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VAGAL PARAGANGLIOMA'S: A RARE CASE REPORT.

Otorhinolaryngology

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ABSTRACT

Vagal paragangliomas are rare tumors which are difficult to differentiate from carotid body tumors and schawannomas of the vagus nerve. We present a 23-year-old male who presented with six-month history of left side neck mass extending to oro-pharynx and difficulty in swallowing. He underwent imaging studies followed by surgical excision of the tumor. Intraoperative findings showed that the tumor was located between the common carotid artery and the internal jugular vein and was arising from vagus nerve. Histopathological examination confirmed the diagnosis of vagal paraganglioma. Patient recovered fully functional without any complications.

KEYWORDS

INTRODUCTION

Cervical paragangliomas are rare tumors of the head and neck of neurovascular origin arising from neural crest cells. Within the head and neck, they are generally defined and named according to the site of origin, including carotid body tumors, jugulotympanic, moreover, vagal tumors.[1] These tumors generally exhibit a slow rate of growth and most often present as a neck mass noted clinically or radiographically. Diagnosis is usually made through a combination of clinical findings and radiographic imaging with computed tomography (CT) and magnetic resonance imaging (MRI).[1] Carotid angiography is usually unnecessary, unless the lesion requires tumor embolization.[2] Positron emission tomog- raphy (PET) scan and Octreoscan may be used to exclude multiple paragangliomas, which can occur in 10% of patients in sporadic cases and up to 40% of familial paragangliomas.[1] PET and Octreoscan are also helpful in excluding metastatic disease from suspected malignant paraganglioma. Catecholamine hypersecretion is rare in head and neck paragangliomas, occurring.

In 1 to 3% of cases.[1] when present, patients may present with symptoms similar to that of adrenal-derived pheo- chromocytomas (i.e., hypertension, paroxysmal head- aches, palpitations, and diaphoretic episodes). Benign cervical nonfunctional paragangliomas can be treated with surgery, radiation, or by observation, although definitive treatment for functional paragangliomas is complete surgical excision. We present a case of func- tional vagal paraganglioma (previously known as glomus vagale tumor) describing the diagnosis, biochemical profile, preoperative management, and surgical approach of this unique type of paraganglioma.

CASE REPORT

A 23- male presented with asymptomatic left neck mass, since 6months with difficulty in swallowing. Past h/o trauma to neck before symptoms started.

The mass was firm, non-tender, non-pulsatile , and measured 10x8 cm on left side of neck along the anterior border of strenocledo-mastoid muscle extending into oropharynx pushing left tonsil and uvula to opposite side. Examinations revealed normal larynx, and nasopharynx.

CT (computed tomography) showed a soft tissue density intensly enhancing on arterial and venous phase with necrotic areas within measuring approximately 3.3x7x8cm (AP X TR X CC) noted over prevertebral space on left side. The tumor was located between the common carotid artery and the internal jugular vein. The lower end of the tumor was elongated and continuous with vagus nerve. The common carotid artery, the internal carotid artery, and the external carotid artery were displaced anteriorly.

The presence of the tumor between the common carotid artery and the internal jugular vein suggested its derivation from the vagus nerve. In addition, the tumor was markedly hyperactive vascular. Based on these findings, a diagnosis of paraganglioma of the vagus nerve was made.

In November, tumor resection was performed under general anesthesia. The tumor was situated between the internal carotid artery and internal jugular vein. Since the tumor enveloped the vagus nerve and the nerve could not be dissected, the upper and lower sites of the vagus were cut, and the tumor was resected. No adhesion was observed between the tumor and the carotid artery or the internal jugular vein. Histopathological examination showed paraganglioma. Left vocal cord paralysis was noted post-operatively

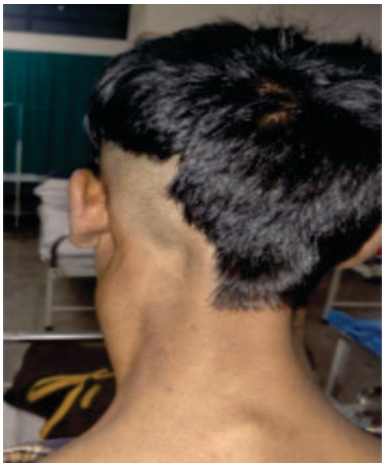

Figure 1: Swelling On Left Side Of Neck

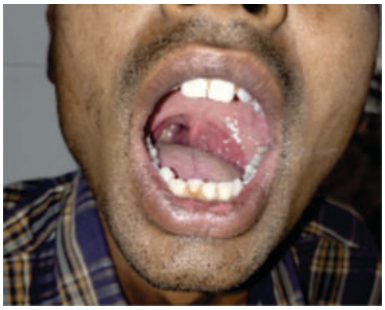

Figure 2: Oropharangeal Extension Of Swelling Pushing Uvula To

Opposite Side

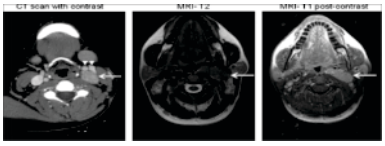

Figure 3: Computed tomography (CT) and magnetic resonance imaging (MRI) scans showing the left carotid space mass. The tumor is contrast enhancing on post contrast CT and T1-weighted MRI scans. Tumor indicated by an arrow, and anteriorly displaced internal carotid artery and internal jugular veins indicated by arrowheads. T2-weighted MRI scan shows characteristic flow voids.

DISCUSSION

Vagal paragangliomas arise from dispersed paraganglia located within or immediately adjacent to the vagus nerve and less frequent in occurrence than both carotid body tumors or glomus jugulare. There is a distinct pre-dilection for female patients, with a painless, slowly growing neck mass. The diagnosis of vagal paragangliomas is relatively easy in the presence of symptoms of vagus nerve paralysis such as vocal cord paralysis or dysphagia. However, when these symptoms are absent, it is said that the preoperative diagnosis is difficult. In particular, these tumors tend to be mistaken for carotid body tumors by angiography due to their hyper vascularity.

The important findings for accurate diagnosis are the presence of tumor at the site of vagus nerve and hyper vascularity of tumor. In our patient, the following US findings strongly suggested the disease. The first finding was that the tumor was preset between the common carotid and internal jugular vein and was continuous with vagus nerve. The second finding was hyperactive vascularity of tumor confirmed by color Doppler study.

Carotid body tumors grow between the external carotid and internal carotid artery, separating these arteries. In vagal paragangliomas, these arteries are anteriorly displaced and no separation between them is observed.

Vagal paragangliomas may arise anywhere along the course of vagus nerve and they grow between the carotid artery and the internal jugular artery, separating the carotid artery and internal jugular vein. Therefore, attention should be given to the position of the tumor in regard of the internal jugular vein.

Schawannomas of the vagus nerve may separate the carotid artery and the internal jugular vein. But vagal paragangliomas can be differentiated from schawannomas of the vagus nerve by abundant blood flow in the former. Indeed, hyperactive vascularity of the tumor was confirmed not by CT, MRI, and angiography but also by the color Doppler study. There have been no studies that suggested of the usefulness of US and Color Doppler sonography for the diagnosis of vagal paragangliomas. These methods are noninvasive and should be widely used for the diagnosis of this type of tumor.

Paragangliomas are often muticentric and familial. Once a paraganglioma is detected, family members as well as other areas of the patient should be screened. Based on the literature, angiography is recommended for this purpose.

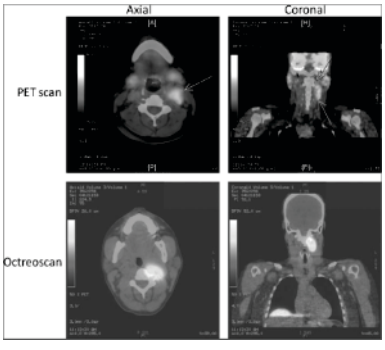

Figure 4: (A) Positron emission tomography (PET) scan and (B) Octreoscan showing the same mass, which is fludeox- glucose (18F) avid and takes up radiolabeled octreotide (axial and coronal sections). Tumor indicated by arrows.Sympathetic trunk. Although vagal paragangliomas are rare tumors (with fewer than 200 cases reported in the literature), 10 to 19% of these are malignant.[3–5] only 1 to 3% of cervical paraganglioma are hormonally active

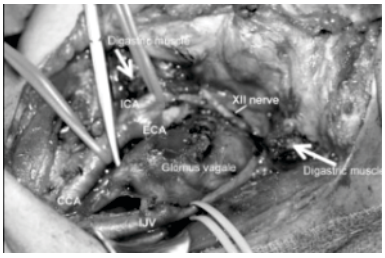

Figure 5: Surgical view showing tumor exposure after division of digastric and styloid musculature.

Carotid artery is retracted medially with red vessel loops, and internal jugular vein is retracted laterally with blue vessel loops. Cut ends of the digastric and styloid musculature are indicated by arrows with catecholamine hypersecretion.[1] Catecholamine se- cretions required for functional activity include epi- nephrine, norepinephrine, and dopamine. Functional paragangliomas (often referred to as extra-adrenal pheo- chromocytomas) lack the enzyme phenylethylamine-N-methyltransferase, which is required for the conversion

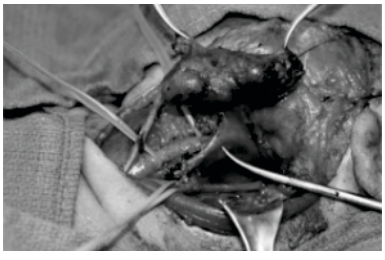

Figure 6: Surgical view showing tumor dissected comple- tely free, except for inferior attachment to vagus nerve. The clamp points to the sympathetic trunk that was preserved.

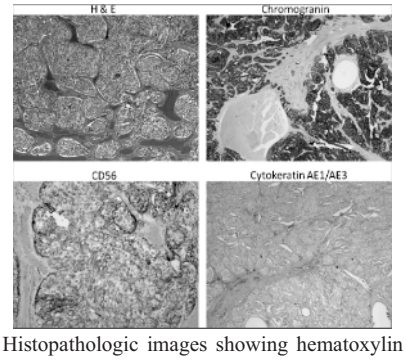

Figure 7: Histopathologic images showing hematoxylin and eosin (H&E) stain (100 × magnification) and immunohistochem- ical staining for chromogranin, cytokeratin AE1/AE3 (40 × magnification), and CD56 (200 × magnification).

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