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Case Report

A challenging case of right sided parapharyngeal cystic schwannoma: a case report

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ABSTRACT

The parapharyngeal space (PPS) is a potential deep space of the neck located lateral to the upper pharynx. The clinical concerns are usually due to encroachment of tumors originating from adjacent structures such as salivary glands. These tumors are considered rare entities in the available literature in otolaryngology, so we appreciate this case and would like to present it to interested readers. A 73-year-old male patient reported swelling in the right side of the neck with difficulty swallowing, choking, change of voice, and right-sided ear pain for 9 months. Clinical examination revealed a well-defined solitary oval mass of more than 6×4 cm below the right angle of the mandible just anterior to the upper part of the right Sternocleidomastoid muscle encroaching the lumen of the oropharynx behind the right posterior tonsillar pillar, displacing the right tonsil and the parapharyngeal wall forward and medially with obvious extension to the nasopharynx and hypopharynx with normal and healthy overlying mucosa. The right side of the soft palate was pushed downward and the uvula was also displaced severely downward and to the opposite side. It was soft and cystic on palpation. A CT scan of the neck was requested which revealed a cystic mass in the right PPS. Surgical excision of the cyst by external transcervical route under general anaesthesia. Histopathological examination features are consistent with Schwannoma.

Keywords: Parapharyngeal tumours, Vagus nerve Schwannoma, Neurogenic tumor, ENT, Otolaryngology, Head and neck surgery

INTRODUCTION

The parapharyngeal space (PPS) is a potential deep space of the neck located lateral to the upper pharynx. This bilateral fat-containing region is shaped as an inverted pyramid with its base attached to the skull base and the apex reaching the level of the hyoid bone, it contains several vital neurovascular and lymphatic structures. These structures are potential origins of many parapharyngeal tumors. The neighboring deep lobe of the parotid may encroach upon the anterior part of the space if enlarged and appear as a parapharyngeal mass. Primary neoplasms arising in this space are quite rare, accounting for only 0.5% of all head and neck tumors.[1] Approximately 70-80% of the tumors originating from PPS itself are benign. Clinical concerns are usually due to encroachment of tumors originating from adjacent structures such as salivary glands (parotid and minor salivary tumors).[2]

The Schwannoma is the most common neural tumor found in the PPS. The vagus nerve is reported to be the origin of 50 % of PPS Schwannoma followed by cervical sympathetic chain.[3]

CASE REPORT

A 73-year–old male patient reported swelling in the right side of the neck with difficulty of swallowing, choking, change of voice, and right-sided ear pain for 9 months.

Clinical examination revealed a well-defined solitary oval mass of more than 6×4 cm below the right angle of the mandible just anterior to the upper part of the right Sternocleidomastoid muscle encroaching the lumen of the oropharynx behind the right posterior tonsillar pillar, displacing the right tonsil and the parapharyngeal wall forward and medially with obvious extension to the nasopharynx and hypopharynx with normal and healthy overlying mucosa. The right side of soft palate is pushed downward. Uvula also showed marked displacement downward and medially to opposite site. The mass was soft and cystic on palpation and trans-illuminable. No other palpable mass was found (Figure 1). In addition, obvious wasting of the right side of the tongue and deviation to the right side (right hypoglossal nerve palsy) are noted.

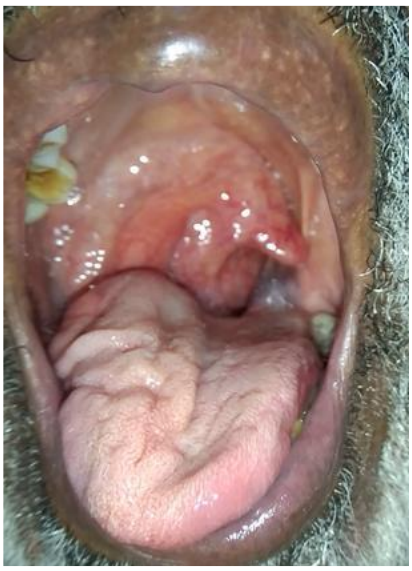

Figure 1: A mass is seen extending from the right parapharyngeal region encroaching to the oropharynx behind the right tonsil and pushing the soft palate and the uvula downward.

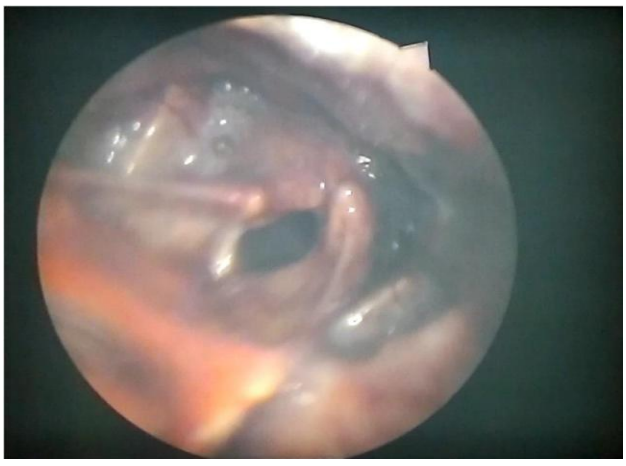

Figure 2: Indirect laryngoscopic view shows a completely paralyzed and bowing right vocal cord and pooling of saliva in the right pyriform fossa.

On working up change of voice, indirect laryngoscopy revealed complete paralysis right vocal cord (right vagus nerve paralysis) (Figure 2). The nasopharynx was visualized by rigid nasoendoscopy, which revealed a bulging from just behind the opening of right Eustachian tube with intact mucosa. The otoscopic examination showed a normal TM on both sides. Other cranial nerves (including the accessory nerve) were intact.

Investigations

The patient was sent for a CT scan with contrast which revealed a well-defined hypo-dense cyst 20×15 cm, with an enhanced capsule in the right PPS extending upward to the nasopharynx and the base of the base of the skull, and downward to the hypopharynx leaving a narrow space in the upper airway. The right carotid sheath with its contents was seen pushed antero-laterally. With no bone encroachment (Figure 3). This is followed by aspiration of 100 ml of reddish fluid which was sent for cytology study which later became inconclusive. Further imaging study with MRI was adopted. This confirmed the cystic nature of tumor and the normal great vessels around it.

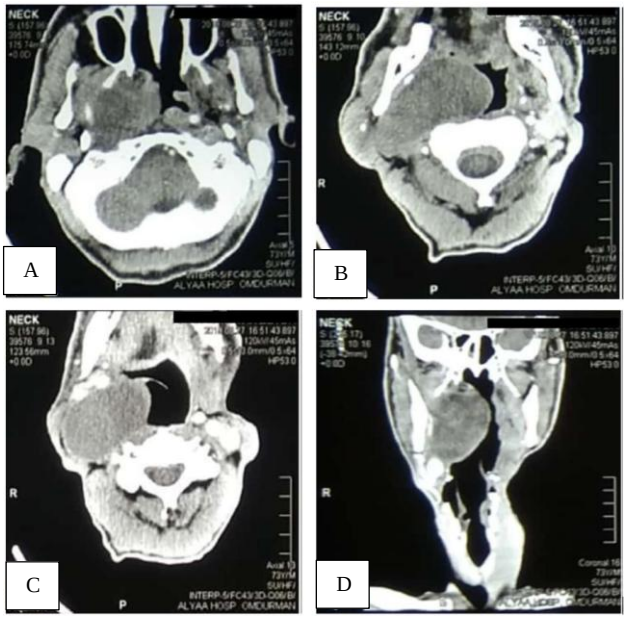

Figure 3 (A-D): Different CT cuts show the extension of the right parapharyngeal lesion from the base of the skull to the larynx.

Surgical excision of the right parapharyngeal cyst by external transcervical route under general anesthesia was performed with no intraoperative or postoperative complications.

Histopathological examination

Hematoxylin and eosin-stained slides show cystic neoplasm composed of bland wavy spindle cells arranged in hypocellular and hypercellular zones with cellular

palisading. Cells are bland with rare mitotic figures noted (Figures 4-6). The features are consistent with Schwannoma.

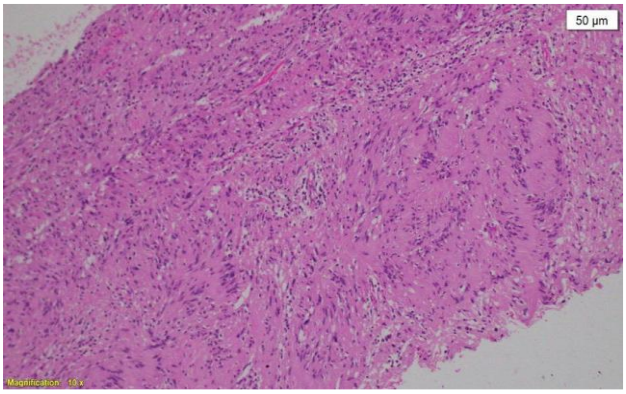

Figure 4: H and E section (10X) shows bland spindle cells forming short fascicles and palisade around the fibrillary process (Verocay bodies).

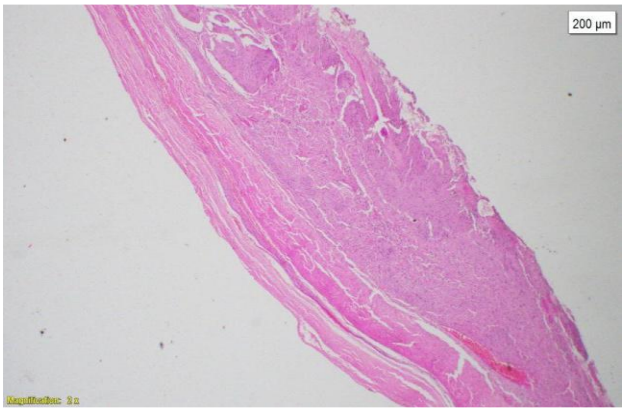

Figure 5: H and E section (X2) shows a well-defined mass with cystic degeneration of tumour confined within dense fibrous tissue.

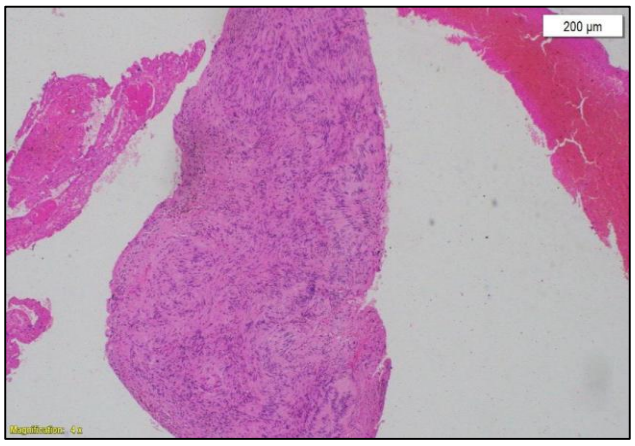

Figure 6: H and E section (X4) shows biphasic spindle cell neoplasm: compact hypercellular Antoni A areas and hypocellular Antoni B areas. Hemorrhage and fibrin deposition are also noticed.

DISCUSSION

PPS tumors account for 0.5% of all head and neck masses.[3] In a study conducted by Riffat et al around 70 different histological subtypes of lesions had been identified. Approximately 82% were benign and 18% were malignant. The most common tumors were of salivary gland origin, accounting for 45% of all lesions. The majority were benign and pleomorphic adenomas were the commonest benign lesion. Adenoid cystic carcinoma and muco-epidermoid carcinoma were the commonest reported malignant lesions.[4]

PPS tumors of neurogenic origin are the most encountered types of tumors in this space. Paragangliomas are the most common among neurogenic tumors, and schwannomas are the second most common PPS neurogenic tumors. Malignant lesions accounted for around 5% and the majority were of neurogenic origin. The major subtype was malignant peripheral nerve sheath tumor.[3]

A wide variety of miscellaneous lesions account for 12% of PPS tumors. These include internal carotid artery aneurysms, branchial cleft cysts, haemangiomas, and meningiomas. Metastatic lesions account for 3% of lesions and 2% of lesions are of lymphoid origin.[2]

Schwannomas are benign neurogenic tumors considered to originate from differentiating Schwann cells. They present in patients between 30 and 70 years of age. They usually arise from either the vagus nerve or the sympathetic trunk. The vagus nerve is reported to be the origin of 50% of parapharyngeal schwannomas and the cervical sympathetic chain is the next common source.[2]

Generally, schwannomas are characterized by slow and asymptomatic growth. However, progressive growth in the parapharyngeal region may result in pressure effects and cause dysphagia, hoarseness of voice and difficulty of breathing. The pre-operative evaluation is critical, with imaging modalities like CT and MRI determining the diagnosis and may indicate the nerve of origin. These tumors are usually fusiform along the long axis of the nerve or may be round. They are of heterogeneous signal intensity with hypointense, isointense, and hyperintense

areas on MRI. Biopsy for histopathology remains the gold diagnostic test to confirm the diagnosis.

Treatment of parapharyngeal schwannomas is challenging and may risk the vital neighboring structures. The goal is to completely remove the tumor while preserving all surrounding structures.[5]

Different surgical approaches are covered in the literature such as transcervical, transparotid, transcervical-transparotid, transmandibular, or intraoral; however, it is prudent to choose the one that best suits the size and the type of mass.[5,6]

Survival of benign parapharyngeal tumors, in general, is excellent, almost 100% when complete removal.[6]

CONCLUSION

PPS schwannomas are extremely rare and usually present with a painless, slowly growing swelling in the neck. Diagnosis and treatment of parapharyngeal tumors is not a straightforward mission. It is a challenge due to its complex anatomy, vital adjacent structures, and risky approachability. Preoperative diagnosis can be difficult and investigations such as CT and MRI can be useful for surgical planning. Complete surgical excision is the treatment of choice which can be achieved via several different surgical approaches. Recurrence is uncommon. This patient has been periodically reviewed for one year with no signs of recurrence.

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