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Case Report

Cervical vagal nerve ganglioneuroma: a rare case report

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ABSTRACT

Ganglioneuromas occurring in the neck are uncommon. Schumacker and Lawrence (1939) stated that cervical ganglioneuroma is a rarest neck tumor. These tumors arise from cells of mantle layer of primitive neural tube and from the neural crest which migrate to a station or intermediate point in the autonomic paraganglionic nervous system. They have thus been described in neck arising elsewhere than from the cervical sympathetic chain e.g. larynx, pharynx and from nodus ganglion of vagus nerve. They may also spread or arise intraspinally. It doesn't show sex predilection. Both sexes almost equally affected. Most often present as painless, slow growing lateral neck mass. At time of presentation they present with Vocal cord paralysis. Family history may be present. Definite preoperative diagnosis may be difficult and investigations not usually helpful. We are presenting a unique case of management of left cervical vagal nerve ganglioneuroma who presented to us left side neck swelling and hoarseness of voice.

Keywords: Ganglioneuroma, Thyroplasty, Vagus

INTRODUCTION

Ganglioneuroma is a rare and benign tumor of the autonomic nerve fibers arising from the neural crest. Sympathetic ganglioneuroma themselves are fully differentiated neural tumors that don't contain immature elements. Ganglioneuroma commonly occurs in abdomen, adrenal gland, paraspinal peritoneum, posterior mediastinum. Ganglioneuroma occurring in the neck are very uncommon.

CASE REPORT

A 25 years male presented to the ENT department on 12-5-2015 complaining of gradual swelling in left lateral aspect of neck with hoarseness of voice since 4 months. The swelling was not associated with pain, difficulty in swallowing, fever, weight loss. The patient underwent FNAC at local hospital which showed granulomatous lymphadenitis for which AKT was started since 1 month but the symptoms didn't resolve. On examination there was a swelling in left lateral aspect of neck of size of 5 cm × 3 cm just below the angle of mandible as shown in Figure 1. The skin over the swelling was normal with no visible pulsation. It was firm in consistency and moved horizontally. Transillumination was negative with no bruit over swelling. Systemic examinations were within normal limits. Hopkins 70 degree scopy showed left side vocal cord palsy. Repeat FNAC was inconclusive.

CT scan from skull base to mediastinum with contrast showed well defined sharply marginated minimally enhancing low attenuated lesion in the carotid space with few areas of mild hypersdensities within, medial to the sternocleiodomastoid muscle with loss of intervening fat plane and lateral to the left carotid vessels causing medial displacement located in the region of the angle of mandible posterior to the left submandibular gland suggestive of nerve sheath tumor with left vocal cord palsy with no significant abnormality in chest as in Figure 2. The patient was operated under general anesthesia. Horizontal skin incision given on the skin

crease approximately 3 cm below the angle of mandible in left lateral aspect of neck. After retracting the skin flaps dissection continued in anterior border of SCM muscle from medial to lateral direction. Spinal accessory nerve identified and preserved. Level II lymph nodes along with peritumor lymphnodes dissected and sent for HP study. Carotid sheath was dissected. As shown in Figure 3 tumor found to be arising from the cervical vagus nerve encroaching the jugular foramen of the skull base dissected, excised and sent for histopathological study. A cut end of the vagus nerve was grafted by greater auricular nerve. Wound closed in layers.

Post-operative period was uneventful however patient had hoarseness with persistent left vocal cord palsy. Type I thyroplasty was conducted to improve voice after one month following which he developed of good quality voice.

Microscopic sections revealed a focally circumscribed tumor composed of rather bland appearing spindle cells amidst loose stroma with focal areas of myxoid change. Interspersed were several ganglion cells, with polygonal shapes, containing abundant amphophilic cytoplasm and prominent nucleoli. On immunohistochemistry, spindly cells and ganglionic cells displayed S100-P positivity. Diagnosis of ganglioneuroma was offered. Level II lymph nodes were reactive, free of tumor deposits as seen in Figure 4.

Figure1: Preoperative picture of swelling on left side of neck.

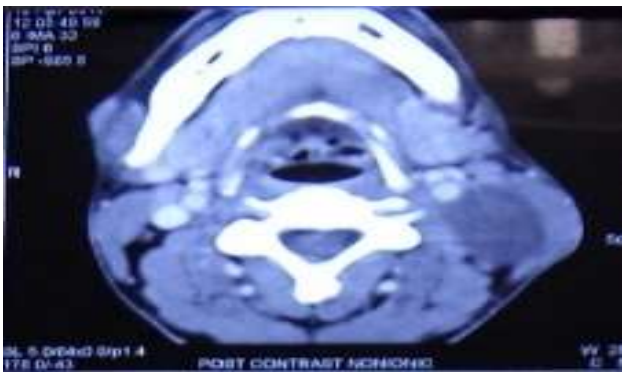

Figure 2: Axial scan with contrast showing the mass on left side displacing carotid medially.

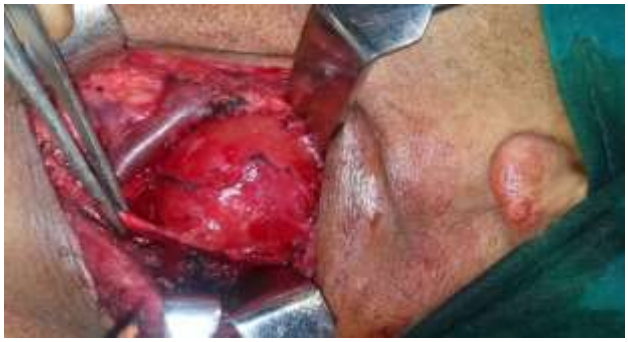

Figure 3: Ganglioneuroma at cervical vagus nerve.

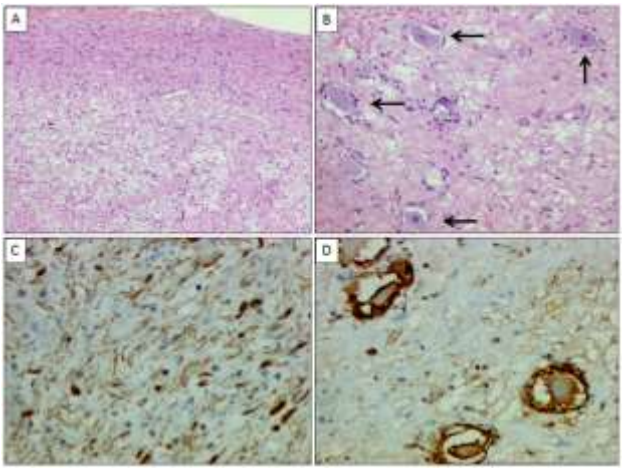

Figure 4: Histopathological features.

A. Focally circumscribed tumor containing spindly cells in a loosely fibrous to myxoid stroma. Hematoxylin and eosin (H and E) staining 200 X. B. Higher magnification showing spindly cells with several interspersed ganglion cells (arrows). H and E staining 400 X. C. Immunohistochemical staining showing S100-P positivity within spindly cells. Diaminobenzidine (DAB) staining 200 X. D. Spindly cells and large ganglion cells displaying S100-P positivity (DAB) staining 400 X.

DISCUSSION

Ganglioneuroma is a slowly growing neoplasm, usually relatively free from surrounding tissue, a factor which simplifies its surgical removal.[1,2] Pre-operative diagnosis of ganglioneuroma is difficult because many vagal ganglioneuroma do not present with neurological deficits and several differential diagnoses for tumor of the neck may be considered, including paraganglioma, branchial cleft cyst, malignant lymphoma, metastatic cervical lymphadenopathy.[3,4] Furthermore, due to their rarity, these tumors are often not even taken into consideration in the differential diagnosis. Pressure on neighbouring structures produces the deleterious effects; the removal of such a lesion from the neck thus becomes all the more imperative. The clinical diagnosis of the precise histologic character of such a tumor is difficult, but a deep lateral cervical mass, relatively fixed, but free from overlying skin and displacing the carotid artery medially, is likely to be a neurogenic tumor. There is no ready

method for distinguishing clinically between a neurinoma and a ganglioneuroma, nor can it be determined before exploration whether the origin of the tumor is from cervical sympathetic chain, vagus trunk, or from accessory or hypoglossal or other nerves in this location. It is noteworthy in the present a case where section of its cervical portion under ether anesthesia had any demonstrable effect on the cardiorespiratory function. This is in general agreement with the findings of the various authors who have previously reported resection of the cervical portion of one vagus.[5-7] Postoperative vocal cord paralysis was, of course, a constant finding in these patients.[8] Approximation by suture of the nerve was not attempted in our case as the gap was more. However we tried great auricular nerve grafting. While insufficient time has elapsed to determine the ultimate success of the suture of the divided vagus in the present instance, it seems unlikely that the cord will regain its motility. We performed type 1 thyroplasty to regain the voice of the patient after one month with improvement of quality of voice.

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Ethical approval: Not required

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