

Case Report

Cervical schwannoma: diagnosis and treatment

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ABSTRACT

Schwannomas are benign, encapsulated, slow-growing, tumours deriving from the peri neural cells located in the nerve sheath. They can arise from any cranial, peripheral or autonomic nerves, and show a predilection for the head and neck region. They may produce secondary symptoms like nasal obstruction, dysphagia, and hoarseness of voice depending upon the location of the tumour. Preoperative diagnosis is difficult, relying on clinical suspicion, and confirmed by surgical pathology. Preoperative imaging or fine needle aspiration cytology may help to reveal diagnosis. The definitive diagnosis is made by histopathological examinations. A 28 years old female patient presented to the OPD with painless swelling on the left lateral side of the neck since 1 year. The swelling was mobile, non-tender, non-pulsatile firm in consistency measuring 4×3 cm with no bruit. MRI neck with contrast was done which revealed a moderately enhancing lesion of size 7×4×3 cm in the left carotid sheath. The tumour was approached by an anterior approach along the medial border of the sternocleidomastoid. Platysma and fascia were dissected followed by carotid sheath to reach the tumour. As the mass was seen arising from a branch of the ansa cervicalis, it had to be sacrificed.

Keywords: Para pharyngeal space, Schwannoma, Benign tumour, Ansa cervicalis, Anterior approach, Horner syndrome

INTRODUCTION

Schwannoma or neurilemmoma are benign neoplasms of Schwann cell origin except optic and olfactory nerves.¹ The Schwann cell surrounds peripheral nerve tissue and is believed to originate from the neural crest. They are typically solitary, well-encapsulated, benign tumours characteristically running along the course of a nerve or attached to peripheral, cranial, or sympathetic nerves.² It is very occasionally malignant. It is the commonest neurogenic tumour found in the parapharyngeal space. Here, schwannomas may arise from the last four cranial nerves or the autonomic nerves, the vagus being the most common site.

The histological appearance can be of 2 types: Antoni A: Consists of pattern of elongated spindle cells forming a palisade of nuclei around central mass of cytoplasm.

Antoni B: Consists of cells in loose myxoid stroma. Cervical sympathetic chain schwannomas are uncommon and most often appear as an asymptomatic, slow-growing, solitary neck mass; Horner's syndrome is rarely apparent on physical examination. Pre-operative diagnosis may prove difficult, for evaluation of neurilemmoma, contrast-enhancing computed tomography (CT) or magnetic resonance imaging (MRI) and ultrasound (US) are the most appropriate, carotid angiography is not necessary. Horner's syndrome is a post-op complication of the surgery, due to the necessary division of the cervical sympathetic chain, but this does not appear to have an adverse effect on the patient.

The choice of the surgical approach depends on the size of the tumour. For tumours of the post styloid compartment of the parapharyngeal space, transcervical approach is adequate.

Injury to the carotid artery and internal jugular vein can result in profuse bleeding intraoperatively.

CASE REPORT

A 28 years old female patient presented to the OPD with painless swelling on the left lateral side of the neck since 1 year. There was no history of dysphagia, generalised weakness, change in voice or neurological deficits. The swelling was mobile, non-tender, non-pulsatile firm in consistency measuring 4×3 cm with no bruit. MRI neck with contrast was done which revealed a moderately enhancing lesion of size 7×4×3 cm in the left carotid sheath. Features were suggestive of a nerve sheath tumour.

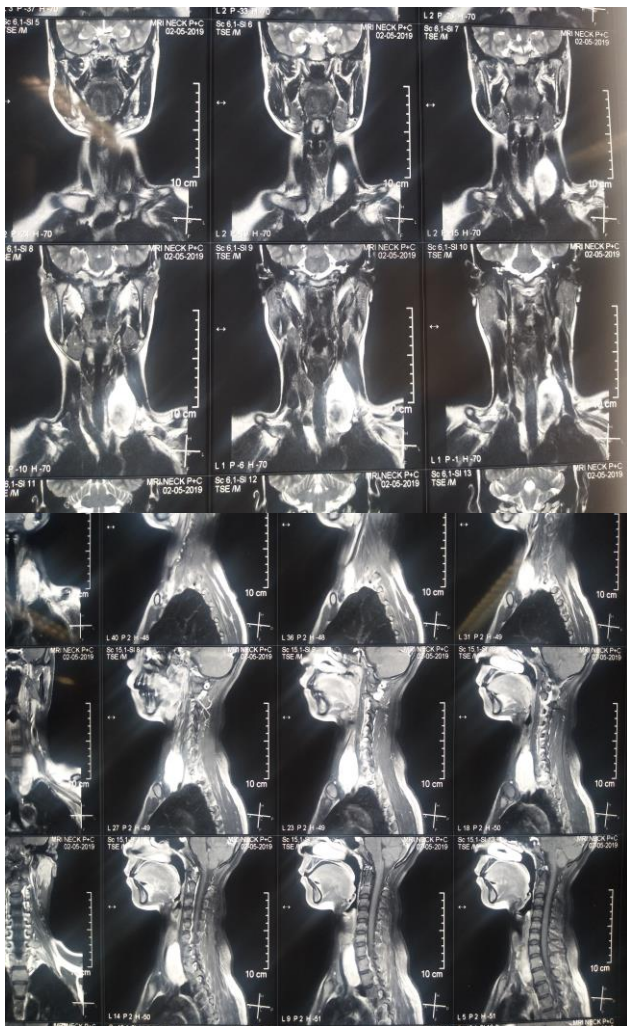


Figure 1: MRI neck with contrast: moderately enhancing lesion of size 7×4×3 cm in the left carotid sheath.

The tumour was approached by an anterior approach along the medial border of the sternocleidomastoid. Platysma and fascia were dissected to reach the tumour. Dissection was done till the carotid sheath was reached, which was opened.

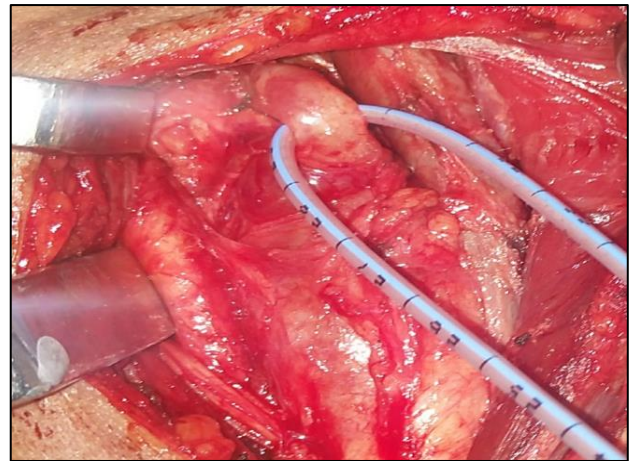


Figure 2: Mass seen arising from Ansa cervicalis in carotid sheath.

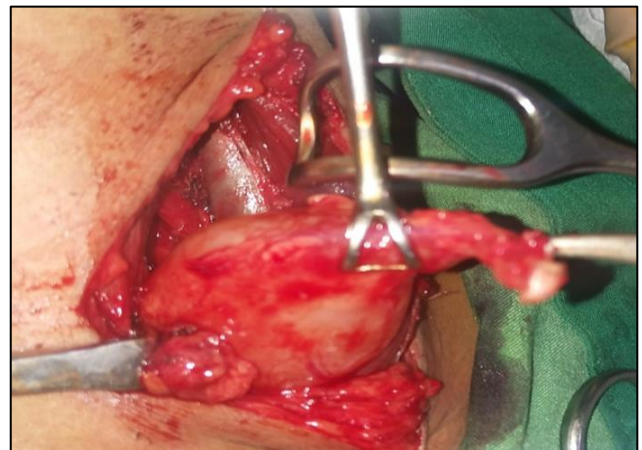


Figure 3: Globular, encapsulated mass.

The mass was found to be arising from a branch of the Ansa cervicalis between the carotid artery and internal jugular vein. It was a globular smooth mass, well encapsulated. As the mass was seen arising from a branch of the Ansa cervicalis, it had to be sacrificed.

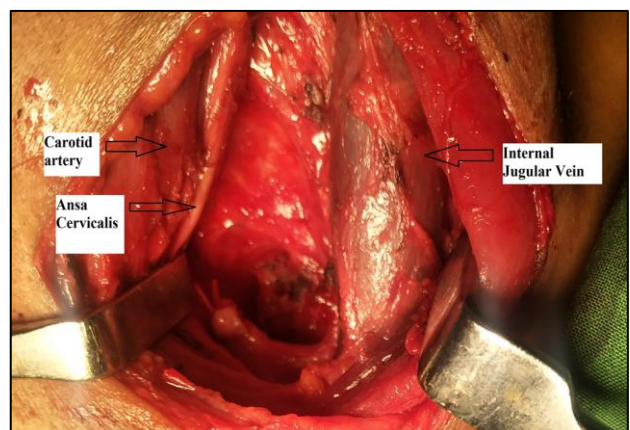


Figure 4: Carotid sheath following removal of the mass.

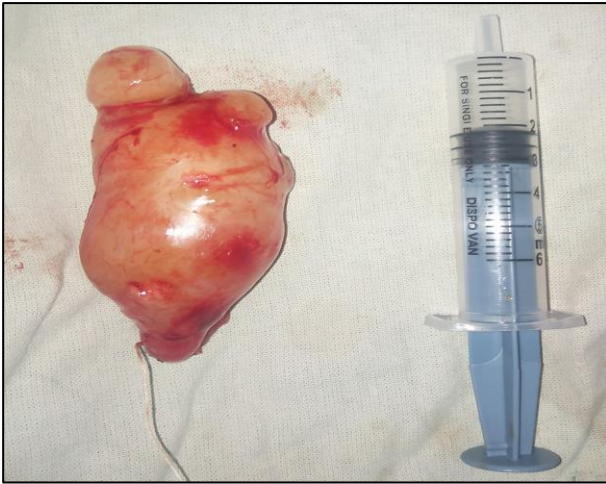


Figure 5: Mass removed and sent for histopathology.

The mass was found to be extending till the left supraclavicular fossa. The mass was separated all around, removed in-toto and sent for HPE. Closure was done in layers over a drain. Drain was removed after 72 hrs. HPE report confirmed schwannoma.

The patient developed Horner's syndrome in the post-op period which gradually recovered. Direct laryngoscopy revealed that both vocal cords were mobile. Horner's syndrome was due to injury to the branch of the Ansa cervicalis from which the mass arose.

DISCUSSION

Neurogenic tumours arise from the neural crest which differentiates into the Schwann cells and the sympathicoblasts. The Schwann cells give rise to neurofibroma and neurilemmoma (Schwannoma).⁵ A schwannoma is a slow growing solitary and encapsulated tumour attached to a nerve. Degenerative changes such as cystic alterations and haemorrhagic necrosis are seen in schwannoma, whereas such changes are not seen in neurofibroma.⁶ Schwannoma may arise from any cranial or spinal nerve that as a sheath i.e., any motor or sensory nerve other than the optic and the olfactory nerves which do not have the Schwann cell sheath. Schwannoma was first established as a pathological entity by Verocay in 1908 who later called it neurinoma in 1910. Later the term neurilemmoma was coined by Stout in 1935. Parapharyngeal space is the site of schwannoma in head and neck region accounting for 25-40% of all reported patients. The first case of neurilemmoma within the parapharyngeal space was reported in 1933 by Figi. Apart from parapharyngeal space other sites in the head and neck like submandibular space, paranasal sinuses, cheek and, oral cavity are rare. The size of the tumour may vary from few mm to over 24 cm. Majority of the patients present with a painless mass and pain may be present only in few cases. Other symptoms may be difficulty in breathing, dysphagia, epistaxis, hoarseness or only a swelling in the neck (Parapharyngeal space). The

swelling is freely mobile in soft tissues, but when it is connected to a large nerve or trunk there is restriction of the movements. The schwannoma may arise at any age and there is no gender or race.⁷ In the neck, the schwannoma has been divided into medial and lateral group. Medial group arises from the last four cranial nerves and cervical sympathetic chain. Lateral group arises from the cervical nerve trunk, cervical plexus and the brachial plexus. Schwannoma may also originate from vagus nerve, sympathetic chain or the glossopharyngeal nerve.⁷ Radiotherapy should be reserved for palliation only in cases of inoperable tumours.⁷ A case has been presented in submandibular region which shows atypical site of origin.⁸ The preoperative diagnosis of schwannoma in the head neck region is difficult. Most of the investigations like CT, FNAC (Fine needle aspiration cytology) or magnetic resonance imaging (MRI) are generally done. MRI has many advantages and has excellent soft tissue contrast. On the other hand, CT is useful where bone is involved like pterygoid plates or bone of posterior wall of maxillary sinus.⁹ Treatment is surgical excision. The tumour is radio resistant and the possibility of the malignant change is extremely rare. Histopathologically, it has typical presentation. Diagnostic features include fibrous capsule, hyaline vessels, cellular (Antoni A) and loose textured (Antoni B) areas, Verocay bodies (opposing rows of spindle nuclei separated by anucleate rows of eosinophilic processes). Retrogressive changes are common in large, old tumours, and include "degenerative nuclear atypia," vascular sclerosis and haemorrhage as well as occasional micronecroses. Microcyst formation, some with a pseudo epithelial lining of plump Schwann cells may also be seen.¹⁰

CONCLUSION

Cervical schwannomas, mostly present as asymptomatic unilateral neck masses, are rare tumours. The preoperative diagnosis may be difficult. The definitive diagnosis relies on clinical suspicion and histopathological confirmation. In the treatment of head and neck schwannomas, complete surgical excision with appropriate approaches is sufficient. There are chances of vagal or sympathetic chain injury during surgery. Local recurrence is extremely rare.

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