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# Paragangliomas of Nasal Cavity: A Case Report

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**Abstract** **Case Report**

Paraganglioma is a rare neuroendocrine tumor. Nasal localization is very rare. Head and neck paragangliomas represent only 0.6% of head and neck tumors and 3% of all paragangliomas. The treatment is based on surgery. Radiotherapy is indicated if the surgery does not provide complete resection. We report the case of a 36-year-old patient who has had left nasal obstruction with epistaxis for 2 years. The CT scan shows a solid mass centered on the left nasal cavity, puffy, without signs of aggression, benign-looking. The patient underwent a complete endoscopic endonasal tumor removal after preoperative external carotid embolization. Pathological analysis with immunohistochemistry confirmed the diagnosis of paraganglioma. The follow-up to the treatment was favorable.

**Keywords:** Paragangliomas, Nasal cavity, Surgery, Radiotherapy, Embolization.



## INTRODUCTION

Paraganglioma is a rare neuroendocrine tumor resulting from the migration of neural crest cells associated with the autonomic nervous system [10, 14]. The sanctuary sites for these tumors are the medullary portion of the adrenal gland (pheochromocytomas), intercarotid glomus, and jugulotympanic glomus [14]. Nasopharyngeal paraganglioma is one of the rare locations of paragangliomas of the head and neck which themselves represent only 3% of all paragangliomas [3]. The aim of this work is to report a rare localization of paragangliomas.

## CASE REPORT

This is a 36-year-old patient, followed for hypothyroidism, who has presented for 2 years with progressive worsening left unilateral nasal obstruction, associated with epistaxis, clear and sometimes purulent anterior rhinorrhea. Without facial pain and without associated ophthalmological signs. Clinical examination found a deformation of the nasal pyramid, with the presence of a fleshy tumor bleeding on contact obstructing the entire left nasal cavity and exerting a mass effect on the contralateral nasal cavity, the nasal flow was abolished on the left side and very reduced on the right side (Figure 1). CT scan of the nasal cavity and cavum shows a lesional process centered on the left nasal cavity, dense, moderately enhanced and heterogeneous. This process blows the bony cortex of the jawbone downwards, the bony skeleton of the inferior turbinate, and the internal wall of the left maxillary sinus laterally. Forward, it fills the nostril. And back, it fills the choana and bombs in the nasopharyngeal lumen. With retentional sphenoidal and left maxillary sinusitis (Figure 2).

An anatomopathological study of a biopsy of the tumor shows a morphological appearance suggesting a paraganglioma of the nasal cavities or an inverse papilloma. An immunohistochemical complement was done, returning in favor of a paraganglioma.

The patient received preoperative embolization. A complete exeresis of the tumor of the left nasal cavity was performed by endonasal endoscopic approach (Figure 3). The definitive anatomopathological study concluded in a paraganglioma. The operative consequences were simple. The evolution was favorable with no signs of recurrence with a 1-year follow-up.

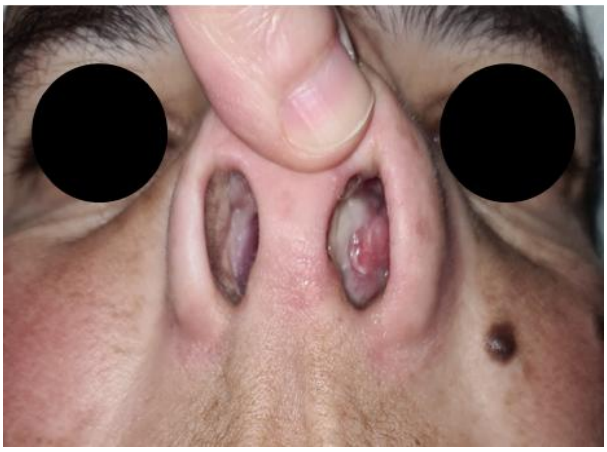

**Fig-1**

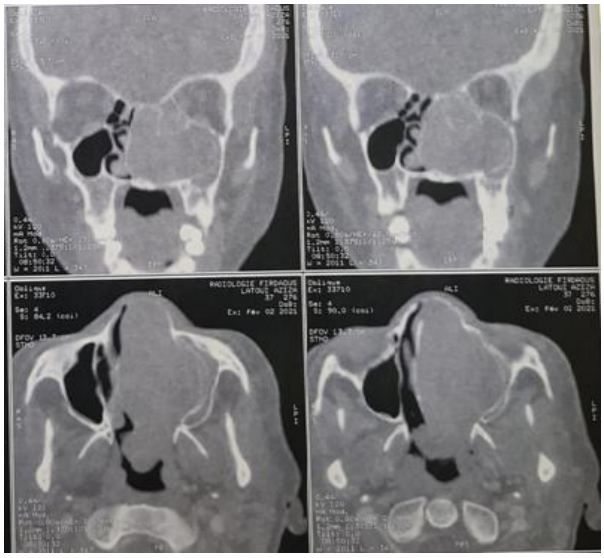

**Fig-2: Nasal sinus CT-scan (coronal and axial sections) showing a solid mass centered on the left nasal cavity, puffing, without sign of aggression, benign in appearance.**

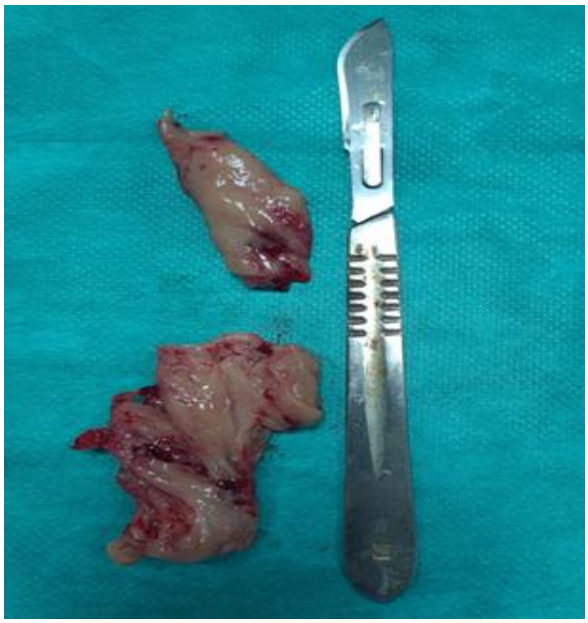

**Fig-3**

## DISCUSSION

Paragangliomas of the head and neck are very rare. They represent 0.012% of all human tumors, 0.6% of head and neck tumors, and 3% of all paraglangliomas [9, 12]. Peltier *et al*. Described a case of cavernous sinus paraganglioma in 2007 in Amiens [14]. Most of these tumors have a single location, but in 10% of cases, they are multiple [3, 9, 16]. Paragangliomas can be sporadic or familial. In 10 to 50%, they are of family origin [13]. The vast majority of reported cases are benign; malignancy is more common in sporadic tumors than in familial tumors [6, 12]. They are richly vascularized tumors, with slow growth and the possibility of infiltration of surrounding vascular, nerve and bone structures [1, 5, 7]. Paragangliomas are not painful on their own. Their clinical manifestations are diverse and depend on their topography and local extension [3].

The preoperative evaluation of these tumors is based on CT and MRI. The CT scan makes it possible to objectify the bone erosion and shows an important increase in contrast of the lesion after injection [4, 8]. On MRI, the paraganglioma appears homogeneous, isointense in T1 and hyperintense in T2 with a not very homogeneous appearance described as "salt and pepper" [3, 4, 14]. Arteriography may show a tumor "blush" or a paucivascularized tumor [14, 16]. The nasopharyngeal location of the paraganglioma can mimic other tumors that grow in the same anatomical region, and manifest with common clinical signs such as epistaxis and nasal obstruction [12]. We can cite the inflammatory nasosinus polyp, lobular capillary hemangioma, cavernous hemangioma of the nasosinus and pharyngeal region, nasopharyngeal angiofibroma. Showing the paraganglioma as "salt and pepper" on MRI may help in the diagnosis [3, 14].

The presence of somatostatin receptors in paragangliomas has recently been used in diagnostic scintigraphy. Radioactive agents such as pentetreotide have a sensitivity of up to 90%, but are less specific due to the presence of the same receptors in other neuroendocrine tumors [3].

Histologically, these are endocrine tumors with specific immunohistochemical characteristics. They express the markers of neuroendocrine tumors such as synaptophysin and chromogranin. They do not express markers for epithelial tumors [1, 12]. The majority of paragangliomas of the head and neck are mild, therefore, treated with surgery alone without radiation therapy [3, 12, 16]. In addition to the endonasal and intraoral approach, transfacial routes have also been described for the treatment of nasopharyngeal tumors [3]. Radiotherapy is only indicated in one category of patients, or postoperatively. It seems to be a useful alternative in the event of unresectable lesions, in patients with a high anesthetic risk, in patients with a paraganglioma with signs of malignancy, and finally in

addition to surgery in the event of incomplete resection [4, 12, 16, 15]. In our case the resection was complete.

The recurrence rate after complete resection is estimated in the literature to be between 1 and 4% (10, 2). Complementary radiotherapy in case of incomplete resection would give excellent results with a local control rate of up to 100% [12, 16, 15]. Embolization is indicated preoperatively for large tumors. It thus helps reduce intraoperative bleeding [12].

## CONCLUSION

Paragangliomas of the nasal cavities are rare benign tumors. Imaging can give signs of direction, but the diagnosis is based on immunohistochemistry. Surgery provides healing if the resection is complete. Radiotherapy keeps its indications if surgery is impossible or when the resection is incomplete.

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