Malignant paraganglioma of the vagus nerve: diagnostic and therapeutic strategies.

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INTRODUCTION: Vagal paragangliomas are rare tumors that develop in the retrostyloid compartment of the parapharyngeal space. They arise from an island of paraganglion tissue derived from the neural crest that is located on the vagus nerve. The growth of these tumors is very variable. A small percentage of these are known to have malignant potential. They arise from the carotid body, jugular bulb or vagus nerves. There is limited literature discussing the management of malignant vagal paragangliomas.

OBJECTIFS

- To report the outcomes of surgical treatment of vagal paragangliomas and to define a management protocol

CASE DETAILS :

A 64 year-old female reported to the emergency department for evaluation of a sever dysphagia. She incidentally complained of difficulty with vocal range and stamina over several months. Family history was negative for head and neck cancer. Physical exam revealed on palpation; a lateral right cervical mass. Flexible fiberoptic laryngoscopic exam revealed normal bilateral true vocal fold motion and appearance. No cervical lymphadenopathy was appreciated and cranial nerves II XII were intact.

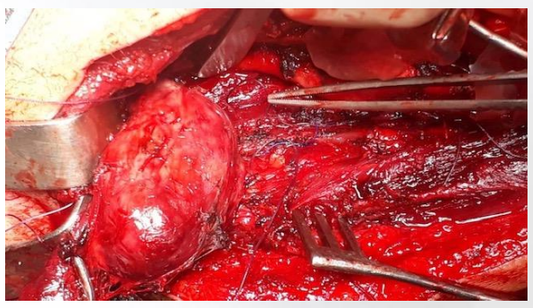

Fig. 2 exposition de la masse tumorale.

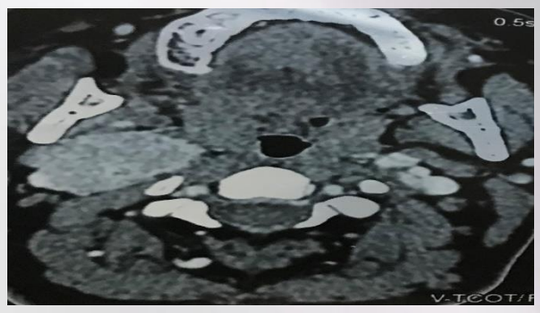

Fig. 1 CT SCAN

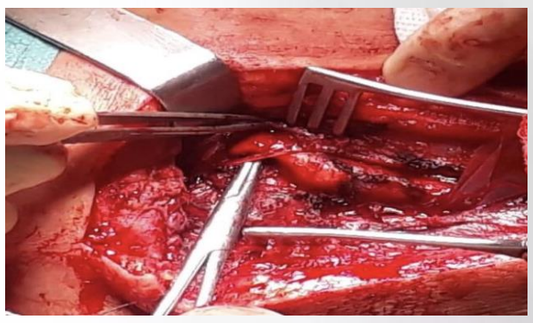

Fig. 2: resection de ma partie retro-styloidienne

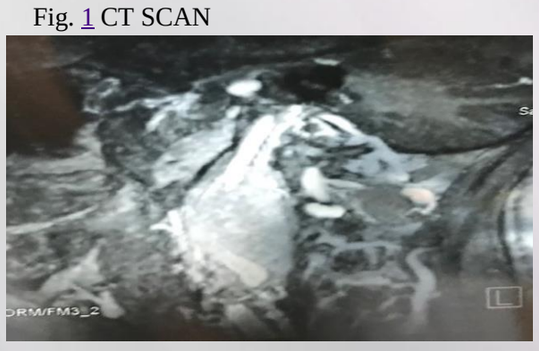

Fig. 4 MRI

Computed tomography (CT) identified a 4,2 × 40 ×2,6 cm right post-styloid parapharyngeal space avidly enhancing mass (Fig. 1). Magnetic resonance imaging (MRI) confirmed a mass within the left parapharyngeal space that was isointense on T1, slightly hyperintense on T2, and showed multiple flow voids with gadolinium enhancement. The mass was posterior to the carotid bifurcation and anterolateral to the internal jugular vein. Radiographic findings were highly suspicious for a vagal paraganglioma or nerve sheath tumor. Urine catecholamines, vanillylmandelic acid (VMA) panel and thyroid function studies were all within normal limits. Succinyl dehydrogenase (SDH) gene testing was negative. The patient was offered surgical excision or radiation therapy, and she elected to proceed with surgical excision., she developed left 10th and 12th cranial nerve deficits.

Reght selective neck dissection was not performed. And to our great surprise, the histologic reveal a malignant paraganglioma with lacal lymph metastasis.

Patient is oriented for radio-chimiotherapy.

DISCUSSION:

Paragangliomas (PGs) are rare neuroendocrine tumors derived from neural crest cells that arise from paraganglia and uncommonly show malignant behavior. They account for 0.6 % of all head and neck tumors [1]. In the head and neck, they arise from the carotid body, jugular bulb or vagus nerves. PGs typically have a benign clinical behavior; however, up to 19 % can be malignant [2]. Less than 5 % of head and neck PGs originate from the vagus nerve [3]; however, vagal paragangliomas (VP) are the most likely to be malignant [2]. These tumors present a management challenge in the head and neck due to their close relation to vital neck and skull base structures.

CT and MRI with angiography provide valuable information to aid diagnosis and preoperative planning [6]. Vagal PGs tend to cause anterior displacement of the internal and external carotid arteries, as seen in this case. When large, these may splay the carotid bifurcation, a finding more typically seen with carotid body tumors. Typically, lesions are avidly contrast enhancing on CT and MRI, reflecting their vascular nature. The classic MRI feature of flow voids gives the tumor a "salt and pepper" appearance. 123I-metaiodobenzylguanidine (123I-MIBG) scintigraphy can be used to detect occult or metastatic PGs [6].

Imaging characteristics of schwannomas include enhancement, however, not to the degree of paragangliomas. However, malignancy is classically defined by the gross or microscopic evidence of regional or distant metastatic disease [1, 3, 8].

Displacement of the common or internal carotid artery is also characteristic of these tumors. They do not demonstrate flow voids [9]. Treatment consists of surgical resection, and nerve preservation is often achieve.

Radiation therapy has been offered as a primary treatment modality, and some studies demonstrate success with local control [7]. Surgical resection may require sacrifice of CN X and poses a risk of damage to surrounding vasculature and cranial nerves [3]. It is important to discuss the clinical implications of these deficits preoperatively.

Conclusion :

In summary, PGs are slow growing tumors which typically demonstrate benign clinical behavior. Malignant PGs can only be diagnosed by demonstrating metastatic disease. Management of malignant PGs is challenging and can include surgical intervention alone or with the addition of adjuvant radiation therapy. Genetic testing is available to assess whether these patients carry the familial variant of the disease and it allows for the screening of family members.

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