

An Extensive Cervical Vagal Nerve Schwannoma: A Case Report

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SUMMARY

Vagal nerve schwannoma is extremely rare. The majority of cases present with a slow growing neck swelling without neurological deficit. Magnetic resonance imaging is the gold standard investigation to establish a pre-operative diagnosis. We report a case of a 32-year-old man with an extensive right vagal nerve schwannoma involving the right jugular foramen and parapharyngeal space. The tumour was resected via a transcervical approach. Complete excision of the tumour is the key to prevent recurrence.

KEY WORDS:

Vagal nerve schwannoma

INTRODUCTION

A schwannoma is a benign tumour of a peripheral or cranial nerve. Approximately one third of schwannomas occur in the head and neck region¹. Vagal nerve schwannoma is an extremely rare tumour. Jugular foramen schwannomas constitute approximately 2.9 to 4% of all intracranial schwannomas¹. On the other hand, parapharyngeal tumours account for 0.5% of all the head and neck neoplasms². To date, there has been no reported vagal schwannoma involving the jugular foramen and parapharyngeal space simultaneously.

CASE REPORT

A 32-year-old gentleman presented to our otorhinolaryngology clinic, complaining of painless right neck mass which was growing in size gradually for the past two years. He complained of dysphagia for six months. There was no hoarseness or other compressive symptoms.

Clinical examination revealed a 6x7cm, firm, and non-tender mass over the right neck which was extended from the angle of mandible superiorly and to the upper one third of the neck inferiorly. There was medialization of the right lateral pharyngeal wall which indicated that parapharyngeal space was involved. The cranial nerves were intact and there was no Horner's syndrome.

Fine needle aspiration cytology (FNAC) was performed and showed spindled cell lesion suggestive of schwannoma. Magnetic resonance imaging (MRI) showed right neck mass likely to be arising from the vagal nerve. The mass was extended from the right jugular foramen, along the carotid sheath and ended at C5 level. The right parapharyngeal space

was obliterated (Fig 1,2). The internal jugular vein (IJV) was compressed while internal carotid artery (ICA) and external carotid artery (ECA) was splayed.

The patient underwent an excision via a transcervical approach under general anaesthesia. There was an encapsulated yellowish mass with a cystic component. The mass extended to the base of skull superiorly, and situated medial to the right sternocleidomastoid muscle and lateral to the carotid artery. The tumour was resected along the surgical plane. However the nerve of origin was unable to be determined intra-operatively.

Histopathological examination revealed typical features of schwannoma, exhibiting spindled shaped cells with nuclear palisading (Figure 3). No nuclear atypia, necrosis or mitosis was seen.

Post-operatively the patient complained of a change of voice. Flexible nasopharyngolaryngeal scope showed right vocal cord palsy with a phonatory gap. Repeated scope was carried out on day three post-operation showed compensated right vocal cord palsy. At one year follow up, the right vocal cord palsy persisted but was well compensated by left vocal cord. There was no recurrence of tumor.

DISCUSSION

Schwannoma is a benign tumour of the peripheral or cranial nerve. It is a rare tumour and only one third of schwannoma are found in the head and neck region³. Head and neck schwannomas are more readily found arising from the eighth cranial nerve, followed by cranial nerve IX, XI, XII and the sympathetic chain. In contrast, vagal nerve schwannoma is extremely rare⁴. It has been hypothesized that Schwann cells of the ganglion area are more prone to tumour formation. The superior ganglion of the vagal nerve is located within the jugular foramen whereas the inferior ganglion is located at the extracranial orifice of the foramen. The former will end up as intracranial extension and the latter prone to extracranial extension¹. Based on this hypothesis, our patient would have the tumour originating from the inferior ganglion of the vagal nerve.

Most patients with vagal nerve schwannoma present with a slow growing painless neck mass without neurological deficit³. This finding is consistent with our patient where no clinical signs were elicited to point to the diagnosis of vagal nerve schwannoma. The presence of dysphagia in our case

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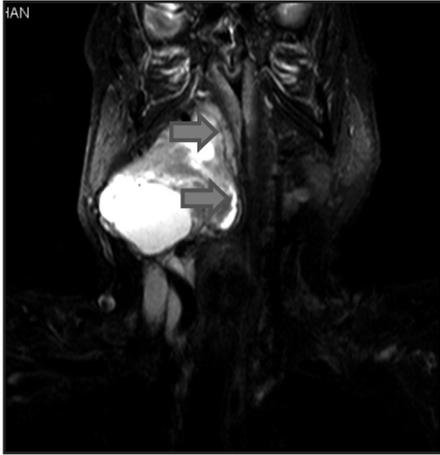


Fig. 1: Magnetic resonance imaging showed heterogenous hyperintense mass with cystic component causing narrowing of oropharynx and laryngopharynx..

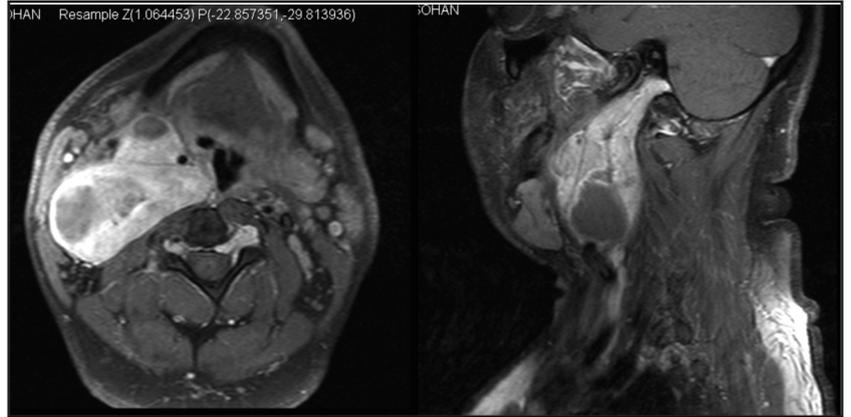


Fig. 2: Magnetic resonance imaging showed the mass obliterated the right parapharyngeal space and superiorly extended into the right jugular foramen

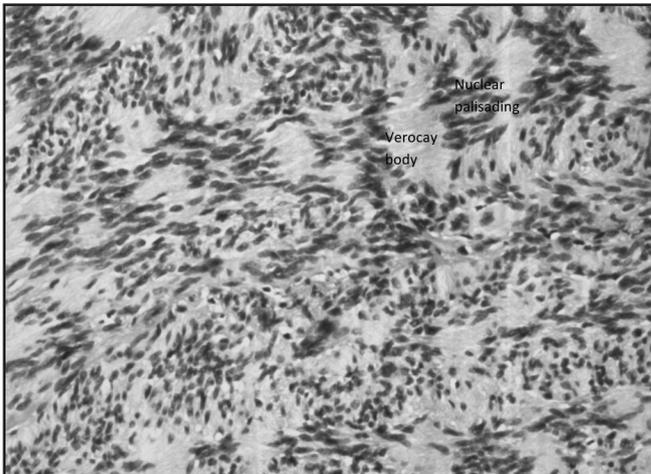


Fig. 3: Histology showed features of typical features of schwannoma, exhibiting spindled shaped cells with nuclear palisading and verocay body. Haematoxylin and eosin stain x 100.

was due to medialization of the right lateral pharyngeal wall secondary to parapharyngeal involvement. It was suggested that paroxysmal cough during FNAC or palpation of the mass was diagnostic for the vagal nerve schwannoma. However, none of these signs was present in our case. The definite diagnosis of our patient was established only after surgery in the presence of the right vocal cord palsy indicated that cranial nerve X was involved. In fact, diagnosis based on pre-operative or post-operative cranial nerve palsy was well reported as not all the nerve of origin could be identified intra-operatively¹.

The role of FNAC is less valuable in schwannoma and most would yield inconclusive result. FNAC is not recommended by some authors². However, FNAC is important to rule out

other causes of neck swelling like metastatic cervical lymphadenopathy, lymphoma or branchial cyst. It should be practiced as a routine for all the neck mass. In our case, FNAC showed features suggestive of schwannoma.

MRI is the gold standard to assess the origin and the extent of the tumour. It was suggested that vagal nerve schwannoma normally displaced the internal jugular vein and the ICA or ECA². This is the important imaging feature to diagnose vagal nerve schwannoma. However, in our case, both the ICA and ECA was splayed at the bifurcation site while IJV was compressed. This was uncommon as there was no reported case of splaying of carotid bifurcation by vagal nerve schwannoma to date. In fact, literature suggested that vagal nerve schwannoma could never separate the ICA and ECA². However, this phenomenon demonstrated in our case. On the other hand, splaying of carotid bifurcation should alert the diagnosis of carotid body tumour².

The management of vagal nerve schwannoma is complete excision. For tumour confined to the neck, few approaches can be used including transcervical, transcondylar, cervical trans-mastoid or via infra-temporal fossa approach. For tumour involving the jugular foramen, subcapsular extirpation via enlarged jugular foramen is advocated⁵. In our patient, transcervical approach was used despite of the extensive tumour involving the right jugular foramen and parapharyngeal space. The tumour was successfully resected along the surgical plane with ease despite of the extensive involvement. Complete excision including the tumour capsule is the key to prevent recurrence³. In our case, however, the patient was complicated by right vocal cord palsy post-operatively. It was reported that hoarseness was almost always present after tumour resection while vocal paralysis happened in 85% of cases³. The patient should be informed of this risk before operation.

CONCLUSION

Vagal nerve schwannoma is rare. The majority of the cases present with a slow growing neck swelling without neurological deficit. MRI is the gold standard for investigation to establish pre-operative diagnosis. Complete excision of the tumour is the key to prevent recurrence.

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