

Schwannoma of Parapharyngeal Space

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ABSTRACT

Parapharyngeal tumors are rare and their surgical excision is challenging because of the anatomical complexity of the area. They are late to present and symptomatology is varied till they become large in size. Herewith, a rare case of schwannoma probably arising from the cervical sympathetic chain is reported presenting as a mass in the parapharyngeal space.

Keywords: Schwannomas, Parapharyngeal space, Parapharyngeal tumors, Cervical sympathetic chain.

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INTRODUCTION

Tumors of parapharyngeal space account for only 0.5% of all head and neck tumors. They comprise varied histological types, the common being salivary gland tumors, schwannomas and paragangliomas. Almost two-third of the tumors are benign and only one-third are found to be malignant.¹ A rare case of schwannoma probably arising from the cervical sympathetic chain is reported presenting as a mass in the parapharyngeal space.

CASE REPORT

A 18-year-old male presented with complaints of asymptomatic swelling in the left side of the neck since 4 years. Examination revealed a diffuse swelling in the left side of the neck in carotid triangle. A pulsating vessel was seen running along the anterior part of the swelling (Fig. 1). There were no palpable neck nodes. Throat examination revealed a visible bulge in the left tonsil and posterior pillar. On indirect laryngoscopy, left lateral pharyngeal wall was displaced medially. Both vocal cords were mobile. Cranial nerve examination was normal. A provisional diagnosis of left parapharyngeal mass was made. Contrast enhanced computed tomographic (CECT) scan revealed an isodense mass in left carotid space with nonhomogenous enhancement measuring 7.5×4.8 cm with central hypodense area pushing the carotid artery anteriorly and compressing the jugular vein with dilatation of the vein below the mass. The lesion is pushing medially the glottis and the lateral pharynx with mild compression of trachea. No neck lymphadenopathy (Fig. 2). Ultrasound guided fine needle aspiration cytology (FNAC) few clusters of oval to

spindle cells having bland nuclear chromatin in myxoid stroma against a hemorrhagic background. Magnetic resonance (MR) angiography was done which showed a well-defined space occupying lesion in the left carotid space displacing left common carotid artery and carotid artery bifurcation anteriorly and internal jugular vein anterolaterally suggestive of vagal schwannoma (Fig. 3). Twenty-four hours urinary vinyl mandelic acid assay found to be within normal range. Patient was taken up for excision of the mass through a transcervical approach. A curvilinear incision made from behind the angle of the mandible to cricoid cartilage. Dissection was made in subplatysmal plane. Left sternocleidomastoid was retracted and carotid sheath was opened. A small nerve twig was attached to the tumor in its inferior aspects and vagal nerve was found to lie separate from the tumor and was intact. The tumor was found to be well-encapsulated and was excised *in toto* (Fig. 4). Postoperatively, patient was found to have pseudoptosis of left eye. Grossly, the tumor measured $7.5 \times 4.5 \times 4$ cm (Fig. 5). Microscopic sections of the tumor areas revealed hypocellular and hypercellular areas with interspread thick- and thin-walled blood vessels. The hypercellular areas show plump spindle-shaped tumor cells arranged in interlacing fascicles, whorls and palisading pattern with formation of Verocay bodies. Impression was of a schwannoma.

DISCUSSION

Schwannomas were first described by Verocay in 1908. Since that time, they have been called neurilemmomas, solitary nerve sheath tumors, perineural fibroblast tumors



Fig. 1: Preoperative photograph of the patient showing left-sided neck swelling

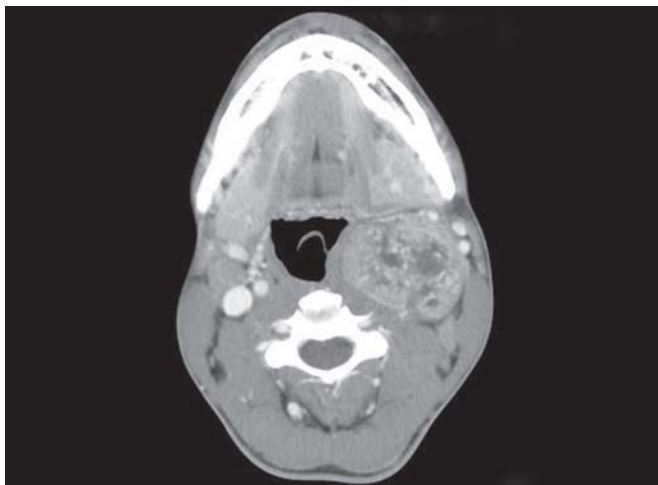


Fig. 2: CECT neck showing the parapharyngeal mass occupying the poststyloid compartment

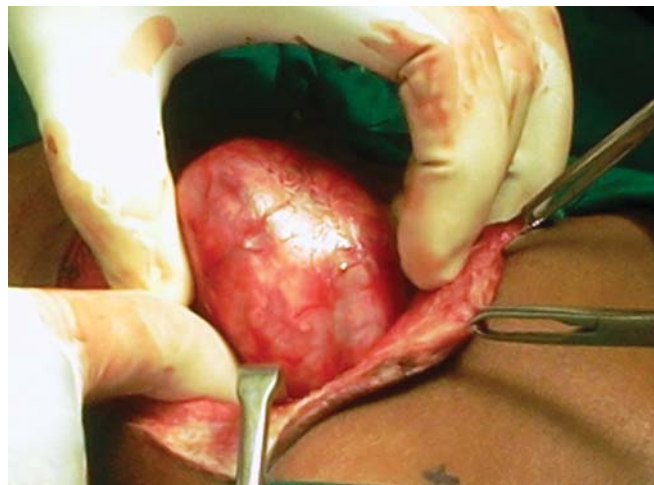


Fig. 4: Intraoperative photograph showing the tumor being exposed

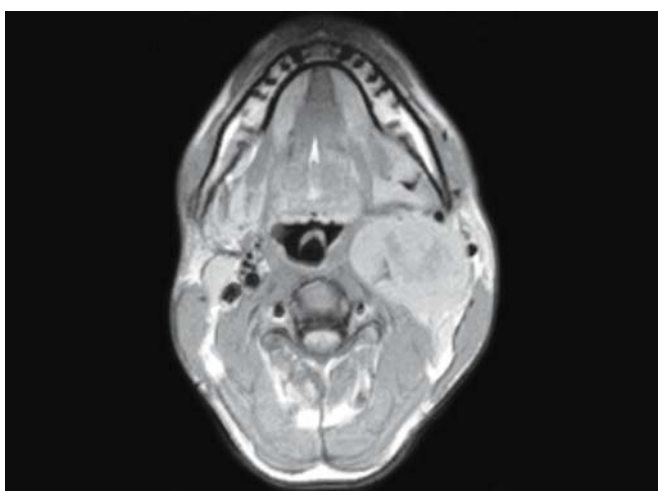


Fig. 3: MR angiography T1-weighted axial section image showing mass occupying left carotid space



Fig. 5: The excised specimen

and, most recently, schwannomas, according to the World Health Organization (WHO) classification.² Schwannomas are uncommon nerve sheath neoplasms that may originate from any peripheral, cranial or autonomic nerves of the body with the exception of the olfactory and the optic nerves.³ In head and neck region they most commonly arise from the parapharyngeal space. The most common nerves of origin were the vagus and the cervical sympathetic chain.⁴ Schwannomas are almost always diagnostic problems because their history and clinical examination are nonspecific and deceptive.³

Neurogenic tumors of head and neck arise from neural crest cells which differentiate into nerve sheath cells and sympathoblast. Schwann cell is the parent cell of both schwannomas and neurofibroma. Neurofibroma also has an origin from the perineurium and is thus linked inseparably from the nerve of origin.⁵ Schwannomas grow longitudinally along the length of the nerve assuming a fusiform appearance without compromising the morphological and

functional integrity of the nerve and can be separated surgically from their nerve of origin. Distinction between schwannomas and neurofibroma can be made microscopically.⁶

Most important aspect of these lesions is distinguishing cervical schwannomas from other pathologies like salivary gland tumors and paragangliomas in the parapharyngeal space. The differential diagnosis of a parapharyngeal mass is based on the division of the space into prestyloid and poststyloid compartments.⁶ Diagnostic modalities in the form of FNA cytological techniques as well as better imaging in the form of MRI or CT scans have lessened the problem of misdiagnosis to some degree.³ CT with contrast medium or MRI is essential to the initial workup for parapharyngeal space schwannoma. A mass pushing the internal carotid artery or common carotid artery anteriorly is suggestive of a schwannomas originating from the sympathetic chain or vagus nerve.⁵ Schwannomas being

well-encapsulated tumors can be completely removed during surgery. Selection of the approach to the parapharyngeal space is based on the site and size of the tumor keeping in mind the adequate exposure of the tumor and exposure of the neurovascular structures in the vicinity to preserve their function and control bleeding. Transcervical, transmandibular and transparotid are the main approaches described to approach the parapharyngeal space.⁶ Although Horner's syndrome is rarely seen in the preoperative period, it is the most common complication postoperatively.⁷ In our case, a small nerve twig was seen attached to the tumor which was sacrificed and patient developed pseudoptosis postoperatively. Hence, schwannomas was probably arising from the cervical sympathetic chain.

CONCLUSION

Schwannomas originating from the cervical sympathetic chain are rare in the neck. These lesions are commonly asymptomatic or present with nonspecific symptoms, and accurate preoperative diagnosis is not always easy. An accurate preoperative workup is very important not only for a correct diagnosis, but also for surgical planning and informing the patient about the possible complications.

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