

Vagal Schwannoma of the Parapharyngeal Space (PPS) Extending to the Skull Base: Case Report

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ABSTRACT

Introduction: Parapharyngeal space (PPS) tumour accounts for 0.5% of all head and neck tumours. PPS tumours have various origins- salivary gland tumours, schwannomas, neurofibromas and paragangliomas. They are usually asymptomatic. 50% of PPS schwannomas are originated from vagus nerve followed by sympathetic trunk.

Case report: A 64-year-old male presented to us with hoarseness of voice for last 3 months. Tongue was deviated to left on protrusion. Bilateral tonsillar enlargement was there, left more than right with absent gag reflex. Contrast enhanced computed tomography (CECT) of neck showed tumor in left carotid sheath. Magnetic resonance imaging suggested vagal schwannoma. Patient was planned for excision of left vagal nerve schwannoma by mandibular swing approach.

Discussion: Vagal schwannoma excision has risk of nerve paresis postoperatively.

Conclusion: PPS tumours present with atypical clinical manifestations and are rare. The choice of optimal procedure should be individualized based on availability, patient characteristics, and the surgeon's experience.

Keywords: Mandibular swing approach, Parapharyngeal space tumor (PPS), Vagal schwannoma.

INTRODUCTION

Parapharyngeal space (PPS) tumour accounts for 0.5% of all head and neck tumours. PPS tumours have various origins- salivary gland tumours, schwannomas, neurofibromas and paragangliomas.¹ 25-45% is the incidence of schwannoma in head and neck region. Schwannoma/ neurilemmoma are slow growing relatively uncommon benign lesion from neural sheath of any myelinated nerve. They are usually asymptomatic.² 50% of PPS schwannomas are originated from vagus nerve followed by sympathetic trunk.² Schwannomas are mostly benign, 8-13.9% can turn malignant.³ Even though PPS is surgically difficult to approach due to complex anatomy, mainstay treatment for such lesions is surgical excision.¹

CASE REPORT

History: A 64-year-old male presented to us with hoarseness of voice for last 3 months. There was no history of voice abuse, trauma, intubation, difficulty in breathing and swallowing. No history of significant weight loss or recent onset of jaundice or bone pain or coughing out of blood.

On Examination: Tongue was deviated to left on protrusion. Bilateral tonsillar enlargement was there, left more than right with absent gag reflex. No neck swelling, laryngeal framework was normal. On indirect laryngoscopy- left side vocal cord had restricted mobility both in adduction and abduction, pooling of saliva in left pyriform sinus.

Investigations: After the history, contrast enhanced computed tomography (CECT) of neck was advised which showed a solid+ cystic component of size ~ 3.4 x 3.6 x 5.1cm in left carotid sheath, widening the space between the internal and

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external carotid arteries. Following this magnetic resonance imaging (MRI) of neck with contrast was advised for better soft tissue details. It showed a left parapharyngeal space cystic mass of size $\sim 4.4\text{cm} \times 3.0\text{cm}$ at C1 and C2 vertebral level with diffuse thickening and enhancement of adjacent left carotid sheath- suggestive of vagal schwannoma (Figure 1A and 1B).

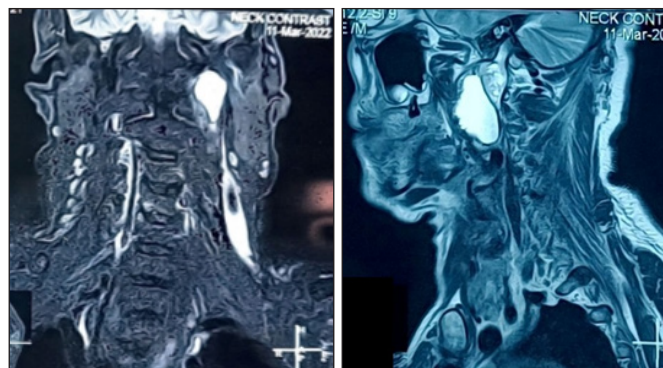


Figure 1: (A) MRI-Axial cut showing left vagal schwannoma
(B) MRI-Coronal cut-left vagal schwannoma

Management: After evaluation, patient was planned for excision of left vagal nerve schwannoma by mandibular swing approach. Lower lip splitting incision was put with extension onto left side of neck till mastoid tip with mandibulotomy. 4cm x4cm swelling was visualized at the PPS between external and internal carotid artery extending till skull base via jugular foramen. Left hypoglossal and spinal accessory nerve preserved. Swelling and involved glossopharyngeal nerve removed. Mandibulotomy repaired with 5-hole plate, 4mm screws, interdental wiring. Romovac drain kept, and wound closed in layers. Drain removed on 4th postoperative day. On histopathological evaluation of the specimen, it was diagnosed as Schwannoma (Figure 2). Postoperatively patient had no palatal movement on the left side with non-progressive hoarseness of voice. No recurrence seen on a year of follow up.

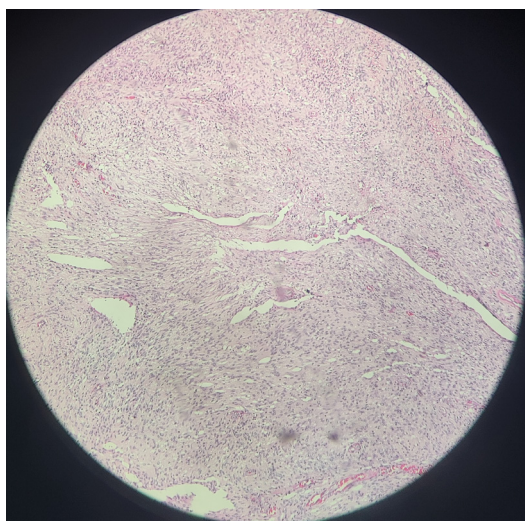


Figure 2: HPE- showing Antoni A and Antoni B areas

DISCUSSION

PPS is an inverted pyramid shaped deep potential neck space from skull base till greater cornu of hyoid. PPS is divided into 2 compartments by a fascia from the styloid process to the tensor veli palatine muscle. Anteromedial is pre-styloid compartment, containing the deep lobe of the parotid gland: retromandibular portion, a branch of CN V to the tensor veli palatine muscle, ascending pharyngeal artery, minor or ectopic salivary gland, and pharyngeal venous plexus, and fat. Posteromedial is post-styloid compartment containing- the internal carotid artery, internal jugular vein, cervical sympathetic chain, CN IX to XII, lymph nodes, and glomus bodies, and sympathetic nerves.² Incidence of vagal schwannoma is 2-3 times more than sympathetic chain schwannoma. Presentation of schwannomas vary according to the site of origin as they compress the maternal nerve fibres.⁴ If arise from sensory nerves, may present as pain, whereas if arise from the vagus nerve may present as hoarseness of voice and globus sensation of throat, others arising from the facial nerve may present as facial palsy.³ Our patient had left VC palsy resulting in hoarseness of voice.

Definitive preoperative diagnosis may be aided with radiographic studies as it is usually difficult to diagnose jugular foramen schwannomas by clinical presentation alone. Frequently done imaging is computed tomography (CT) or magnetic resonance imaging (MRI). For improved evaluation of the surrounding anatomical soft tissues, MRI is preferred.³ Contrast enhanced CT mainly shows the degree of enhancement-homogeneously solid or heterogeneous and patchy. Whereas, benefits of MRI are superior soft-tissue contrast resolution and no exposure to ionizing radiation. T2 weighted MRI, schwannomas are homogenous well-circumscribed masses exhibiting high-signal intensity, but on T1weighted MRI, a relatively low-signal intensity signal is visualized.^{4,5} Currently MRI is the imaging of choice for schwannomas of PPS.

In our case both we have done both CECT neck and MRI neck which helped us to locate the schwannoma better.

Out of all investigations the gold standard investigation is the histopathological examination (HPE) of the resected specimen. The HPE of schwannomas demonstrates two varying amounts of microscopic patterns: Antoni A and Antoni B. Tissue consisting of fascicles of spindle shaped Schwann cells is Antoni A area. These are cells arranged in palisades around central acellular, eosinophilic areas known as Verocay bodies. Antoni B area is less cellular and less organized; the spindle cells are randomly arranged within a loose, myxomatous stroma.³ Similar findings were note in HPE of our operated specimen.

Treatment of choice for this kind of schwannomas are complete tumour removal with preservation of nerve function as they are benign in nature and radio-resistant. Recurrence rates are extremely low in these cases.⁶ Postoperative neural function can be achievable as the schwannomas generally displace

the nerve they grow in but is not always guaranteed Even in cases of nerve transplantation, reversal of function should not be expected.⁷ It is evident that the neural fascicles were frequently splayed over the tumors, making preservation of the nerve of origin a difficult task for cervical schwannomas.

Various approaches are there for schwannomas or tumors of PPS. The PPS being a compact space comprising of cranial nerves and vessels; the transcervical approach provides an excellent access and/ or visualisation to the middle and inferior compartments of the PPS. On the other hand, tumors close to the deep lobe of the parotid and those involving the facial nerve are better approached through the transcervical-parotid approach. Apart from these two methods, the mandibular split approach, is applied for malignant, recurrent or large tumors involving the internal carotid artery and the skull base. For tumors involving the skull base or jugular foramen, intratemporal approach is adopted.⁸ Mandibular swing operation gives enough exposure from cervical region to skull base, with control of proximal vessels. Patient can have trismus and nerve paresis postoperatively.⁹ In our case complete excision is done using mandibular swing approach, as the tumour is extending till skull base via jugular foramen. Nerve function was not reversed.

CONCLUSION

PPS tumors have atypical clinical presentation with rare occurrence. Surgery is the choice of treatment. The approach of surgery should be based on availability, patient characteristics, and the surgeon's experience. Common complications of resection of schwannomas from head and neck region is nerve damage. Utmost importance should be taken in diagnosing of origin and extension preoperatively with proper risk explanation to the patient. Complete excision is needed to prevent recurrence.

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