Original Research Paper

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Internal Medicine

A NECK SWELLING THAT TURNS OUT TO BE A CAROTID BODY TUMOUR – PARAGANGLIOMA (CASE REPORT)"

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ABSTRACT This is a case report of a 41-year male having a swelling over right neck. Initially thought to be of an enlarged node probably infective in nature. However, it turns out to be paraganglioma arising from carotid body with characteristic histological and radiological appearance such as Zellballen pattern and splaying of ICA and ECA respectively.

KEYWORDS : Carotid body tumour; Paraganglioma; Parasympathetic system; Zellballen; Splaying

INTRODUCTION

Carotid body tumors and paragangliomas of the head and neck are typically painless, slow growing tumors that are often present for years prior to the patient seeking medical attention. Carotid body paragangliomas are vascular lesions, and this is reflected in their imaging appearance. Carotid body paragangliomas arise at the bifurcation of the internal and external carotid arteries and have classic radiographic features.

Case Report

A 41-year-old gentleman with hypertension has presented with complaints of swelling over right retromandibular region for 4 months. There was no history of pain. No history of discharge/ ulceration or discoloration of overlying skin. No history of loss of weight or appetite. No bladder or bowel incontinence. No dysphagia or hoarseness of voice. It was slow growing mass which is pulsatile in nature. A fine needle aspiration was performed earlier from right level 2 cervical LN; which was inconclusive showing few atypical cell clusters.

CT neck with contrast enhancement showed a well-defined oval heterogenous intensity enhancing soft tissue lesion measuring approximately 4.6 x 3.1 x 5.8 cm noted in right carotid space posterior to carotid vessels. The lesion is causing displacement and partial splaying of internal and external carotid artery anteriorly. Multiple sub-centimetric borderline enlarged cervical lymph nodes are noted. With CT imaging; possibilities of carotid body tumour, Castleman's disease and schwannoma were considered.

His basic blood workup was in normal range. Hb-12.9; TC-6.01; PLT-217; Creatinine – 0.7 Normal electrolytes and liver function tests. Thyroid function test was in normal range. MRI Neck with gadolinium contrast enhancement was suggestive of vagal schwannoma. Urine vanillyl mandelic acid was in normal range.

Surgical Management

Since the lesion was vascular, pre-operative embolization and balloon occlusion testing of RCA was planned. The procedure was performed under local anaesthesia. About 70 to 80% of the tumour was embolized using 500-micron ivalon particles. Remaining tumour supply is coming from external carotid artery directly without identifiable feeders. Right common carotid artery was occluded temporarily using 7x20mm balloon. Though right A1 segment is hypoplastic, there was good cross circulation through anterior communicating artery on left internal carotid injection. Venous phase, in both hemispheres was almost congruent (1 sec delay in early venous phase). Post-embolization, 5x4 cm tumour encasing right CCA, ECA and ICA was removed by surgery. The procedure was done under near infrared spectroscopy monitoring.

Histopathology and Immunohistochemistry

Histopathological sections show Sections show a circumscribed encapsulated tumour which is cellular and composed of cells arranged in multiple foci in a Zellballen pattern. In other areas the cells are somewhat spindled and arranged in sheets. The cells possess vesicular nuclei with foci of degenerate atypia. There is much fibrosis in the stroma. No mitotic figures: are seen. In some foci, there are areas of myxoid change and hyalinisation seen. Together histopathological features were consistent with paraganglioma. By immuno-histochemistry, cytokeratin (ae1 + ae3): positive vimentin: positive; chromogranin-a: positive; synaptophysin: positive in sustentacular cells; ki-67: less than 1%; s100: negative which is consistent with paraganglioma, right carotid body tumour.

Paraganglioma

Extra-adrenal paragangliomas are neoplasms of the paraganglia located within the paravertebral sympathetic and parasympathetic chains. It is well known that paragangliomas may be hereditary and may be part of genetic syndromes such as Von Hippel-Lindau syndrome, neurofibromatosis type I (von Recklinghausen disease), MEN 2A and MEN 2B. In the head and neck region the normal paraganglia are associated with the parasympathetic nervous system and paragangliomas arising from these parasympathetic sites account for up to 70% of extra-adrenal paragangliomas. The most common site is the carotid body. Carotid body paragangliomas arise at the bifurcation of the internal and external carotid arteries and have classic radiographic features. Carotid body paragangliomas are vascular lesions, and this is reflected in their imaging appearance. These lesions splay apart the internal (ICA) and external carotid arteries (ECA), and as it enlarges, it will encase, but not narrow the ICA and ECA. Upon contrast administration the lesions avidly enhance reflecting their vascular nature. Surgical resection is the treatment of choice, but these neoplasms are very vascular making resection challenging.

Histologically, carotid body tumors, and paragangliomas in general, have a characteristic growth pattern often referred to as a "zellballen" growth pattern. This refers to a well-developed nested or organoid growth pattern of the tumor cells with an intervening stromal component of delicate fibrovascular tissue and supporting cells or "sustentacular" cells at the periphery of the zellballen or cell nests.

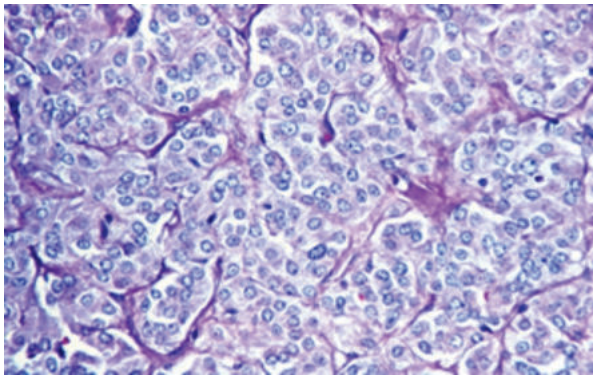

Figure 1: Representing Classical Zellballen Arrangement Of Cell Nests In Paraganglioma

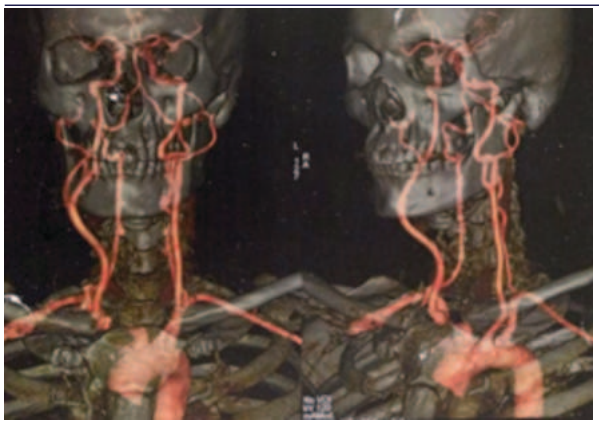

Figure 2: Characteristic Splaying Of ICA And ECA By The Carotid Body Tumour

Carotid body tumors and paragangliomas of the head and neck are typically painless, slow growing tumors that are often present for years prior to the patient seeking medical attention. They may attain large size and infiltrative growth and local recurrence may lead to death. Although it is estimated that less than 10% of paragangliomas are malignant, in some studies malignancy rates are as high as 50%. It is important to remember that all have malignant potential and it is not always possible to predict malignant behavior based on histologic features alone.

The treatment of choice for carotid body tumor paragangliomas is surgical resection. Pre-operative adrenergic blockade should be considered and due to the high vascularity of these tumors pre-operative embolization is prudent. Overall prognosis is quite good with complete surgical resection. Continued follow-up is necessary, however, as recurrence and metastasis may occur years later. It is estimated that malignant paragangliomas have less than a 50% 10-year survival rate. Surgery remains the treatment choice for these malignant tumors since chemotherapy and radiation do not appear to be of significant benefit.

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