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Case Report
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Management of a Case of Carotid Paraganglioma in an Under Medicalized Context: Review of the Literature

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Abstract

Introduction: Paragangliomas, also called glomus tumors or chemodectomas, are neuroendocrine tumors that arise from glomus cells in paraganglia. They are rare tumors, accounting for about 0.6% of head and neck tumors and about 0.03% of all tumors. We want, through a clinical case, to highlight the clinical and diagnostic aspects, the therapeutic difficulties and to review the literature.

Case Report: A 38-year-old woman, admitted to the department for a right submandibular tumefaction progressively evolving for four (4) years without any notion of dysphagia, dyspnea, dysphonia, or peripheral facial paralysis, nor rhinology. He had no notion of fever or associated pain. She had no surgical medical history. The physical examination found a subangular mandibular tumefaction about 5 cm in diameter on its long axis, of regular outline, not painful on palpation, pulsatile, the overlying skin looked normal. Examination of the oral cavity found a tumor developed behind the right posterior pillar which pushed the amygdala forwards and inwards causing a slight deviation of the uvula towards the contralateral side. Cervical CT showed a right laterocervical hypervascularized mass. The tumor was classified as Shamblin II. Surgical excision without embolization was performed under general anesthesia and local healing of the wound was obtained after 10 postoperative days. Histological examination revealed a carotid paraganglioma. No recurrence was noted with a follow-up of 12 months.

Conclusion: Early diagnosis and surgery are guarantees of a good prognosis.

Keywords: Paraganglioma; Carotid; Surgery; Embolization

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Introduction

Paragangliomas are the most common Carotid Body Tumors (CBT) of the head and neck, with an approximate incidence of less than 1 case in 30,000 people, particularly affecting women with an average age of 43 years [1]. These are rare neuroendocrine tumors that develop at the expense of the carotid body at its bifurcation [2]. They exhibit extensive growth and can compromise and enclose vital neurovascular structures in the neck, such as the external and internal carotid artery, making surgical resection extremely difficult [3]. Although his tumors are neuroendocrine, they rarely produce catecholamines and most of these tumors are benign [4]. In the literature, there is little data on the subject in Black Africa, however Tall et al. [5], published in 2005 a series of 8 cases, 2 of which were of carotid location. The management of carotid paragangliomas recognizes several issues:

- Isolated mass discovered late and asymptomatic.
- Lack of familiarity with its management due to its rarity.
- A subsequent lack of embolization makes management difficult at the advanced stage.

The objective is to report our experience in the management of a case of carotid body tumor and to make a comparative analysis with the literature.

Case Presentation

It was a 38-year-old woman, a housewife admitted to the service on 12/31/21 for right angular mandibular swelling that had been evolving for four (4) years. No notion of fever and no notion of

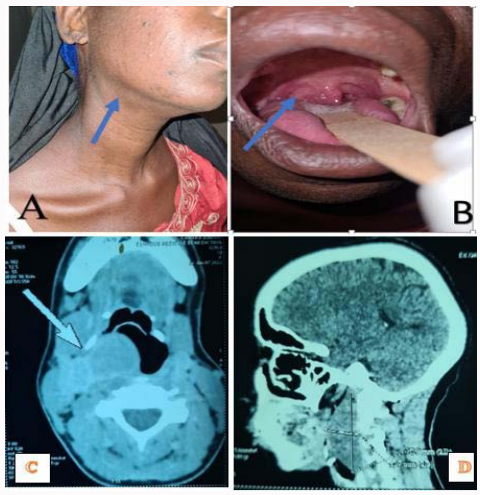

Figure 1: A, B) Preoperative images of the patient; C) CT injected axial section showing a mass vessel at the expense of the right carotid with extension into the oropharynx; D) Sagittal CT scan visualizing a well-circumscribed hypervascularized mass between C1-C7.

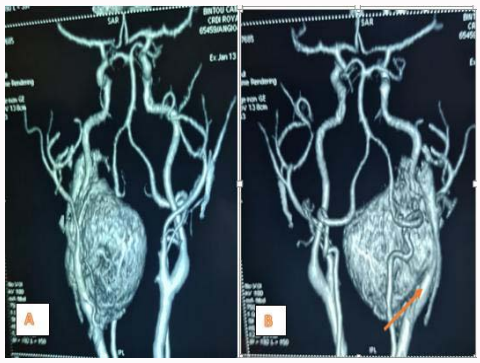

Figure 2: A) Preoperative angiographic image: Identification mass at the level of the right carotid bifurcation without wrapping of the internal carotid arteries and external (front view); B) posterior view.

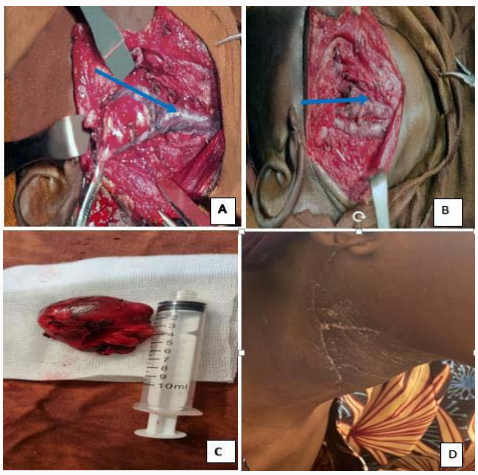

Figure 3: A) Intraoperative image showing the tumor at the level of the right carotid bifurcation before excision; B) Per-operative image showing the macroscopic aspect of the right carotid bifurcation after tumor excision; C) Image of the surgical specimen; D) Image after healing.

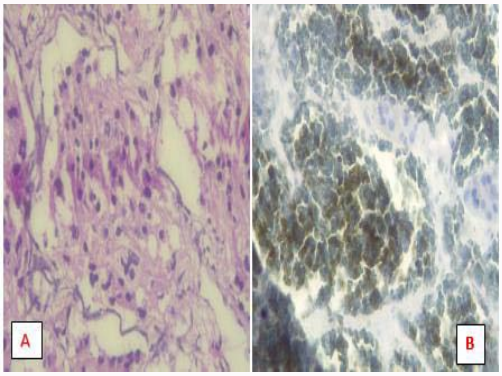

Figure 4: A) Histopathological image in magnification (x100) showing the microscopic appearance of the paraganglioma with clusters of cuboid cells separated by septa richly vascular fibrous arranged in small nests (Zellballen). B) Cytoplasmic staining for neuroendocrine marker chromogranin Ki67 positive at 15%.

pain. It evolved progressively without notion of dysphagia, dyspnea, dysphonia or facial paralysis or rhinolalia. She has no family history; medical and surgical personnel; or local trauma; or endaural surgery; nor of local irradiation and resided since his childhood in a region of low altitude. The patient was in good general condition (stage 1 of the WHO mobility index); Temperature: 37°C; BP: 110/80 mmHg; pulse: 80 beats/min well colored conjunctivae.

Cervical physical examination reveals a subangular-mandibular swelling about 5 cm in diameter on its long axis, of regular contour, mobile in relation to the horizontal plane and fixed in relation to the vertical plane, of firm consistency, not painful to the palpation, pulsatile, the skin opposite was of normal aspect. Examination of the oral cavity found a tumor developed behind the right posterior pillar which pushes the tonsil forward and inwards, causing a slight deviation of the uvula towards the contralateral side, painless, of firm consistency, not bleeding on contact, the overlying mucosa looked healthy (Figure 1).

The CT angiography revealed a deep hypervascularized tumor mass of the carotid sinus at the level of its bifurcation under the 2 carotid branches without intersecting them; compressive with oropharyngeal extension and taking the contrast product, the appearance of which suggests a priori a paraganglioma, classified Shamblin II (Figure 2). The thoracoabdominal CT did not reveal any lesions in favor of a secondary localization.

The patient's standard blood operability assessments returned to normal (NFS, blood crisis). The dosage of urinary catecholamines: The levels of norepinephrine and dopamine were normal. Adrenaline was also within normal limits.

We performed surgical excision without prior embolization by cervicotomy under general anesthesia. The dissection was under adventitial and the coagulation was ensured by the bipolar scalpel, until the total excision of the mass. It was a mass about 5 cm in diameter on its long axis and was located under the 2 carotid branches at the level of its bifurcation without sheathing them. Simple point closure separated from the skin and under the skin after placement of a suction drain. The hospitalization lasted 7 days. The postoperative course was simple and healing was achieved after 10 days of progression (Figure 3). Histological examination of the surgical specimen confirmed the diagnosis of carotid paraganglioma (Figure 4). The patient did not receive radiotherapy; or chemotherapy. With a follow-up of 12 months, we did not note any recurrence.

Table 1: Review of cervical paraganglioma data.

Authors	Year of publication	Number of cases	Age	Sex	Side reached	Classification	Operational Data
Tall [5]	2005	3 cases/8	28; 20; 41 years	F	Unilateral	Shamblin II	Surgical resection without embolization
Ifeoluwa [12]	2017	1 case	8 years	M	Unilateral	Shamblin II	Primary embolization followed by surgical resection
Cimpean [18]	2017	1 case	69 years old	M	Unilateral	Shamblin III	Surgical resection without embolization
Kallel [14]	2018	2 cases/4	49 and 37 years old	F	Unilateral Bilateral	Shamblin III and II	Surgical resections without embolization
Makeieff [16]	2019	2 cases	16 and 15 years old	F/M	Unilateral	Shamblin II	Surgical resection without embolization
Meng-QI [17]	2019	1 case	63 years old	F	Unilateral	Shamblin III	Primary embolization followed by surgical resection
Moscona-Nissana [1]	2022	1	61	F	Bilateral	Shamblin II	Primary embolization then surgery
Our Case	2022	1 case	38 years old	F	Unilateral	Shamblin II	Surgical resection without embolization

Discussion

Hospital frequency aspect

Paragangliomas are rare neoplasms, generally predominant in women with an average age of about 43 years or 8 women for 1 man [1 (Table 1). Of the 832 cervical surgeries performed in the department over its last three years, this is the only case, i.e., 0.12%. The exact incidence is largely unknown as the clinical patterns are usually described in conjunction with pheochromocytomas. The combined estimated annual incidence is approximately 0.8 per 100,000 people in the United States [5,6]. Since the majority of these tumors are unilateral (95%), bilateral Carotid Body Tumors (CBT) represent approximately (5%) of all cases and are generally described in familial forms [7,8].

There are three main etiologies: sporadic, familial and hyperplastic. The sporadic etiology corresponds to 85% to 90% of the total cases. The family form represents 10%. The hyperplastic form of carotid body paraganglioma is mainly related to high altitudes and chronic diseases that cause hypoxemia, such as Chronic Obstructive Pulmonary Disease (COPD) due to possible chronic stimulation of the carotid body [3].

In our case, it was a sporadic case and the patient came from a low altitude region with no history of chronic hypoxia and no family history. Although the most common cause of paragangliomas is sporadic, 30% to 50% of cases have a hereditary component [9]. Most hereditary paragangliomas, especially those of the head and neck, have been associated with pathogenic variants in which they encode different subunits of the Succinate Dehydrogenase (SDH) enzyme complex [9].

SDH plays a central role in energy metabolism as an enzyme of the tricarboxylic acid cycle and as complex II of the mitochondrial respiratory chain, catalyzes the oxidation of succinate to fumarate in the Krebs cycle and the couple with the transfer of electrons to the terminal ubiquinone acceptor in the electron transport chain [10]. Five inherited syndromes linked to SDH mutations have been described all characterized by an Autosomal Dominant (AD) mode of inheritance with variable penetrance, tumor risk and malignancy rates [9]. The SDH gene includes several subunit genes (such as SDHA, SDHB, SDHC, SDHD) and cofactors (SDHAF2) [10]. We did not carry out a Genetic Test on the patient, due to a lack of financial means and a technical platform.

Clinical aspects

The diagnosis is made by combining clinical and imaging findings. Carotid body tumors are rarely secreting tumors. Therefore, they usually present as a painless, slowly growing neck mass. The circumstance of discovery of the paraganglioma in our case was a swelling under the angular-mandibular evolving in a slow way and increasing progressively in volume without any notion of associated pain. Many authors have highlighted the slow and asymptomatic evolution, hence most often its late diagnosis [11,12]. On examination of the neck, we find the "Fontaine sign" where the mass is mobile horizontally but not vertically and can be pulsatile. The fountain sign was present in our patient. An important aspect of CBT evaluation is examination of the cranial nerves, as large CBT can cause cranial nerve compression (IX-XII), transient ischemic attacks, and even stroke [2,4]. We did not find any signs of compression in our patient.

In the case of the secreting form, the signs and symptoms are caused by the excess of catecholamines. Patients may experience headache, palpitations, swelling, flushing, hyperglycemia, fever, nausea, paleness, hypertension, arrhythmias, stroke, or even anxiety that can lead to heart attack myocardium [13]. None of these symptoms were present in our patient.

In imaging, easy to perform and no invasive Doppler is the main diagnostic method since it confirms the anatomical relationship of the tumor with the carotid bifurcation and its vascularization, but its interest in distinguishing the exact boundaries of the tumor from the vessels remains limited [14]. We did not perform ultrasound because the patient was seen with a CT scan that suggested a paraganglioma.

Besides Doppler ultrasound, there are different imaging methods recommended for preoperative evaluation such as Magnetic Resonance Angiography (MRA), Magnetic Resonance Imaging (MRI), Computed Tomography (CT) and CT angiography, Computed Tomography (CT angiography).

Characteristic signs on MRI are "salt and pepper" T2 imaging and lesions that are isointense on T1 and hyperintense on T2 [14]. On CT, CBT are characterized by hypervascular masses located at the carotid bifurcation. CT angiography findings are a hypervascular mass with enlarged arteries, tumor blush, early draining veins, and a lyre sign (flaring of the internal and external carotid arteries). Although the ARM is the benchmark exam for CBT, we do not currently have one within our structure. Our means of diagnosis was therefore Computed Tomography angiography (CT angiography) which enabled us to identify the tumor with precision and to classify it.

Therapeutic aspect

The main treatment for paragangliomas is surgical resection of the tumor. The cure rate after complete resection of a benign tumor of the carotid body is 89% to 100%. Although conservative treatment

can be used for asymptomatic patients, the vast majority will become symptomatic in the future. Shamblin et al. developed a surgical grading system that predicts surgical morbidity. The Shamblin classification divides CBT into three different groups based on operative notes and tumor relationship to vessels [12,15].

The first Shamblin I group includes easily resectable tumors with minimal adhesion to vessels. Group two Shamblin II tumors are partially surrounded by vessels and adhere to the adventitia. Finally, group three Shamblin III tumors are adherent and intimate with the carotid bifurcation. The third group of tumors of the Shamblin classification are the most difficult to remove and present frequent neurological consequences after surgery [2,3]. In our case the tumor was classified Shamblin II.

CBT show extensive growth and can enclose vital neurovascular structures in the neck, making surgical resection extremely difficult due to hypervascularization (with an average blood flow of 200 ml/g per minute) and possible loss of significant blood [14]. For highly vascularized and large CBT, embolization prior to surgery has demonstrated safer resection by decreasing vascularization and tumor size. Occasionally, CBT are unresectable before embolization, but shrink and become resectable [15-17]. The goal of embolization is to selectively obliterate vascular structures after evaluating which artery provides the most blood supply to the tumor [16,17]. Some criteria used by different authors include: Height (usually >3 cm), disease stage and Shamblin III scale [18].

Despite a size >5 cm and although the literature is in favor of embolization before the management of these tumors, we performed an extirpation of the tumor without embolization in our patient, because embolization is not available in our context.

Given its complexity, paraganglioma surgery is not without complications. They are most often neurological. They are unavoidable when they result from the mass ablation of an advanced tumor, from a resection of the X and in the event of definitive ligation of the Internal Carotid Artery (ICA). Accidental complications occur either during the dissection or because of the approach. Medialization of the vocal cord, early postoperative rehabilitation and protection of the airways by temporary nasogastric tube limit the morbidity of such lesions. Neuroendocrine complications are of two types: Either blood pressure decompensation during excision of secreting tumors or due to the unrecognized presence of a pheochromocytoma, or blood pressure lability observed in the event of bilateral excision of a paraganglioma due to removal of the carotid paraganglioma-carotid sinus complex [6]. Paragangliomas are highly vascularized tumors and resection can result in significant blood loss during the procedure associated with high rates of cranial nerve palsy and even stroke and mortality. The risk of these complications varies from 0% to 13% [15,18]. In our case, we did not encounter any intraoperative or postoperative complications. The postoperative course was simple with a healing time of 10 days.

These paragangliomas are considered to be not very radiosensitive, in particular for carotid paragangliomas. Radiotherapy is indicated in the event of surgical contraindication (paraganglioma with basal cranial extension, advanced cervical paraganglioma or bilateral form in the elderly) or in addition to surgery (recurrence, malignant form or residual tumor) [13,19].

Gross features involve a tumor that rarely exceeds 6 cm in diameter and grows near or envelopes the bifurcation of the common carotid artery. It has a gross fleshy appearance and the tissue is red-pink to brown due to bleeding or fibrosis [9,20]. Histologically, the tumor consists of polygonal or spindle-shaped chief cells with eosinophilic and uniform cytoplasm arranged in small nests (Zellballen) surrounded by sustentacular cells. The nests are separated by a delicate fibrovascular stroma. The nuclei are round or oval and have prominent nucleoli [9,20]. Principal cells stain strongly for neuroendocrine markers, including chromogranin, synaptophysin, neuron -specific enolase, CD56 and CD57. The sustentacular cells are positive for the S-100 protein [15,20].

Conclusion

Paraganglioma is a rare and benign vascular tumor that most often manifests as a laterocervical mass with an asymptotic evolution. The echo Doppler, Angio scanner can be of a capital contribution for the diagnosis. Its management is mainly surgical and must be early to avoid complications. The surgical methods depend on the tumor stage (Shamblin).

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