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Case Report

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Glomus carotideo complications - when the patient refuses treatment

Abstract

In 1941 Rufus Guild was the first using the term glomus jugular or jugular body tissue and demonstrated the 3 locations of it (half the time in the adventitia of the jugular bulb dome, and with the same frequency in the tympanic artery branch of the jugular glossopharyngeal nerve or the auricular branch of the vagus nerve). It was only much later that these tumors were reported to occur at any of these places. The carotid body tumor is also known as carotid glomus, paraganglioma or chemodectoma. Although many consider the tumor of the carotid body as a pheochromocytoma outside the adrenal gland, in 2004 the WHO classified it as a neuroendocrine tumor of extra-renal localization regardless of its secretory function. The paraganglioma is generally a benign tumor, but remains potentially locally aggressive with growth rates of 2 cm every 5 years, which may lead to symptoms by local compression. They are most commonly located in the carotid bifurcation, jugular foramen, along the vagus nerve and inside the middle ear. The tumor of the carotid body is the most common tumor of paragangliomas, with a local incidence approximately of 1: 1,700,000.[1,2]

Keywords: symptomatic glomus carotideo, refused treatment, pheochromocytoma, carotid body, tumor, carotid bifurcation, jugular foramen, adrenal gland, carotid glomus

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Introduction

Carotid glomus is a neuroendocrine tumor, originating in the mesoderm in the third branchial arch, of the ectoderm that makes up the neural crest, from which the paraganglionic cells are derived. It is rare, slow-growing, a symptomatic until it manifest as a mass in the cervical region or neck. In 1941 Rufus Guild was the first using the term glomus jugular or jugular body tissue and demonstrated the 3 locations of it (half the time in the adventitia of the jugular bulb dome,[3] and with the same frequency in the tympanic artery branch of the jugular glossopharyngeal nerve or the auricular branch of the vagus nerve). It was only much later that these tumors were reported to occur at any of these places. The carotid body tumor is known also as carotid glomus, paraganglioma or chemodectoma. The etiology can be sporadic 2/3 of the cases and 1/2 are hereditary with mutations that encode the different subunits of the succinate dehydrogenase enzyme. Although the carotid body tumor is the most common paragangliomas of the head and neck, it represents 0.6% of tumors in the neck in general. A Classification may be according the tumor location (Shamblin Class I located in the carotid bifurcation, but without adherence to the carotid vessels; Class II partially involves the carotid vessels and Class III closely adherent to the carotid), according to extension and relation to morbidity and mortality (Glasscock-Jackson and Fish - Type A, B, C1, C2, C3, D1, and D2). It is more frequent in females, with age of 30-70 years, in a proporcion of 3-6/1. The incidence is uncertain being about 1 in 1.3 million people per year.[4,5]

Case report

Female 81-year-old, Caucasian, born and resident in the city of Guarda - Portugal, diagnosed and treated for hypertension and chronic polyradiculoneurritis of the upper and lower limbs, followed in consultation with Neurology. Forwarded to the Internal Medicine department nausea, regurgitation, diarrhea, tachycardia and sweating. In Internal Medicine evaluation, she presented a mass in the left side of the neck that she never valued, mentioning appearance of the same approximately +/-5 years asymptomatic in the beginning, but the last year she began with dysphagia, odynophagia,[6] anorexia and weight lost +/-5 kg. At the physical examination, there was a mass in the left anther-lateral side of cervical region related with the carotid, with +/-4 cm painless, hard elastic, immobile, adhered to the deep planes. This mass did not present with flow or murmurs, the research of adenomegalias was negative. The patient underwent cervical CT imaging, which suggested carotid glomus confirmed by cervical MRI (ovoid formation in the carotid space, very vascularized 4* 6 cm). After the Angio-TaC of the vessels of the neck was perfomed (an ovoid mass that intensively absorbs contrast, with areas that capture less contrast and other areas of necrotic aspect, the carotid artery is displaced laterally by the mass, the jugular is surrounded by mass or collapsed). Analytically the patient had mild iron deficiency anemia (justified by anorexia),[7] the dosage of vanillandelic acid and metanephrines were negative, as was the stool occult blood test. Endoscopy and colonoscopy did not present alterations (Figure 1).[8] The patient was forwarded to the vascular surgery department, where a surgical proposal was made, but refused and she abandoned all the medical appointments (Figure 2). After 9 months, the patient was admitted to the Emergency Room with renal colic, reporting in the last 6 months frequent episodes of hypotension, profuse sweating and sometimes syncopes and dysphagia. Physical examination was emaciated, with marked asthenia, with cutaneous and hypotensive pallor (57/48 mmHg in the right arm and 67/53 mmHg in the left, heart rate 68 bpm). She was hospitalized with suspension of the antihypertensive medication, for stabilization and support of dietician, having started a pasty diet. Analytically the changes were the same as the dosage of vanillandelic acid and metanephrines, the ECG and Holter had no alterations. After the stabilization of her condition, the surgical proposal was made again and the refusal was repeated,

she kept the follow-up with Internal Medicine, Nutritionist and Psychology. At her discharge the antihypertensive medications was suspended the episodes of hypotention, syncopes remained.[9]

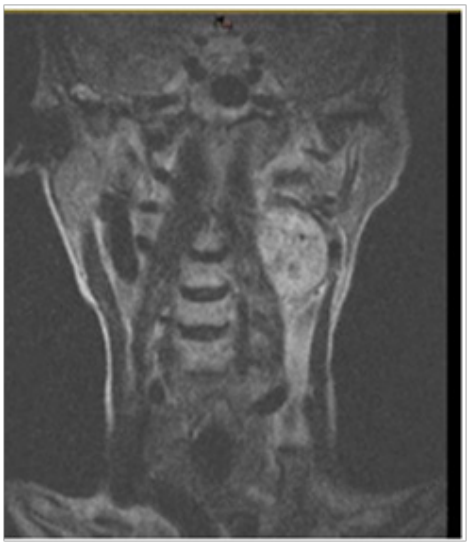

Figure 1 RMI of Neck.

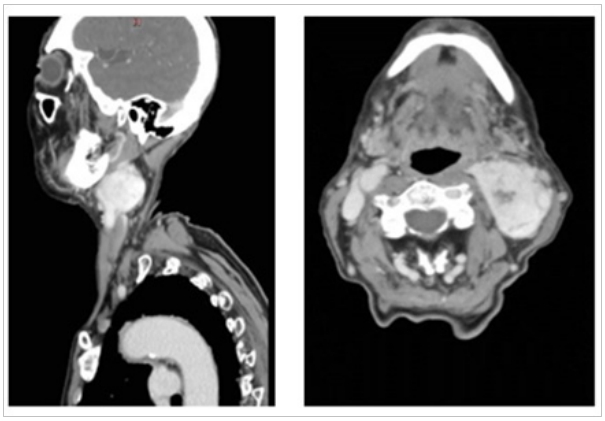

Figure 2 TAC of Neck.

Discussion

Although many consider the tumor of the carotid body as a pheochromocytoma outside the adrenal gland, in 2004 the WHO classified it as a neuroendocrine tumor of extra-renal localization regardless of its secretory function. The paraganglioma is a generally benign tumor, it remains potentially locally aggressive with growth rates of 2 cm every 5 years, which may lead to symptoms by local compression. Less than 6-15% are malignant tumors. They are most commonly located in the carotid bifurcation, jugular foramen, along the vagus nerve and inside the middle ear. Its hereditary form may present at younger ages (associated with familial paraganglioma syndrome, disease of Hippel-Lindau, multiple endocrine neoplasia type 2, neurofibromatosis type 1, Dime Carney-Stratakis) and represents 7-9% of all paragangliomas of the carotid body, can be unilateral or bilateral in 32%, seems to have equal prevalence in both genders. Sporadic is includes the hypertrophic cases that are related to exposure of low amounts of oxygen (such as patients with COPD, individuals living in high altitude areas, hypoxemic heart disease), is the most frequent form, represents 90% of cases, occurs predominantly in women, mostly with unilateral presentation and only 5% is bilateral.[10,11] The tumors can be nonfunctional, function or varying going from nonfunctioning to functioning. When functional, it may be associated with symptoms suggestive of excessive production of catecholamines, such as hypertension, flushing, headache, palpitations and diarrhea.[12,13] The growing of the carotid glomus can cause hoarseness and difficulty of swallowing by extrinsic compression and the hypersensitive carotid body syndrome.

This syndrome is a consequence of hyper stimulation of the baroreceptors, causing postural hypotension, accompanied by diaphoresis, pre-syncope or syncope. According to the literature, the standard diagnostic image is MRI, which evaluates better the vascularity and vessels that feed it, as well as the neural origin. Accuracy of the diagnosis is given by CT angiography that determines the best vessels that feed the tumor, the high risk by the Shamblin scale as well as the preoperative embolization. Doppler ultrasonography is used early in the investigation of the tumor, although it may sometimes make the diagnosis possible, it is not considered the best imaging test. The CT of the diagnosis, showing tumor hypervascularity, but does not have the same properties as NMR. Due to its vascular characteristics, BAF is not recommended considering the bleeding risk, the biopsy of the surgical specimen is preferred. The treatment of choice is surgical for definitive cure even though if the medical treatment is considered if the tumor is functional. The Shamblin scale is widely used for stratification of the surgical risk, assessment intervention as already mentioned. In small and asymptomatic paragangliomas, conservative treatment is the most indicated. Radiotherapy is used alternatively in the impossibility of resection of the tumor or there are possibilities of recurrence of the same.[14,15]

Conclusion

With this case, the authors elucidate the difficulty in managing patients with this pathology and the necessity of a multidisciplinary team. Although the tumor is rare, it is necessary to offer alternative treatments, mainly non-surgical, that improve the quality of the patients facing the complications, in our case the dysphagia that led to a malnutrition state and hypersensitive carotid body syndrome. Palliative Cares must be considered, if the complications get worst.

Acknowledgments

None.

Conflicts of interest

The author declares there is no conflict of interest.

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