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Vagal paraganglioma: A case report

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Abstract
Vagal paragangliomas (VPs) are normally benign, highly vascularized tumours that derive embryonically from the neural crest and represent a rare pathology in which surgery is usually recommended, but where experience is crucial to reduce the number of post-operative complications. In this study, we present our experience and a review of the literature.

Keywords: paraganglioma, vagal paraganglioma, vagus nerve

Introduction
Vagal paragangliomas (VP), also known as vagal glomus tumors, are characteristically slow growing benign tumors with low morbidity. It is a tumor arising of cells that have a common embryological origin in the neural crest from where they migrate beyond the sympathetic chain to acquire glandular characteristics described as paraganglia [1].
Surgery is the standard treatment, but some tumors are inoperable due to the patient's general status, or the tumour size or local extension [2]. We present a new case of benign vagal paragangliomas.

Case report
A 50 year- old –woman presented with an asymptomatic right neck mass, firm and fixed to the underlying structures in the jugulo-digastric area which lasted for three years.
There was no medial displacement of peritonsillary structures and no vocal cord paralysis.
Ultrasonography revealed a moderately vascular tumor near the carotid artery bifurcation measuring 6.0- 4.7 cm.
A Contrast-enhanced CT scan of the neck revealed a right mass 50 x 40mm at level of carotid bifurcation, with high contrast enhancement, compressing internal jugular vein and displacing anteriorly external and internal carotid arteries.
The tumor's unusual location and the inherent increased risk of postoperative vagal dysfunction were recognized preoperatively and discussed extensively with the patient.
Operative resection was performed through a standard anterolateral cervical incision along the anterior border of the sternocleidomastoid muscle.
We had found a highly vascular lesion displacing forward the internal carotid artery and backwards the jugular vein without occupying the carotid bifurcation angle. The lesion, found to arise from the right vagus nerve, was excised together with part of the nerve involve.
Three months postoperatively the patient was presenting a vagus nerve paralysis with hoarseness and without dysphagia.

Discussion
Vagal paraganglioma (VPG) or glomus vagale is a rare neoplasm representing fewer than 5% of all head and neck paragangliom [3], with a female predominance and a mean age at the time of diagnosis between 48 and 50 years, our patient fulfills both criteria.
Familiar VGP involve additional risk to multifocality and malignancy [4]. Although there are 8-20% cases of familiar VGP, it was not the case for our patient.
The glomus vagale is a slow growing tumor with few symptoms, however the clinical presentation of VP is related to the location in the parapharyngeal space.
The most frequent symptoms are a neck mass (74%), hoarseness (37%), pharyngeal fullness (26%), dysphagia (21%), pain (21%), and coughing (16%).in our case the patient presented only a right neck mass [5, 6]. According to literature 20 cases of metastatic VP has been reported. Vagal paragangliomas have a higher propensity to metastasize, with an incidence that approaches 20%. Both hematogenous and lymphatic spread are possible [7]. In our case there was no distant metastasis.
Surgery remains the treatment of choice for VP, with the risk of sacrificing the vagus nerve.
Several studies have concluded that there is no surgical advantage to performing preoperative embolization especially with the possibility of neurologic sequelae caused by cerebral artery occlusion [8], however there are other studies that recommend a preoperative embolization and claim the benefits of decreasing the tumor's vascularity and the risk of hypoglossal nerve paralysis. Our patient didn't have a preoperative embolization.
Radiotherapy is an alternative when surgery is contraindicated and in selected cases, metastatic disease, especially bilateral or unrespectable tumours [9].

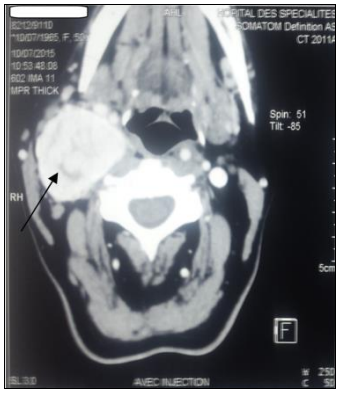

Fig 1

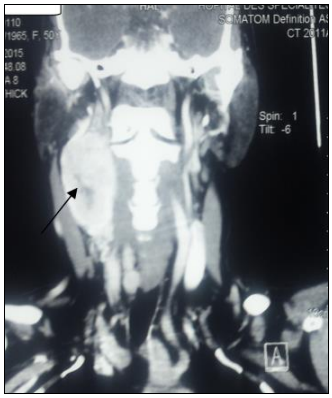

Fig 2

Fig 1 & 2: A CT scan confirmed the location and vascularity of the tumor

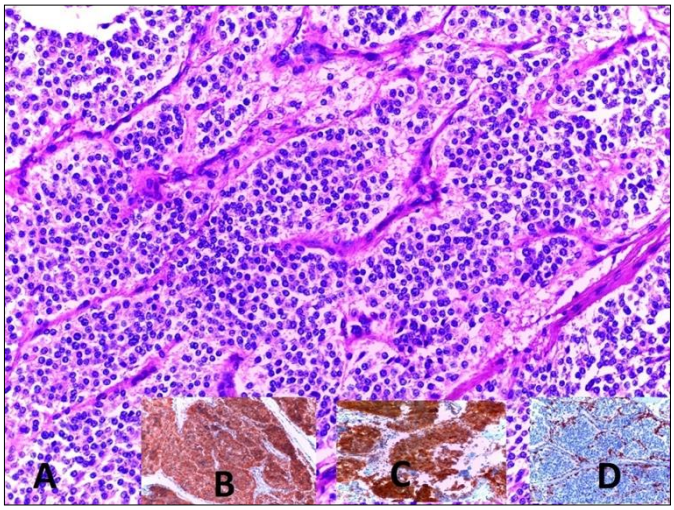

Fig 3: A) Vagal paraganglioma with fibrous septa and nerve fibers embedded in the tumor. H E G ×20, **B)** synaptophysin and, **C)** chromogranin a are positive in the tumor, **D)** S-100 protein is positive in sustenticular cells

Conclusion

The surgery is considered the treatment of choice for vagal paragangliomas, with a major risk of neurological complications.

Radiotherapy can be proposed when surgery is contraindicated.

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