4x4 cm, Figure 1C and D, it was firm and the cut surface was grey-white.

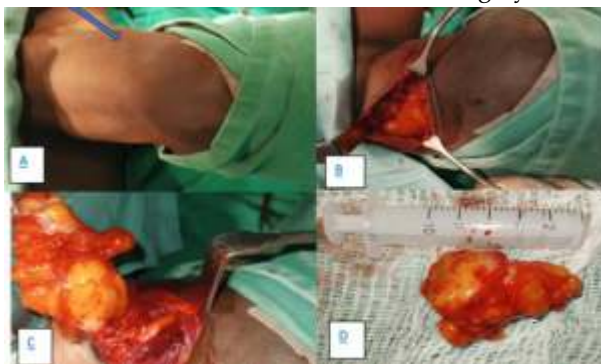


Figure 1: A: Picture of the lateral neck mass as indicated by the blue arrow. B: mass exposed intraoperatively. C: dissection of the mass of surrounding tissues. D: the excised mass

The histology result reported a circumscribed mesenchymal neoplasm disposed in alternating hypercellular (Antoni A) and hypocellular (Antoni B) zones. The hypercellular areas were composed of fascicles of spindle-shaped cells that have hyperchromatic ovoid to elongated wavy serpentine nuclei and moderately fibrillary cytoplasm. Foci of nuclear palisading with occasional Verocay body formation were present. The hypocellular areas comprised of scant ovoid to spindle-shaped cells within a loose and myxoid stroma. Areas showing hyalinized vasculatures and stromal hyaline were also present as shown in Figure 2. The patient was followed up for six months and was doing well as at the time of this report.

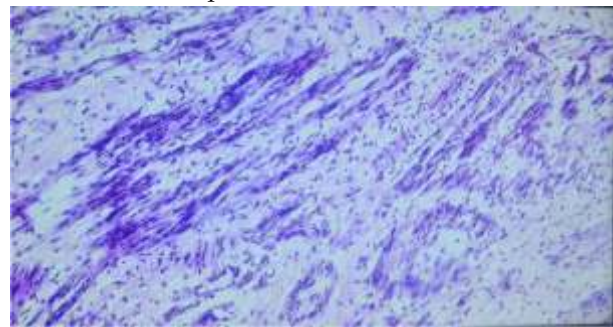


Figure 2: photomicrograph of lateral neck mass using the H & E stain (X10)

Discussion

Schwannomas are rare peripheral nerve sheath tumours which occur more commonly in the head and neck region. It is a benign tumour originating from cranial, peripheral, or autonomic nerves except for the optic and olfactory nerves.^{10,12} It occurs more commonly in the 4th to 6th decades of life, the index patient was in her 5th decade of life. Schwannomas usually present as a painless slow-growing lateral neck mass which may be palpated along the medial border of the sternocleidomastoid muscle similar to the index case.^{2,3,5} They are usually asymptomatic, but when symptoms occur, hoarseness and Globus sensation are the most common symptoms of vagal schwannomas.^{1,5,6} Other symptoms include dysphagia, dyspnoea, and neural deficit. Pain may be a feature of sensory nerve schwannomas.^{5,6} Facial nerve paralysis and Horner's syndrome may be seen in schwannomas of the facial nerve and cervical sympathetic chain respectively.^{3,4,5} The index patient had no obstructive, neurologic or toxic symptoms.

The diagnosis of schwannomas pre-operatively may be difficult as the history and examination may be

non-specific and there are many tumours in the neck which should be differentiated from schwannomas.^{4,5} Differential diagnoses of vagal schwannomas include paragangliomas, metastatic lymph nodes and schwannomas of the cervical sympathetic chain.^{3,4,6,15} These tumours are rare and may not be thought of initially.^{4,5} Fine needle aspiration cytology (FNAC) is usually non-specific and inconclusive.^{6,7,18,19} The index case had FNAC that is suggestive of spindle cell neoplasm. The neck ultrasound scan finding revealed a well-defined hypoechoic mass posterior to the right lobe of the thyroid gland was initially suggestive of a parathyroid adenoma. Similarly, Pankratjevaite *et al.*¹³ reported that the neck USS they performed misdiagnosed the neck mass to be a malignant lymph node. This emphasizes the fact that neck USSs are unreliable in assessing vagal schwannoma. CT scan or MRI were not done in the index patient. MRI is considered the best imaging modality to assess the tumour's origin and extent.^{15,16,17} Vagal schwannomas on MRI typically appear as a well-circumscribed mass between the internal jugular vein and the carotid artery. It displaces the internal jugular vein laterally and the carotid artery medially. Schwannoma appears as an isotense or hypointense lesion on T1-weighted images but hyperintense on T2-weighted images.^{1,14}

The mainstay for the treatment of schwannomas is surgical excision. Tumours confined to the neck can be approached through transcervical, transcondylar, cervical trans-mastoid or infra-temporal fossa approach.

The goal of surgical treatment is total removal of the nerve sheath tumour while preserving the intact fascicles, even though there is a risk of injury to the nerve of origin.^{2,3,5} To reduce the risk of nerve injury and recurrence, intracapsular enucleation of the tumour was advocated.¹ The rate of recurrence is low if a complete surgical excision is done.^{15,16,19} The index case had a lateral cervical approach via a skin crease incision for total excision of the tumour.

Macroscopically, schwannomas appear as a well-circumscribed yellowish-white mass with a well-defined capsule.^{1,11,13} They are solid or cystic mesenchymal tumours. There are two patterns microscopically the Antoni A and Antoni B patterns.^{6,15} The Antoni A pattern where there is a cluster of spindle-shaped cells arranged in bundles, whorls or palisades.

The Antoni B pattern contains fewer cells and also has other features, like cystic changes or xanthomatous changes.^{6, 15,18,19,20} These features have been demonstrated in the index case. Necrosis, haemorrhage and cystic degeneration are other specific features observed as histologic findings^{19,20}, though not demonstrated in the index case.

Conclusion

Vagal schwannomas are rare tumours of the nerve sheath. They should be considered when patients present with a slow-growing lateral neck mass, especially when symptomless and in close relation to the sternomastoid. There is usually a diagnostic dilemma and diagnosis is only confirmed on histology post excision. Complete surgical excision is the treatment of choice of these tumours. There is a need to counsel the patients on neurological deficits should the nerve be damaged iatrogenically.

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