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A Pulsatile Neck Mass of Unknown Etiology

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1. Introduction

Carotid body tumors, also known as paragangliomas or chemodectomas. are of a rare type of neoplasms and generally benign, that arise near the carotid bifurcation within glomus cells derived from the embryonic neural crest. The reported incidence of 1-2 per 100,000 [1]. The incidence of malignant tumors below 10% [2-3]. Approximately one-third are inherited as a part of a genetic syndrome. Genetic screening is advised because of the implication for malignant behavior and multifocality. The majority of these tumors are asymptomatic and initially noticed by inspection and palpation of neck swelling during the physical examination, it manifests as a pulsatile and generally painless cervical mass with firm consistency, located below the angle of the jaw, typically mobile in the lateral plane with restricted mobility in the cephalocaudal direction (Fontaine sign) or more commonly as incidental findings on radiological imaging studies. Nonetheless, the most observed symptoms are pain, dysphagia, and autonomic dysfunction in symptomatic cases [4]. If the tumors produce catecholamines, patients may complain of symptoms such as headache, hypertension, and palpitations [5].

2. Case Report

We present a case of a 77-year-old male patient who presented with a Vertebrobasilar Transient Ischemic Attack on a newly diagnosed atrial fibrillation. During the clinical examination, we noticed a pulsatile, painless lateral left neck swelling, mobile in the lateral plane with restricted mobility in the cephalocaudal direction. (Fontaine's Sign) And faint bruit was heard on auscultation.

Due to the swelling, an echography with duplex scan was performed demonstrating a solid mass of a highly vascular nature, located at the region of the carotid bifurcation (FIG. 1).

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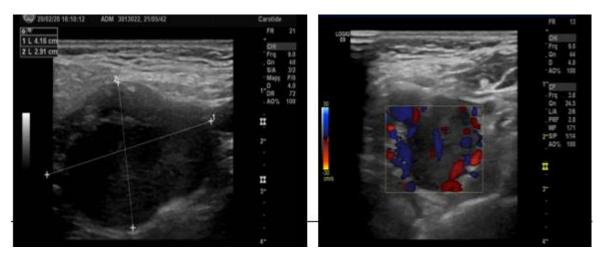


FIG. 1. Echography with duplex scan shows a well-defined mass of $4.2 \text{ cm} \times 2.5 \text{ cm} \times 3 \text{ cm}$, with a highly vascular nature located at the carotid bifurcation between the internal (ICA) and external carotid artery (ECA).

A magnetic resonance imaging angiography (FIG. 2) showed a homogenously enhancing mass at the carotid bifurcation, in keeping with the diagnosis of a carotid body tumor. On imaging, the tumor was classified as Shamblin type I. A Pre-operative hormonal check-up showed no abnormalities in catecholamines level. Because of the vascular nature of the tumor, a preoperative endovascular embolization was made. (FIG. 3). After all this pre-operative investigations, a Referral to the Otolaryngologists and the vascular surgeons for removal of the carotid body tumor was made. The patient underwent surgery under general anesthesia. The neck was explored by a vertical incision along the anterior border of Sternocleidomastoid Muscle and after careful incision, A tumor with a smooth contour and approximately 4 cm × 3 cm in diameter was observed and removed. The material removed from the surgery was forwarded to the anatomy-pathologic clinic. The histological result was compatible with benign carotid paraganglioma. The postoperative period was uneventful, and the patient was discharged with no neurological deficits.

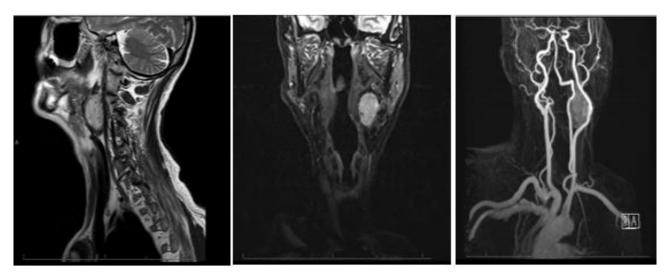
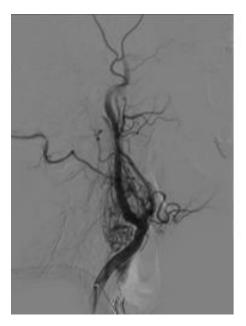


FIG. 2. Magnetic resonance imaging angiography showed a well-defined mass in the left carotid space causing splaying of the internal carotid and external carotid arteries, compatible with the diagnosis of a carotid body tumor (paraganglioma).



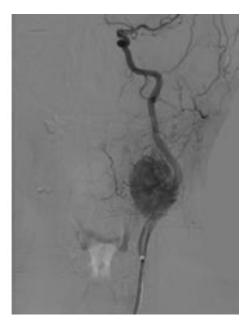


FIG. 3. Preoperative endovascular embolization of the carotid body tumor in the left carotid bifurcation.

3. Conclusion

The carotid body tumors are rare, slow growing, hypervascular neuroendocrine tumors located at the carotid bifurcation. They are mostly benign but can undergo malignant transformation. They are usually identified by clinical examination or found incidentally on imaging studies. The surgical approach is the treatment of choice due to its high anatomy-surgical complexity, it requires criterial clinical evaluation, the need to know the differential diagnosis to establish the definitive one, and the participation of several professionals from different medical specialties.

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