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Case Report

Carotid Body Paraganglioma: A Case Report

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Abstract

Paragangliomas are rare neuroendocrine, highly vascular tumors arising from extraadrenal paraganglia with the potential to secrete catecholamines. Carotid body paraganglioma is a frequent site for this kind of tumor in the head and neck. Imaging, biochemical and often genetic testing is advised for patients suspected of this disease. We review a case of a 56 years old woman presenting with Shamblin III carotid body paraganglioma. Complete surgical resection without arterial embolization or carotid artery ligation was achieved.

Keywords: Carotid Body Paraganglioma; Paraganglioma; Head and Neck Surgery

Introduction

Paragangliomas are rare neuroendocrine, highly vascular tumors arising from extraadrenal paraganglia with the potential to secrete catecholamines [1]. There can be sympathetic or parasympathetic paraganglioma depending upon the site of origin [2]. Skull base and neck paraganglioma is usually a parasympathetic, non-catecholamine secreting tumor, often arising from branches of the vagus, glossopharyngeal nerve, glomus jugulare or carotid body, being the latter the most frequent [1]. Although there is a genetic background for these disease, two thirds approximately of paragangliomas present as sporadic tumors [3]. The other third tends to be related to hereditary syndromes like von Hippel Lindau or neurofibromatosis type I [3].

The mean age at diagnosis is 47 years, with a reported rate of 2-8 cases/million.yr [2]. In the case of hereditary paraganglioma there is no gender predilection. Although with sporadic tumors women encompass 71% [4]. Usually benign in nature albeit rare cases of malignancy made evident by metastatic behavior [4]. Among the risk factors known to these pathology, there is the well described hereditary factor and also a higher prevalence among patients subject to chronic hypoxia such as those living in high altitudes or suffering from chronic obstructive pulmonary disease [5].

Case Presentation

Our patient was a 56 years old woman who presented with a painless, gradually enlarging mass on the right upper neck. She did not complained of dysphagia, hoarseness or any neurological symptoms. No episodes of "spells" were related. Upon examination a rubbery, painless, freely mobile pulsatile mass of approximately 4 cm of diameter was found. No Fontaine's sign was apparent [6]. Cranial nerves were normal at examination.

Giving the characteristics of the mass, an Angio CT was made with findings consistent with carotid body paraganglioma Shamb-

lin III (Figures 1,2). Laboratory tests ensued by the endocrinology department with measurements of urinary catecholamines, blood glucose levels as well as the pre-surgical test of coagulation and complete blood count. No high concentration of catecholamines or glucose were found.

Figure 1: CT vascular reconstruction showing a right carotid region paraganglioma with displacement of carotid vessels.

Figure 2: CT vascular reconstruction showing a right carotid region paraganglioma with displacement of carotid vessels.

The decision was made to operate on the patient. The surgery was done in the Hospital de Clínicas Dr. Mauel Quintela in Montevideo, Uruguay. A complete resection with mono cautery and bipolar cautery was achieved without carotid ligation or preoperative arterial embolization (Figures 3 - 5). The patient recovered favorably and was discharged shortly after surgery. The pathology study afterward confirmed the tumor to be a paraganglioma.

Figure 3: Neck dissection showing mass in carotid region.

Figure 4: Dissected carotid region with complete tumor resection.

Figure 5: Complete en bloc resection of carotid body paraganglioma.

Discussion

Carotid body paraganglioma most frequently presents as in this case, with a painless mass that could add neurologic, vascular or aerodigestive symptoms due to expansion. [6-7]. Other symptoms that should be taken into account are the ones related to the production of vasoactive catecholamines. Such is the case of the "spells" consisting in sporadic hypertensive crisis with sweating, headache and tachycardia [6-8].

The initial evaluation relies on imaging test with CT and MRI being the most accepted options that are able to identify, describe and classify the tumor [6]. All paragangliomas should be tested for high concentrations of catecholamines and metanephrines in blood or urine to prepare for the consequences and evaluate the risk of surgery [8]. Due to the highly vascularized nature of this tumors, a incisional or fine needle aspiration biopsy is contraindicated [10]. As for genetic testing, it is suggested in cases where there is clinical suspicion for hereditary cause as in paraganglioma syndrome [11].

There are different options in how to approach these tumors. Initial observation, surgical resection, permanent embolization, conventional radiation therapy, stereotactic radiosurgery, or a combination of those modalities could be used [6]. The Shamblin criteria classifies these tumors in three categories [12]:

- Class I tumors are localized with splaying of the carotid bifurcation but little attachment to the carotid vessels
- Class II tumors partially surround the carotid vessels
- Class III tumors intimately surround the carotids.

Class three tumors are considered to be of high surgical risk. However, resection is feasible with experienced surgical team as in this case.

Conclusion

Carotid body paragangliomas are rare tumors were surgical planning with imaging and multidisciplinary approach with endocrinologic team is needed in order to provide the patient with the best tailored treatment.

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