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Parapharyngeal Tumours – A Report of 3 Rare Cases

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ABSTRACT
Parapharyngeal space is a potential inverted pyramidal space in the neck containing vital neurovascular structures. It has a complex and crowded anatomy and can house a variety of tumours. Diagnosis of such tumours can sometimes be a clinical as well as a radiological challenge and surgical excision and histopathological examination provides the confirmative diagnosis, often leading to surprises. We present 3 such cases, namely, a leiomyoma of parapharyngeal space, a trigeminal nerve schwannoma presenting as a parapharyngeal mass, and a synovial sarcoma of the parapharyngeal space, which were a diagnostic and surgical challenge

KEYWORDS: *Parapharyngeal, schwannoma, trigeminal, synovial, sarcoma, leiomyoma.*

INTRODUCTION

Parapharyngeal space tumors comprise approximately 0.5% of head and neck tumors[1], with more than 80 percent being benign. Common tumours of the parapharyngeal space are salivary gland neoplasms, paragangliomas and neurogenic tumours.

The parapharyngeal space has a complex anatomy, containing a variety of tissues, any of which could potentially turn neoplastic. Many rare tumours of the parapharyngeal space have been reported including lipomas, sarcomas and extracranial meningiomas. Owing to the structural crowding and silent presentation, parapharyngeal space tumours are a diagnostic and a surgical challenge.

Here, we present 3 rare tumours of the parapharyngeal space which presented to us between April 2017 and June 2017, and were successfully managed.

CASE 1

A 47 year old lady presented with a history of a large swelling in the left side of neck since 6 years. She also complained of snoring and dysphagia since 1 year. The swelling was painless and gradually progressive. On examination, the swelling was 10x12cm in size extending deep to the sternocleidomastoid laterally, deep to the ramus of the mandible superiorly, upto the level of cricoid cartilage inferiorly. The swelling was firm, nontender, with restricted mobility, non-pulsatile

and the skin over the swelling was pinchable. On oropharyngeal examination, the left tonsil was found to be pushed medially almost upto the midline. Contrast enhanced CT scan showed a partially circumscribed enhancing ovoid mass with some calcifications and lobulations. Fine needle aspiration revealed inadequate and haemorrhagic smears. In view of the unclear nature of the swelling and the suspicion of an unresectable malignancy, Core needle biopsy was performed which showed a spindle cell neoplasm arranged in fascicles composed of hyperchromatic nuclei with a pinkish eosinophilic cytoplasm with patchy areas of loose fibrovascular tissue. Based on the above report, transcervical excision of the tumour was performed. Intraoperatively, the tumour was identified lying deep to the internal jugular vein, which was found dilated and tortuous. The capsule of the tumour was found adherent to the internal jugular vein. Hence, the internal jugular vein was ligated and subcapsular excision of the tumour was performed preserving the vagus nerve. On removal of the tumour, 2 remnant lobulations of the tumour were seen, a larger lobulation lateral and posterior to the common carotid artery which was dissected and excised after identifying and preserving the sympathetic trunk. The other small highly vascular looking lobulation near the carotid bifurcation was left behind and followed up. The final histopathology of the specimen was surprisingly reported as a circumscribed mass composed of interlacing fascicles of spindle cells with eosinophillic fibrillary cytoplasm, oval nuclei having blunt ends, consistent with a leiomyoma.

CASE 2

A 34 year old lady presented with a history of a gradually progressive swelling in the right parotid region for 5 years. On examination, a 5x4 cm firm non-tender swelling was noted extending deep to the ramus of the mandible. A clinical diagnosis of a parotid tumour was made and a CT scan and FNAC was performed. To our surprise, CT scan showed a large mass involing the parapharyngeal space extending into the infratemporal fossa, severely thinning out the skull base indenting the greater wing of sphenoid, with a ring lesion in the right temporal lobe of brain. FNAC was suggestive of a neurilemmoma. However, to rule out a malignant parotid neoplasm and for better soft tissue demarcation, MRI with contrast was performed. MRI showed a 5.2x6.7x8.6cm lobulated mass, in the right masticator space involving the infratemporal fossa region epicentred on the mandibular division of the trigeminal nerve. The mass was also seen on the proximal part the themandicular nerve extending intracranially to the ganglionic segment in Meckel's cave also to the preganglionic segment in the ambient cistern. The mass was abutting and medially displacing the right internal carotid artery in the cavernous sinus. A neurosurgical opinion was taken and after thorough neurosurgical workup, a multidisciplinary approach right subtemporal craniotomy and transcervical approach near total excision of the tumour was performed. Final histopathology of the specimen showed spindle cells arranged in short bundles and interlacing fascicles, Verocay bodies, which confirmed the diagnosis of Schwannoma of the trigeminal nerve.

CASE 3

A 36 year old gentleman presented with a history of left sided neck swelling which was painless and gradually progressive since 3 years. On examination, there was a large firm swelling deep to the sternocleidomastoid with transmitted pulsations. On oropharyngeal examination, left tonsil was pushed medially upto the midline. MRI was performed which showed an encapsulated 6.7x 5.8x2.7cm lesion displacing the carotid bifurcation anteriorly. A diagnosis of Schwannoma of the parapharyngeal space was made and transcervical excision was performed. Intraoperatively, a friable tumour was identified posterior to the carotid sheath, extending laterally deeper to the sternocleidomastoid muscle, which was removed piecemeal. To our surprise, the

histopathological examination showed a spindle cell neoplasm arranged in lobules composed of predominantly epithilioid cellular neoplasm alternating with patchy areas of hypocellularity. There was increased mitosis. The intervening stroma showed biphasic pattern, suggestive of a malignant epitheliod and spindle sell neoplasm. On, immunohistochemistry the tumour was positive for Vimentin, PAN CK, S100, and EMA, confirming the diagnosis of a synovial sarcoma. Medical oncology opinion was taken, and due to complete resection of the tumour on postoperative imaging, patient was kept in follow up for future chemoradiation.

Figure1

Case1: Leiomyoma of the parapharyngeal space. Surgical specimen showing bilobed nodular mass measuring 10x8.5x6cm, with intact glistening capsule.

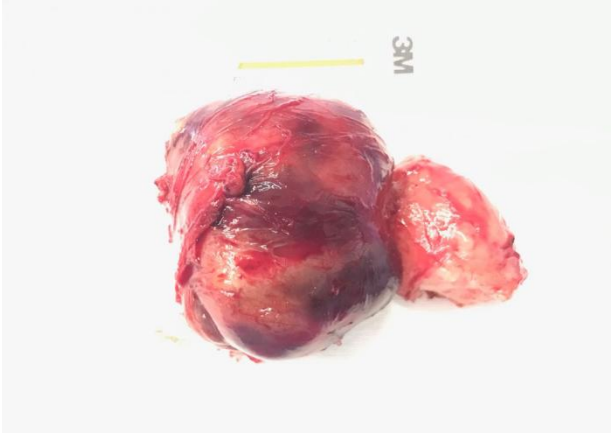


Figure 2

Case 2: MRI showing Schwannoma of Trigeminal nerve

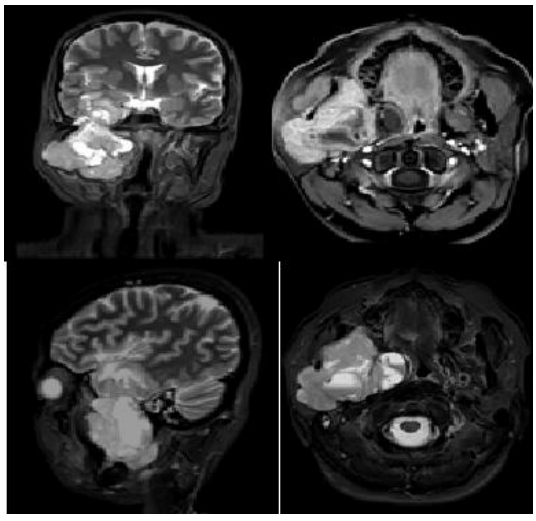


Figure 3

Case 2: Schwannoma of trigeminal nerve, near total excision by combined subtemporal craniotomy and transcervical approach

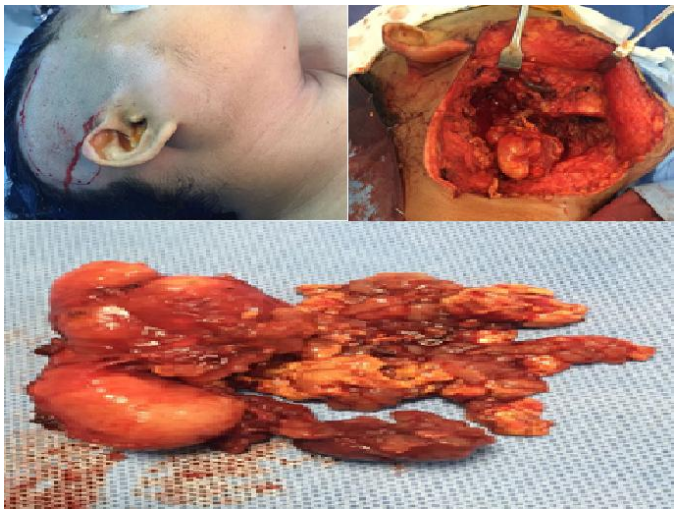
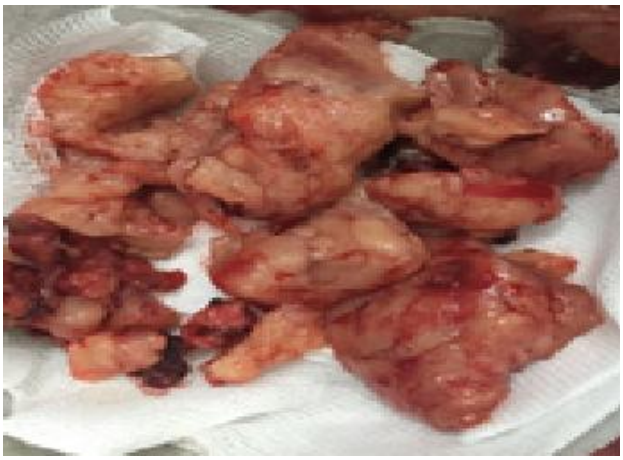


Figure 4: Synovial sarcoma of parapharyngeal space



DISCUSSION

Parapharyngeal space is a potential space in the neck in the form of an inverted pyramid extending from the skull base to the hyoid bone. The medial boundary is formed by the pharyngobasilar fascia and the superior constricter muscle and lateral boundary by the ramus or the mandible. The posterior boundary consists of the prevertebral structures. The styloid process with its muscles and condensations of fascia divide the parapharyngeal space into an anterolateral prestyloid and a posteromedial poststyloid compartment.

The structures contained in the anterior compartment include the pterygoid and tensor

palati muscles, fat and the deep lobe of the parotid gland. The poststyloid compartment contains the carotid sheath, with its contents. The compartment also contains the sympathetic trunk, the IXth, XIth and XIIth cranial nerves and the major part of the internal maxillary artery. Tumours of the parapharyngeal space are the tumours arising from the contents of the compartments.

Salivary gland neoplasms represent the majority of the prestyloid compartment tumours, most common being pleomorphic adenoma of the deep lobe of parotid. Less commonly, salivary neoplasms of the extraparotid tissue including Warthin's tumour, oncocytoma, and lymphoepithelial lesions have been reported. Tumours of the poststyloid compartment include paragangliomas like glomusvagale, carotid body tumours, neurogenic tumours like schwannoma and neurilemmoma from the vagus or sympathetic trunk. Malignancies in the parapharyngeal space are rare. Cases of malignant salivary gland tumours including adenoid cystic carcinoma, mucoepidermoid carcinoma soft tissues sarcomas of neurogenic or muscular origin have been reported.

Due to the anatomical complexity, crowding of structures and silent presentation, the diagnosis of the exact nature of the tumour is a challenge. FNAC (with or without image guidance), CECT, and MRI, all have their roles, but, not rarely, as in the 3 cases described in our report, surgical excision and histopathological examination alongwith immunohistochemistry provides the final diagnosis.

Leiomyomas are benign nonepithelial smooth muscle cell tumors. There are three histological types of leiomyomas: solid, vascular (angioleiomyoma) and epithelioid (leiomyoblastoma)[2,3]. Primary leiomyomas of the neck are extremely rare, and involvement of parapharyngeal space has only been described in countable case reports. Surgery is the recommended treatment modality for leiomyomas, with a very low recurrence rate.

Synovial sarcomas are soft tissue sarcomas that occur near the large joints of arms and legs and account approximately for 8% of all soft tissue sarcomas[4]. Head and neck synovial sarcomas are uncommon and parapharyngeal space involvement is extremely rare. There are no definite management options described but wide surgical margin resection followed by chemoradiation has been considered.

Schwannomas are benign nerve sheath tumours. Trigeminal Schwannomas are second most common intracranial Schwannomas, and can arise from the root, ganglion, or intracranial portion of the peripheral divisions of the nerve – ophthalmic (CN V1), maxillary (CN V2) and mandibular (CN V3)[5]. Preganglionic trigeminal schwannomas arising from the root are confined to the cerebellopontine angle. Ganglionic trigeminal schwannomas are the most common and confined to the Meckel's cave. Postganglionic trigeminal schwannomas are rare, and can arise from any of the peripheral divisions, commonly, the ophthalmic division. Few case reports describe peripheral trigeminal schwannomas extending extracranially into the infratemporal fossa and the parapharyngeal space. Diagnosis and treatment planning is challenging and multidisciplinary expertise is required. Surgical excision is the treatment of choice for Schwannomas with low recurrence, however, the outcome and morbidity in such cases is highly variable.

CONCLUSION

Parapharyngeal space can house a wide variety of tumours, most of them being benign in nature. Diagnosis and treatment approach can sometimes be a challenge owing to the silent nature of these tumours and the anatomical complexity of the region, which can provide with misleading radiological findings. Surgical expertise is required to successfully excise these tumours preventing injury to vital structures, which can cause high morbidity and sometimes, can even be fatal. Histopathology of the surgical specimen often provides a diagnostic surprise.

SOURCES OF SUPPORT: nil

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