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A case of cervical Vagal Ancient Schwannoma

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Abstract **Case Report**

Schwannomas arising from cervical vagus nerve are extremely rare. Approximately 25% to 40% of all schwannomas occur in the head and neck region. Usually, they present as asymptomatic neck lumps in the 3rd to 4th decades of life. Diagnosis is mainly done with radiological imaging such as MRI. The best treatment option is to perform surgical excision of the tumor preserving the nerve function where feasible. Here we present a case of ancient schwannoma arising from cervical vagus nerve located in the parapharyngeal space, excised en-block with the vagus nerve, reconstructed with a great auricular nerve graft. Presentation, differential diagnosis, investigation, and the treatments discussed.

Keywords: Vagus nerve, Ancient schwannoma, excision.

INTRODUCTION

Schwannoma is a rare neoplasm arises from the neural sheath cells called Schwann cells and are seen commonly in peripheries [1]. Approximately 25% to 40% 0f the Schwannoma occur in the head and neck region [2]. Histologically schwannoma classified into several types like common, plexiform, cellular, epithelioid and ancient [3]. From all the above types, ancient schwannoma is the rarest form [3]. Head and neck schwannoma mostly occur in the eighth cranial nerve followed by IX, XI, XII, sympathetic chain, cervical plexus, and brachial plexus [4]. Parapharyngeal space is the commonest space involved in schwannomas [5]. In the literature schwannomas involving the vagus nerve is rare. A Slow progressive painless swelling in the neck is the commonest presentation. In rare instances, pressure symptoms like hoarseness of voice can be seen. Magnetic resonance imaging is the imaging technique of choice. Trans-cervical approach and the removal of the tumor with the preservation of the nerve trunk is the standard treatment.

Herein we present a case of ancient schwannoma arising from vagus nerve located in the parapharyngeal space surgically excised with the vagus nerve trunk.

CASE REPORT

A 29-year-old lady presented to maxillofacial and regional cleft center, teaching hospital karapitiya complaining of a painless neck lump. Two months back she noticed the neck lump while washing the face. (Figure 1) Thereafter lump progressively enlarged. She did not have any changes in the voice, dysphagia or obstructive sleep apnea. She did not have symptoms like fever, weight loss or night sweats. She was not having a remarkable medical or surgical history. On the examination, she was having a lump in the left side submandibular region measuring 3cm x 2cm size. It was firm in consistency and not tender to palpation. It was not attached to skin, muscles or bone in all dimensions. The lesion was not compressible, not fluctuant or non-pulsatile. Intraoral examination tonsillar region in the left side was slightly displaced medially. When the pressure applied to the neck lump medial movement of the tonsillar region was seen (Curtain sign). And the lesion was bimanually palpable. Rest of the physical examination was normal.

Routine blood investigations were normal. Ultrasonography was done which revealed a large anechoic lesion in the left side of the neck. Fine needle aspiration cytology was performed and it revealed a pleomorphic high-grade malignancy. Then magnetic resonance imaging (MRI) of the head and neck performed. It revealed a well-defined oval-shaped mass lesion in the left side carotid space. (figure 2,3) It was hyperintense to surrounding muscles in T2 weighted

images and iso-intense in T1 weighted images. Mass was displacing the carotid sheath contents posteriorly, medially it was protruding into nasopharynx and oropharynx. Mass was in closer proximity with the cervical vertebrae C_1 to C_4. All the tissue planes surrounding the mass was preserved. Radiological diagnosis was neurogenic tumor most likely a schwannoma. Surgery was planned, informed written consent from the patient obtained for the surgery under general anesthesia.

Surgical procedure

Surgery was done under general anesthesia with orotracheal intubation. The cervical incision was placed in the first neck skin crease two fingerbreadths below the lower border of the mandible. Flaps elevated in the subplatysmal plane. The Anterior border of the sternocleidomastoid was skeletonized and retracted posteriorly. Dissection advanced into the carotid space. Then the tumor visualized. (Figure 4) Lingual nerve and the hypoglossal nerve were overlying the mass. After meticulous dissection lingual and hypoglossal nerves were retracted superiorly. A larger portion of the mass was covered with the mandible. Then ramus osteotomy was done and mandible retracted to increase the visualization. The tumor was bluntly dissected with the fingers. Then the vagus nerve was identified entering the mass. It was difficult to shell-out the nerve from the tumor, therefore the mass was removed with the segment of the vagus nerve. (Figure 5) Then the vagus nerve repair was done with the great auricular nerve cable graft. The patient recovered uneventfully from general anesthesia. Specimens sent for histopathology. The patient was having a change in voice. Laryngoscopy revealed restricted right true cord mobility with no features of aspiration. Histopathology report revealed an ancient schwannoma.

Figure 1

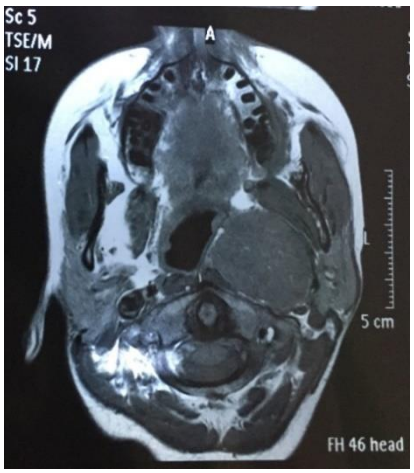

Figure 2

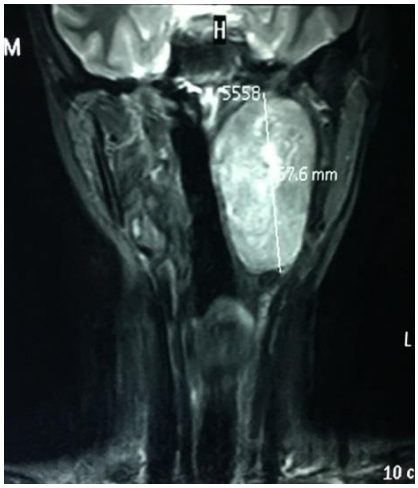

Figure 3

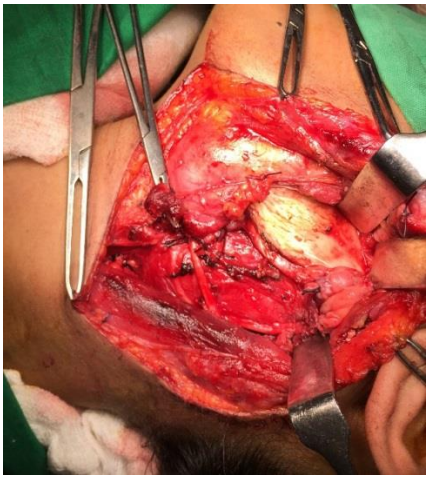

Figure 4

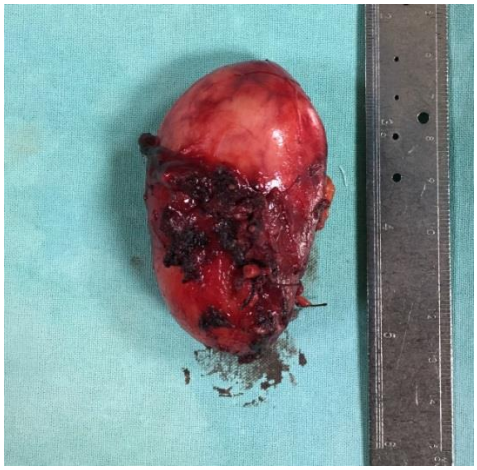

Figure 5

DISCUSSION

Schwannoma is a solitary, well encapsulated, slow-growing benign neoplasm of the arising from nerve sheath cells. They can arise from peripheral, cranial nerves or sympathetic nerves (Not from optic or olfactory nerves as they don't have a nerve sheath) [6]. Schwannoma was first described by *Verocay* in 1908. Approximately 25% to 35% of schwannomas occur in the head and neck area. The most reported region in the head and neck is the parapharyngeal space [7]. These tumors cause minimal symptoms therefore mostly diagnosed at later stages. Due to this long-standing nature of the tumor, it develops degenerative (ancient) changes like cystic changes or myxoid changes [8].

Diagnosis of these tumors is challenging due to the slow growing nature. Cervical lymphadenitis (due to inflammation, metastatic deposits, reactive), lymphoma, lipoma, cystic hygroma, carotid body tumor, laryngocele are the differential diagnoses. Inflammatory lymphadenitis present with a short history, tender to palpation and mostly respond to antibiotics. If it's tuberculosis node will be classically matted and the patient will have fever and night sweats. Lipomas are in superficial tissue planes. Cystic hygromas are non-compressible swellings in the neck with transillumination. Carotid body tumors are pulsatile masses. Sometimes schwannomas of the vagus give rise to cough on palpation which is characterized. Presenting case patient did not have a cough or any other symptoms except the firm neck swelling.

In the management radiological investigation play an important role. Computed tomography (CT), magnetic resonance imaging (MRI) are the main investigation of choice. These modalities aid in the correlation of the tumor to the important vessels and nerves. Up to date, there were no major studies to evaluate the efficacy of radiological imaging techniques in schwannoma. *Isobe at al* studied 24 cases of ancient schwannoma and presented MRI is the most useful imaging study [9]. In MRI vagal schwannomas seen as well-circumscribed ovoid masses hypointense on T1 images and hyperintense on T2 images. Generally, the nerve of origin can be anticipated with the pattern of major vessel displacement. If it arises from sympathetic trunk both carotid and internal jugular vein displace laterally. Vagal schwannomas typically push internal jugular vein laterally and carotids medially. Widening of the carotid bifurcation happens in carotid body tumors which is called the "lyre sign".

Our case-patient presented to us with firm neck lump therefore initial imaging done was ultrasonography. It revealed well defined hypoechoic mass with intervening septae. This report is non-conclusive as other non-inflammatory differential diagnoses too can give the same findings. Then an MRI was performed as the sensitivity and specificity are higher for soft tissue lesions. MRI is considered to be the gold standard in identifying schwannomas. In the MRI report, the main radiological differential diagnosis was the neurogenic tumor.

Ancient schwannomas show characteristic histopathological features. An encapsulated lesion showing degenerative cystic or myxoid areas with occasional bizarre spindle cells and even a few mitoses [8]. Histology display two patterns of areas, areas with hypercellularity (Antony type A) and areas with hypocellularity (Antony type B) [10]. These areas of hypercellularity can be misdiagnosed as malignancy in FNAC [11].

In presenting case FNA report came as a pleomorphic malignant tumor. In the literature use of the FNAC in schwannoma is questionable.

Macroscopically schwannomas of the neck can be classified into 4 categories. Type 1 nerve of origin can be identified, type 2 nerve or origin passes on the tumor, type 3 nerve fibers dilated on the surface of the tumor and in type 4 tumor surface is covered by thinned nerve fibers [12]. In types, 1 to 3 tumor can be dissected out from the nerve-preserving a functional nerve. But in type 4 it may not possible to dissect out the tumor leaving a functional nerve. In the presenting case nerve fibers could not be identified therefore vagus nerve was

sacrificed and nerve anastomosis was done at the same time.

Treatment preferred for vagal ancient schwannoma is complete surgical excision. As the tumor is well encapsulated the dissection is not difficult. The primary aim should be to completely dissect the tumor preserving the vagus nerve. In all the cases attempt must be to carefully perform intra-capsular dissection to shell out the tumor. In type 4 cases shelling out the tumor is not a possibility. According to *Zbaren et al*, there's no difference of recurrence rates in intra-capsular dissection and complete resection as an en-block [13]. An attempt was made in our case to reconstruct the nerve with greater auricular graft after the nerve resection. Post-operative results of the nerve reconstruction are uncertain. Hoarseness of voice and ipsilateral vocal cord paralysis seen in all most all the cases [14].

Our case was presented with type 4 schwannoma, therefore vagus nerve sacrificed and cable grafting is done. Postoperative hoarseness of voice was present. With the speech therapy patients speech improved in a few weeks' time.

CONCLUSION

Ancient schwannoma in the parapharyngeal space is a rare case presentation. Clinical examination combined with MRI is the best investigating method. Results of FNA may be misleading. Complete resection of the tumor is the best treatment modality up to date.

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