

## Rare Carotid Bifurcation Tumor

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### Abstract

**Introduction:** Paraganglioma or chemodectoma of the carotid corpuscle or carotid glomus is a relatively rare neoplasm of slow progression, late diagnosis and uncertain prognosis. This tumor originates from the chemoreceptors of the carotid bifurcation and clinically presents as a laterocervical mass. It represents the majority of paragangliomas of the head and neck (60-70%). **Observation:** We report the observation of a 56-year-old female patient who consulted for a right laterocervical swelling without any other associated clinical signs. A complete radiological workup (Angioscanner/Angiography) will confirm the vascular nature of the lesion, characterize the size of the mass, its nature, its mapping, and its relationship to adjacent organs. Surgical excision had confirmed the diagnosis on histopathological examination. From this observation, we review the clinical, radiological and histopathological features of this tumor, as well as its uncertain evolutionary mode and the therapeutic modalities. **Conclusion:** Lateral cervical paragangliomas remain rare and poorly known affections which require an early diagnosis and a multidisciplinary management: ENT surgeon, vascular surgeon, oncologist and radiotherapist according to the evolutive forms.

**Keywords:** Paraganglioma - Chemodectoma - Carotid - Surgical treatment.

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## INTRODUCTION

Paragangliomas (PG) are rare tumors that develop at the expense of cells from the diffuse neuroendocrine system [1-3]. 85-95% of cases are benign tumors [4, 5]. These cells have a common embryological origin at the level of the neural crest from where they migrate beyond the sympathetic chain to acquire glandular characteristics taking the term paraganglion.

Many synonyms were used to describe these tumors: chemodectoma, non-chromaffin paraganglioma and glomus tumor [1]. Currently, only the term paraganglioma is validated by all histologists [2].

The difficulty of management is related to the secretory potential, multi-locality and malignancy of these tumors [7, 8]. We report an observation of a carotid paraganglioma discovered in a 56-year-old female patient

## OBSERVATION

Mrs T.B., 56 years old, native and resident in Marrakech, admitted in February 2019 for a right laterocervical mass, with no particular pathological history, and no similar case in the family. The onset of the symptomatology dates back to 12 years before her admission, by the installation of an isolated painless right laterocervical swelling; gradually increasing in volume without other associated signs. The patient initially consulted a general practitioner where an ultrasound coupled to the doppler of the cervical region was requested, which objectified a right latero-cervical tissue mass at the sub-angulo-mandibular level, heterogeneous, hypoechoic, well limited, well vascularized, located at the level of the carotid bifurcation of 5cm/6cm of great axis and associated with bilateral jugulo-carotid and sub-maxillary ganglions, of sub-centimetric size; then referred to the ENT department where she was hospitalized for a biopsy. Given the hemorrhagic nature of the mass, the surgeon performed hemostasis with stitches without

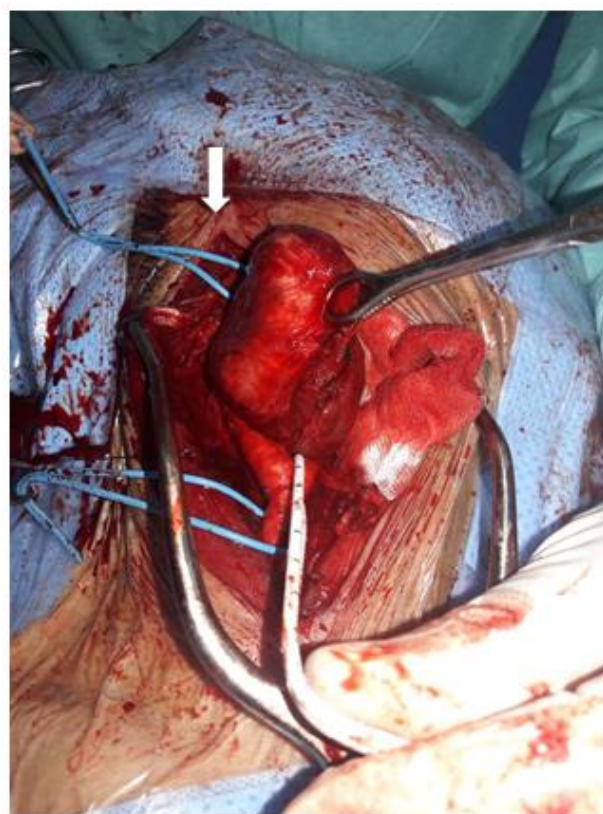
being able to perform this biopsy. She was then referred to us. The clinical examination showed a pulsatile right laterocervical mass that was mobile in relation to the deep plane plus a 2 cm incision with inflammatory signs around the incision and a hematoma (the location of the biopsy). The rest of the clinical examination was unremarkable. An angioscan was performed, which showed a right laterocervical tissue mass located at the level of the carotid bifurcation between the external carotid artery and the right SCM muscle outwardly and pushing the internal carotid artery inwardly, which is sheathed in the form of a sleeve, and the internal jugular vein outwardly with a separating fatty border, This formation is well limited with a roughly oval shape, clearly and homogeneously enhanced on arterial time, receiving several tortuous arterioles, measuring approximately 3.6 cm/4 cm in length and 5.5 cm in height, raising the suspicion of a paraganglioma classified as Shambien stage III (Figure 1). Additional arteriography showed a mass in the carotid bifurcation, well vascularized by the branches of the external carotid artery, suggesting a right carotid paraganglioma (Figure 2). The ENT examination (looking for tympanic localization) was normal. The dosage of methoxylated derivatives of plasma catecholamines was normal (Normetanephrine: 37ng/l; Metanephrine: 32ng/l), an abdominal ultrasound targeted on the adrenal loges was not performed. The patient underwent surgical treatment, which consisted of a total resection of the tumor through a longitudinal pre-sternocleidomastoid incision without sacrifice of the carotid axes (Figure 3,4). A transfusion of 2 CG of was done given the intraoperative bleeding. Postoperative follow-up was marked by mild dysphonia regressing for three months. Anatomopathological examination of the surgical specimen confirmed the diagnosis of carotid paraganglioma, the volume of which was 81 cm<sup>3</sup> or 6cm / 4.5cm / 3 cm (L/I/H) (Figure 5). The follow-up ultrasound was normal. The follow-up CT scans at 1 and 2 years were without abnormalities, including no tumor invasion.



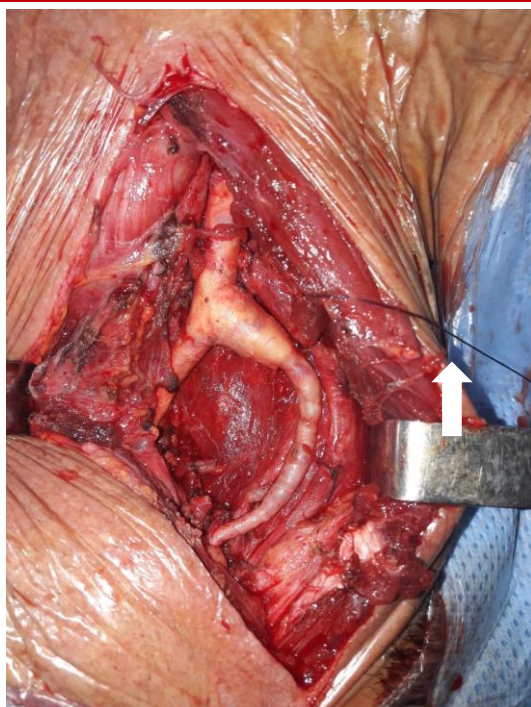
**Fig-1: Angioscan of the cervical arteries showing a hypervascularized lesion process in the right jugulocarotid region (arrow), centered by the carotid bifurcation**



**Fig-2: Arteriography shows a mass at the level of the right carotid bifurcation with vascular blush (arrow)**



**Fig-3: Intraoperative view of the carotid paraganglioma (arrow) after dissection of the primary, external and internal carotid arteries and before complete resection of the tumor mass**



**Fig-4: Intraoperative view after resection of the tumor (paraganglioma) with respect to the carotid vessels (arrow)**



**Fig-5: Macroscopic appearance of the carotid paraganglioma with a volume of 81 cm<sup>3</sup>.**

## DISCUSSION

Paragangliomas or chemodectomas are rare tumors that represent 0.03% of all neoplasia in the human body [9]. The largest series is the Mayo Clinic series by Hallet *et al.* published in 1988: 153 PGs over a 50-year period (1935 - 1985) [10]. Among head and neck PGs, carotid PGs are the most frequent (60 to 70%) [11].

These endocrine tumors arise from the paraganglionic system consisting of neuroepithelial cells derived from the neural crest and possessing

catecholamine neurosecretory granules in their cytoplasm. These cells may be located outside the adrenal medulla in structures belonging to the para- and orthosympathetic system [9]. Thus, they can be occasionally multifocal (2-15% of cases) and associated with multiple endocrine neoplasia (NEM I and NEM II) [12]. Only 1-3% of PGCs have been reported to be secretory [13]. For many authors, urinary and blood catecholamines and their metabolites should be systematically measured preoperatively, especially if the patient presents symptoms suggestive of possible tumor activity: excessive sweating, palpitation, tachycardia, hot flashes, headache, pallor, tremor, nausea or hypertension.

The predominance of women is found in most series. The female to male sex ratio in carotid PGs is 8.3:1 in the series by Rodriguez [14], 1.9:1 in the series by Patetsios [15]. Carotid paragangliomas are often observed in women between 40 and 78 years of age with a mean age of 52 years [16].

The first reason for consultation is the appearance of a painless, palpable, latero-cervical, sub-angulo-mandibular tumefaction, of progressive appearance, not very mobile in the vertical direction but mobile laterally as in the case of our patient (Fontaine's sign), not following the movements of swallowing.

The appearance of a complication may be inaugural: signs of compression (dysphonia,odynophagia...) or signs of catecholamine secretion (fluctuating arterial hypertension, palpitations, sleep apnea...). Such a symptomatology is not very specific and the diagnosis of paraganglioma is generally not the first hypothesis evoked. We most often think of a cervical adenopathy, a carotid aneurysm, a swelling of the salivary glands, a tonsillar hypertrophy, more rarely congenital laterocervical cysts or neurinomas.

The different imaging means allow a non-invasive diagnostic approach with high sensitivity and specificity, so that their information facilitates the therapeutic decision. Imaging is necessary for the diagnosis and also allows for an extension assessment. It will specify the exact location of the tumor, its lateral extension (towards the parapharyngeal space) but especially upwards, in relation to the base of the skull, the repression and/or invasion of adjacent structures (Vx carotid and jugular), the presence of other associated paragangliomas, the presence of adenopathies and distant metastases. The most commonly used examinations are Angioscanner with 3D reconstruction and MRI Angiography which reveal the PG as a hypervascularized tumor. Arteriography is the most efficient examination for diagnosis, it remains the "Gold standard" of morphological explorations. It allows to determine the vascular nature of the tumor and possibly to perform a preoperative embolization with the theoretical aim of decreasing postoperative

bleeding, it gives a hypervascularized tumor image with BLUSH effect [17]. Ultrasound coupled with Doppler remains a clearing examination that shows the hypervascularization of these tumors and restricts the differential diagnosis to other solid tumors such as metastatic adenopathies of thyroid and kidney cancers. The Shamblin classification (carotid PG) is specified at the end of this imaging workup (Table).

Macroscopically, the tumor presents as a well-limited, sometimes lobulated nodule. It is elastic in consistency and reddish-brown in color with hemorrhagic areas. Light microscopy shows round cell nests of variable shape and size surrounded by a richly vascularized and more or less fibrous stroma. The main cells are rounded or polygonal "of epithelioid type". Their cytoplasm is quite abundant, eosinophilic and finely granular. The nucleus is central, often regular. The supratentacular cells, corresponding to modified Schwann cells or satellite cells of the autonomic nervous system, are difficult to identify apart from immunostaining with the anti-protein S100 antibody. Besides this classical histological form of relatively easy diagnosis, other morphological variants are much more difficult to recognize, such as the solid form, the pseudoglandular form or the sclerosing form with hyalinized stroma. Local invasion of adjacent anatomical structures is a criterion of local aggressiveness. The immunohistochemical study confirms the neuroendocrine nature of this tumor [6].

Finally, patients should have a urinary and blood metanephrine assay and a blood pressure assessment [18]. The role of preoperative embolization remains controversial. For some, it would reduce the hemorrhagic character of the tumor and facilitate its resection, the operation having taken place at the latest 48 h after embolization [19]. Carotid stents were initially used as alternatives to embolization in order to exclude tumor vascularization. Currently, they are used in carotid PG surgery with the aim of devascularizing the tumor without interrupting arterial flow and facilitating tumor resection. Published results are in limited series of patients with advanced tumors or patients who have already undergone carotid sacrifice [20].

The approach must be sufficient to allow control of the vascular-nervous elements. At a minimum, it is a conventional cervicotomy approach. The classic incision is most often oblique along the anterior border of the sterno-cleido-mastoid along a line from the tip of the process behind the earlobe to the medial tip of the clavicle, which can be enlarged into a cervico-parotid or even cervico-parotidomastoid approach. Lymphadectomy is not systematic, but in some cases, lymph node involvement is demonstrated [21].

The procedure is performed under general anesthesia with endo-tracheal intubation. The anesthetic risk is increased if the lesion secretes catecholamines. The patient is installed in dorsal decubitus position, a slight cervical extension is obtained by sliding a medium-sized log under the shoulders. The operating field must include the thigh in order to harvest a saphenous vein if carotid repair is necessary.

Radiation therapy may be recommended preoperatively for large tumors or to treat metastases and tumor recurrence. Its objective is to stop tumor growth with clinical and radiological stabilization. The evolution is usually benign and slow.

## CONCLUSION

Latero-cervical paragangliomas remain rare and poorly understood conditions that require early diagnosis and multidisciplinary management: ENT surgeon, vascular surgeon, oncologist and radiotherapist depending on the evolving forms.

Carotid bifurcation chemodectoma is the most frequent paraganglioma of the head and neck (60 to 70% of casent). It is most often a benign solitary tumor with a slow evolution. The diagnosis is made on the basis of clinical and angiographic findings and confirmed by histopathological examination and immunohistochemical studies. Only the appearance of metastases confirms the malignancy of the tumor in the absence of clear histological criteria of malignancy.

The gold standard treatment for cervical PG is surgery. Complications related to surgery, particularly nerve damage, are not negligible. Radiation therapy is an alternative in cases where surgery is contraindicated and in selected cases, notably bilateral and/or non-removable forms.

## REFERENCES

1. Del Guercio, L., Narese, D., Ferrara, D., Butrico, L., Padricelli, A., & Porcellini, M. (2013). Carotid and vagal body paragangliomas. *Translational Medicine@ UniSa*, 6, 11.
2. Peric, B., Marinsek, Z. P., Skrbinc, B., Music, M., Zagar, I., & Hocevar, M. (2014). A patient with a painless neck tumour revealed as a carotid paraganglioma: a case report. *World journal of surgical oncology*, 12(1), 1-5.
3. Baysal, B. E., Ferrell, R. E., Willett-Brozick, J. E., Lawrence, E. C., Myssiorek, D., Bosch, A., ... & Devlin, B. (2000). Mutations in SDHD, a mitochondrial complex II gene, in hereditary paraganglioma. *Science*, 287(5454), 848-851.
4. Patetsios, P., Gable, D. R., Garrett, W. V., Lamont, J. P., Kuhn, J. A., Shutze, W. P., ... & Thompson, J. E. (2002). Management of carotid body paragangliomas and review of a 30-year

- experience. *Annals of vascular surgery*, 16(3), 331-338.
5. Maves, M.D. (1993). Vascular tumours of the head and neck. In: Bailey BJ, Johnson JT, Kohut RI, Pillsbury HC, editors. Head and neck otolaryngology. Tardy medical management. Philadelphia: JB Lippincott, 1397-409
  6. Bougrine, F., Maamouri, F., Doghri, R., Msakni, I., Znaidi, N. S., Chouchane, O., & Bouzaine, A. (2008). Une tumeur rare du glomus carotidien. *Journal des maladies vasculaires*, 33(4-5), 214-217.
  7. Van Den Berg, R. (2005). Imaging and management of head and neck paragangliomas. *European radiology*, 15(7), 1310-1318.
  8. Baillet, F., Delanian, S., Rozec, C., Housset, M., Lamas, G., & Poignonec, S. (1990, January). The value of radiotherapy in the treatment of tympano-jugulare chemodectoma. Apropos of 18 cases. In *Annales D'oto-laryngologie et de Chirurgie Cervico Faciale: Bulletin de la Societe D'oto-laryngologie des Hopitaux de Paris* (Vol. 107, No. 5, pp. 315-318).
  9. Kliewer, K. E., Wen, D. R., Cancilla, P. A., & Cochran, A. J. (1989). Paragangliomas: assessment of prognosis by histologic, immunohistochemical, and ultrastructural techniques. *Human pathology*, 20(1), 29-39.
  10. Hallett Jr, J. W., Nora, J. D., Hollier, L. H., Cherry Jr, K. J., & Pairolero, P. C. (1988). Trends in neurovascular complications of surgical management for carotid body and cervical paraganglionmas: A fifty-year experience with 153 tumors. *Journal of vascular surgery*, 7(2), 284-291.
  11. Sobol, S. M., & Dailey, J. C. (1990). Familial multiple cervical paragangliomas: report of a kindred and review of the literature. *Otolaryngology—Head and Neck Surgery*, 102(4), 382-390.
  12. Pellitteri, P. K., Rinaldo, A., Myssiorek, D., Jackson, C. G., Bradley, P. J., Devaney, K. O., ... & Ferlito, A. (2004). Paragangliomas of the head and neck. *Oral oncology*, 40(6), 563-575.
  13. Snitzer, J. L., Sheeler, L. R., Bravo, E. L., Thacker, H. L., & Lavertu, P. (1995). A carotid body and glomus jugulare paraganglioma secreting norepinephrine. *Endocrine Practice*, 1(2), 82-86.
  14. Rodríguez-Cuevas, S., López-Garza, J., & Labastida-Almendaro, S. (1998). Carotid body tumors in inhabitants of altitudes higher than 2000 meters above sea level. *Head & Neck: Journal for the Sciences and Specialties of the Head and Neck*, 20(5), 374-378.
  15. Patetsios, P., Gable, D. R., Garrett, W. V., Lamont, J. P., Kuhn, J. A., Shutze, W. P., ... & Thompson, J. E. (2002). Management of carotid body paragangliomas and review of a 30-year experience. *Annals of vascular surgery*, 16(3), 331-338.
  16. Luna-Ortiz, K., Rascon-Ortiz, M., Villavicencio-Valencia, V., Granados-Garcia, M., & Herrera-Gomez, A. (2005). Carotid body tumors: review of a 20-year experience. *Oral oncology*, 41(1), 56-61.
  17. Rodríguez, A. M. C., Delgado, G. H., Caballo, M. R. B., & Morte, S. G. (2007). Head and neck paragangliomas: imaging diagnosis and embolization. *Acta Otorrinolaringologica (English Edition)*, 58(3), 83-93.
  18. Martin, T. P., Irving, R. M., & Maher, E. R. (2007). The genetics of paragangliomas: à review. *Clin Otolaryngol*, 32(1), 7-11.
  19. LaMuraglia, G. M., Fabian, R. L., Brewster, D. C., Pile-Spellman, J., Darling, R. C., Cambria, R. P., & Abbott, W. M. (1992). The current surgical management of carotid body paragangliomas. *Journal of vascular surgery*, 15(6), 1038-1045.
  20. Scanlon, J. M., Lustgarten, J. J., Karr, S. B., & Cahan, J. I. (2008). Successful devascularization of carotid body tumors by covered stent placement in the external carotid artery. *Journal of vascular surgery*, 48(5), 1322-1324.
  21. Makeieff, M., Raingeard, I., Alric, P., Bonafe, A., Guerrier, B., & Marty-Ane, C. (2008). Surgical management of carotid body tumors. *Annals of surgical oncology*, 15(8), 2180-2186.