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Case Report

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An Unusual Case of Horner's Syndrome: A Rare Case of Cervical Chain Schwannoma

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Abstract

Schwannomas are benign, solitary nerve sheath tumours arising from the Schwann cells. They can arise from any of the cranial, spinal or autonomic nerve except for olfactory and optic nerve. The commonest site for head and neck schwannomas is the parapharyngeal space with vagus nerve being the most common the site of origin. On the other hand, schwannomas arising from cervical sympathetic chain is very rare entity. These tumours usually present as slow growing neck masses without any neurological deficit. Contrast enhanced imaging of neck helps in delineating the exact location and the nerve of origin. Surgical excision is the treatment of choice for cervical schwannomas. Malignant transformation is rarely seen. Here, we report a rare case of cervical chain schwannoma in a young female presenting with a bulge in oropharynx associated with a neck mass, managed surgically.

Introduction

Schwannomas which are also called as Neurilemmomas are benign tumours arising exclusively from Schwann cells. The tumour can originate from any myelinated central or peripheral nerve. Schwann cells are glial cells that form the myelin sheath. Schwannomas are usually solitary, therefore multiple schwannomas should arise the suspicion of a syndrome. They are sporadic in nature in 90% of cases. Schwannomas constitute 25-45% of tumours of the head and neck [1]. Parapharyngeal space is the commonest site for head and neck schwannomas. In the parapharyngeal space, these tumours can arise from any of the last four cranial nerves or cervical sympatheric chain. The cervical sympathetic chain schwannomas are rare. These tumours usually originate from the superior or middle part of the cervical chain which is typically located in the retrostyloid compartment of the parapharyngeal space [2]. Contrast enhanced computed tomography (CECT)/Magnetic Resonance imaging (MRI) of neck is important for management of these tumours. Histologically, Antoni type A and type B cells are seen with S-100 positivity on immune histochemistry. Here, we describe an interesting case of cervical sympathetic chain schwannoma (CSCS), presented as an oropharyngeal mass. The mass was excised surgically and patient developed Horners syndrome postoperatively.

Case Report

A 27-year-old female presented to Otorhinolaryngology Department of our Hospital with complaint of discomfort while swallowing since 2 yrs, which was insidious in onset, gradually progressive, more towards solids and not relieved on medication. Patient noticed bulge in throat on right side since 1 year which was progressive in nature. There was no complaint of change in voice or difficulty in breathing. On general examination, patient was comfortable. There was no respiratory distress. There was no halitosis or drooling of saliva. The cranial nerve examination was normal. On oropharyngeal examination, there was a mass in lateral pharynx which was pushing the right tonsillar fossa medially

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Figure 1: Clinical examination showing the oropharyngeal bulge on right side.

and uvula to the left side thereby remarkably reducing the oropharyngeal space and obscuring the posterior pharyngeal wall (Figure 1). On palpation, the mass was firm, non-tender, non-pulsatile, and non-compressible. On examination of neck, there was no obvious swelling seen on inspection but on palpation, an ill defined fullness could be palpated behind the angle of mandible. On fibre optic laryngoscopy, vocal cords examination was normal.

CE MRI neck was done which suggested well defined encapsulated oval shaped mass lesion of size 3 cm × 4.2 cm × 5.3 cm in right suprahyoid, parapharyngeal space extending into retropharyngeal space showing fascicular sign with heterogenous post contrast enhancement. Lesion was seen displacing right internal carotid artery (ICA) and internal jugular vein (IJV) laterally without any splaying of vessels (Figure 2). Intraoral FNAC was done which showed stromal cells interspersed with spindle cells, suggestive of nerve sheath tumour. Therefore, a preoperative diagnosis of nerve sheath tumour arising from parapharyngeal space was made. Transcervical approach was planned for excision since it provides better control of neck vessels. Skin incision was given at level of hyoid on the right side and subplatysmal flaps were elevated. Sub-mandibular gland was retracted superiorly and parapharyngeal space was exposed. Hypoglossal nerve was identified and preserved. The mass was seen occupying the poststyloid compartment of parapharyngeal space with great vessels pushed laterally. The mass was found extending to the retropharyngeal space as well. It was dissected from its attachments. Mass was found very closely adhered to ICA where it was separated by sharp dissection. Extracapsular dissection was done and finally it was removed by cutting its attachment from the neural structure, the cervical sympathetic chain. Vagus nerve was identified and preserved. Whole of the mass was removed in toto (Figure 3) and was

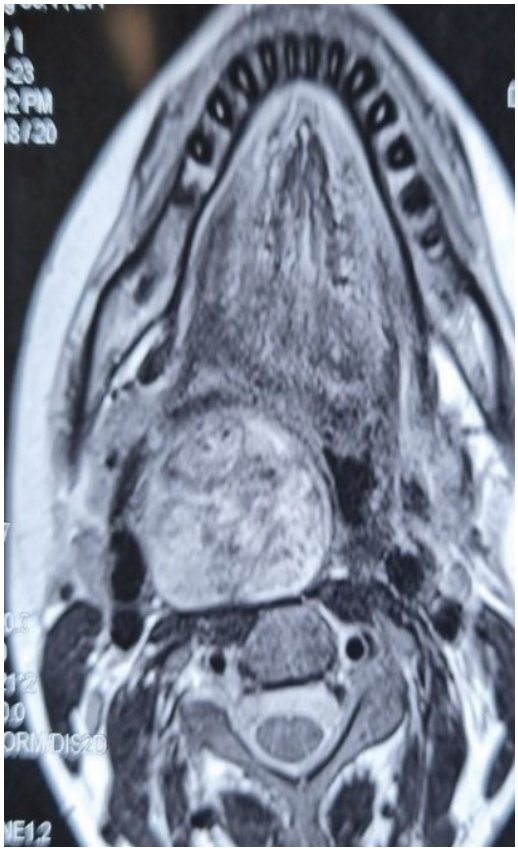

Figure 2: MRI neck showing the mass in parapharyngeal space.

sent for histopathological examination. Postoperative period was uneventful. Neurologically, all cranial nerve examination were normal. On post op day 1, patient developed mild ptosis of right eyelid which progressed on day 3. There was associated miosis. Therefore, a diagnosis of Horners syndrome was made. On gross examination, the mass measured 4 × 6 cm. Post-operative histopathology showed features suggestive of schwannoma (Figure 4). Patient is doing well without any recurrence at 6 months follow up. There is no major disability to patient due to Horners syndrome.

Discussion

Schwannomas are benign, painless tumours that grow slowly. Verocay published the first description of it in 1908, and Antoni followed suit in 1920 with a classical histopathological description of these tumours. The most frequent location for head and neck schwannoma is the parapharyngeal space, which is followed by the nasal cavity, oral cavity, and paranasal sinuses [3]. Parapharyngeal schwannomas usually present as asymptomatic masses in neck without cranial nerve weakness. Pain and neurological impairment might occasionally be signs of malignancy or degenerative changes in the tumour [4]. Though parapharyngeal schwannomas usually present as slowly growing mass in neck but in our case the swelling in the neck was ill defined and not very obvious. Rather it presented as swelling in the lateral pharyngeal wall. Since, there is usually no nerve deficit preoperatively, the nerve of origin is difficult to identify in cervical schwannomas. The presence of Horner's syndrome before excision is unusual

Figure 3: Gross specimen.

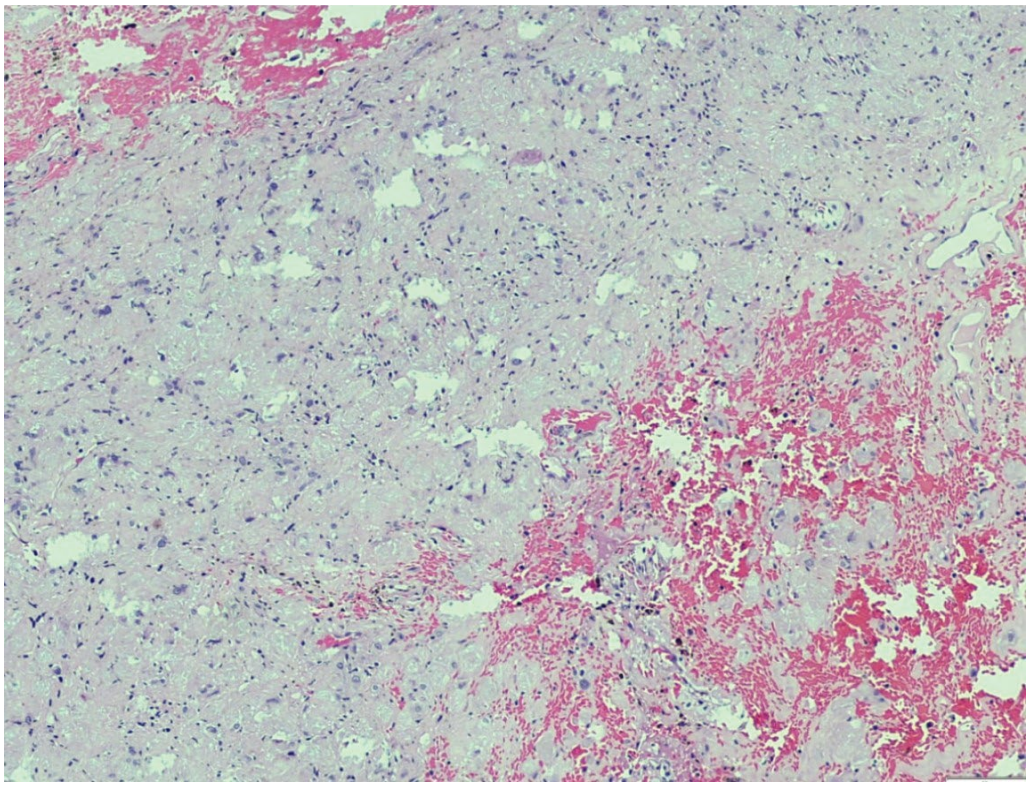

Figure 4: Histopatholgical photo.

and has only been reported in few cases of CSCS [5]. In our case also, there was no neurological deficit preoperatively.

Radiological investigations play an important role in ascertaining the extent of tumour as well as the nerve of origin. Radiologically, vagal schwannomas are seen splaying the Carotid artery and IJV as vagus lies in between them whereas no such separation is seen between them in CSCS.

Atypical presentations of schwannomas have also been reported. Samet Ozlugediketalv reported a case of cervical chain schwanoma presenting as a pulsatile neck mass associated with Horner's syndrome. Although splaying of the internal carotid artery and external carotid artery at the level of the bifurcation ("lyre sign") is accepted as a typical sign of carotid body tumours, recent papers have reported that a similar phenomenon can be found in CSCS. Therefore, the vascularisation degree of the lesion must be evaluated on angiography.

Demonstration of contrast agent accumulation, absence of arteriovenous shunts, and low-degree vascularity suggest schwannoma [6]. Microscopically, spindle-shaped cells in Antoni-A and Antoni-B arrangement interspersed with Verocay bodies are the characteristic features of schwannomas. These tumour cells typically show a positive immune reactivity for S-100 protein.

The treatment of choice of schwannoma is surgical excision with periodic follow-up. The choice of operation is mainly determined by the relationship between the tumour and the nerve of origin. Treatment of CSCS is total excision of the lesion. The majority of surgeons prefer intracapsular enucleation [7]. During surgery, the appearance of the tumour can suggest the right diagnosis, whether it presents as a fusiform mass or eccentric mass to the nerve. Complete surgical removal of the mass, without sacrificing nerve fibre, is possible only when the capsule is easily separable from the underlying fibers. Therefore, patient should be warned of the nerve deficits postoperatively. When dissection of the capsule from the nerve is not easy and there are no signs of malignancy, functional loss can be minimized by opening the capsule longitudinally and removing the tumour from inside. Most sympathetic chain schwannomas arise from superior cervical ganglion, with sympathetic ganglia being embedded in a meshwork of fibers. Thus, if a sympathetic nerve is the origin of the tumour, the preservation of neurologic function is difficult [8] as was seen in our case. Since cervical sympathetic chain damage is well tolerated, restoration of the nerve is usually not performed; while in vagal schwannomas, nerve reconstruction is recommended [5]. Peroperative neuro-monitoring may be useful in cervical nerve tumour, allowing partial tumour resection if need be - as in case of preoperative diagnosis of vagal schwannoma [9].

Conclusion

Schwannomas originating from the cervical sympathetic chain are rare in the neck. These lesions are commonly asymptomatic or present with non-specific 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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