CASE REPORT PROF-1449

SCHWANNOMA OF THE CERVICAL SYMPATHETIC CHAIN;
CLINICAL AND THERAPEUTIC CONSIDERATIONS

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ABSTRACT ... Schwannomas are benign, slow-growing tumors that arise from Schwann cells of the nerve sheath. Those originating from the sympathetic cervical chain are extremely rare and usually presents as an asymptomatic neck mass. Here we report a case of schwannoma of the cervical sympathetic chain in a 32-year-old man who had asymptomatic neck mass for 6 months. But it appeared pulsatile due to the anterior displacement of carotid sheath by the mass and thus mimics a carotid body tumor. As discussed in this report CT scan with contrast is enough for ruling out paraganglioma and imposing extra expense for MRI and angiography is unnecessary .The only rare complication encountered after surgery was Horner's syndrome, which required no treatment.

Key words: Schwannoma, Sympathetic chain, Head and neck.

INTRODUCTION

Schwannomas are typically benign, slow growing, solitary tumors arising from the nerve sheath of a Schwann cell. Schwannomas can arise from any cranial, sympathetic, or peripheral nerve. Cervical schwannomas are uncommon; those arising from the cervical sympathetic chain are extremely rare[1]. Cervical sympathetic chain schwannomas, Like most schwannomas, are most often solitary, benign, and slow-growing, with rare malignant degeneration[2,3]. Patients commonly present with an otherwise asymptomatic neck mass. Surgery is the treatment of choice and major complications are infrequent. However postoperative Horner's syndrome and vagus nerve dysfunction or first bite syndrome has been reported[4,5]. Here we have presented a cervical sympathetic chain schwannoma which was pulsatile and postoperatively, patient developed Horner's syndrome.

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CASE REPORT

A 46-year-old man presented to the outpatient department of otolaryngology of the University of Shiraz (Iran) complaining of a painless, enlarging, left-sided neck mass that was first noticed approximately 6 months before presentation. Physical examination revealed a 6×4 cm pulsatile mass at the level of angle of mandible. In oral examination lateral wall of pharynx was pushed medially.

Computed tomographic (CT) scans of the skull base and neck revealed a un-enhanced non homogenous circumscribed lesion in the left parapharyngeal space that has caused anterior displacement of carotid and jugular sheath with separation between common carotid and jugular vein (Fig. 1). Fine needle aspiration was not diagnostic.

The patient was taken to the operating room, where a

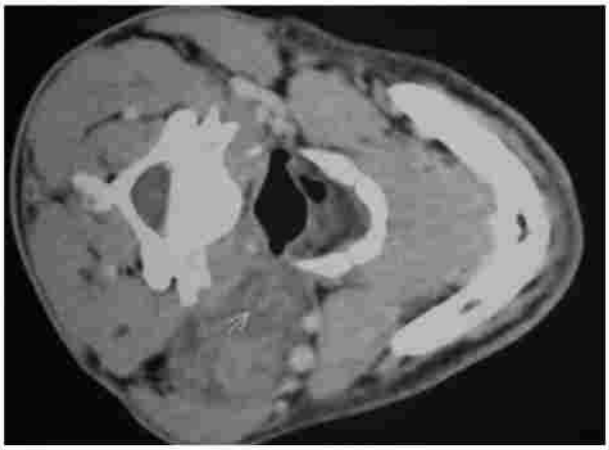

Fig-1. An axial post-contrast CT scan showing left-sided parapharyngeal space non enhancing lesion with the common carotid artery and jugular vein displaced together in an anterior direction.

transverse cervical approach was undertaken. The tumor was found to arise from the sympathetic chain. The mass was unable to be resected without sacrificing a portion of the chain. The excised mass was well circumscribed, creamy, ovoid mass with smooth shiny external surface and measured 6 × 4 × 3cm (Fig. 2). Final pathologic evaluation was consistent with a benign schwannoma.

Fig-2: Surgical specimen of well circumscribed creamy ovoid sympathetic chain schwannoma

Histologic sections show benign spindle cell proliferation with hypercellular and hypocellular areas. The spindle cells are arranged in fascicles. The nuclei show wavy appearance with in some areas show palisading and forming verocay bodies. Some areas of myxoid degeneration were also noted (Fig. 3).

Postoperatively, the patient exhibited left miosis and mild

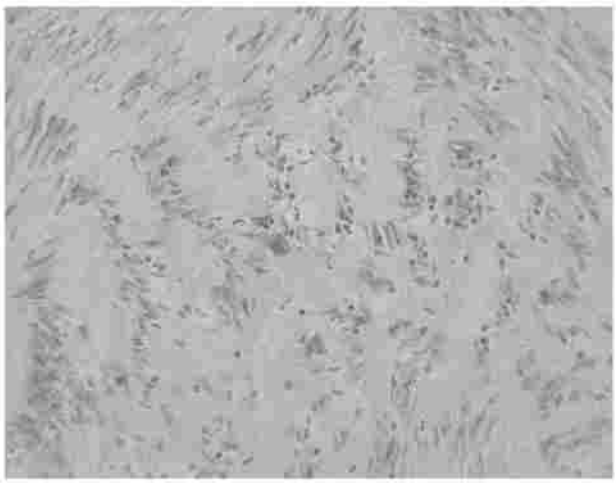

Fig-3. Histologic section of sympathetic chain schwannoma demonstrating palisading of spindle cell nuclei and verocay bodies. (H&E; x 160)

left ptosis, and conspicuous compensated right mydriasis which was worrisome to the patient but it is resolving slowly (Fig. 4).

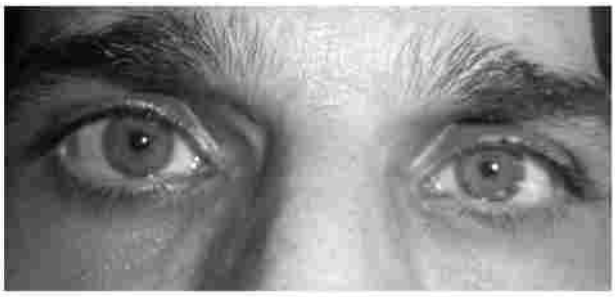

Fig-4. A view of patient with left enophthalmos, miosis, and ptosis that partially recovered 4 months after surgery.

DISCUSSION

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Schwannomas are slow-growing, encapsulated tumors arising from the perineural cells located in the peripheral nerve sheath. The reported sites of origin of cervical schwannomas are the cranial nerves IX-XII, the sympathetic chain, the cervical plexus, and the brachial plexus[6].

Fine needle aspiration, while conclusive in many cases of neck masses, is much less valuable for the compact neural tumor, and failed to reveal the diagnosis in most cases in whom it was performed[7]. So Imaging studies play a central role in the diagnosis of the head and neck schwannoma. The most important task is to distinguish between a vagal or sympathetic schwannoma and a paraganglioma[9].

As in our case; schwannomas regardless of the nerve of origin in general are hypodense with comparison to muscle on CT without contrast and post contrast, these lesions show at least some degree of enhancement, often at the periphery[8]. Paragangliomas, on the other hand, are classically isodense when compared to muscle on precontrast CT, with more reliable homogeneous enhancement postcontrast[8].

Saito et al has noted the schwannomas of the cervical sympathetic chain were found to displace both the carotid and jugular vessels without separating them but interestingly in our case they has been separated[10]. On the other hand vagal nerve schwannomas were found to separate the carotid arteries from the internal jugular vein. A vagal nerve schwannoma may also displace the sheath vessels posteriorly, without splaying them[10].

Recommended treatment is usually surgical resection but because of the intimate relationship to the nerve of origin it is often difficult to preserve the function of the nerve[11]. In patients with sympathetic chain schwannomas post operative Horner's syndrome is almost seen in other reports. Horner's syndrome, which presents with miosis, blepharoptosis, enophthalmos, and anhidrosis, derives from an interruption of the sympathetic pathway at any point along its course between the hypothalamus and the orbit and the most common factor producing the syndrome is tumors[12].

Post operation Horner's syndrome although may be worrisome for the patients but it seems to not cause any adverse effects or complaints for the patients[13].And as our patient their clinical finding is relieved gradually.

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