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Bilateral Carotid Paraganglioma: Case Report; Successful Surgical Excision, Radiological Image and Review of the Literature

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Abstract **Case Report**

Introduction: Carotid body tumors are rare neoplasms that generally present as asymptomatic slow-growing masses in the neck, commonly called cervical paragangliomas, highly vascularized whose cells originate from the embryonic neural crest, carotid glomus tumor is uncommon, representing 1 out of every 30,000 head and neck tumors, they are located in the adventitia of the vessels, they usually grow surrounding the arteries of the carotid bifurcation and the nerves that surround it, in Ecuador the prevalence is at 55 years of age with greater frequency in women [1]. ***Clinical Case:*** A 33-year-old female patient with no significant clinical history, presented with a painful hard mass in the right cervical region that progressively increased in size, for the past 6 months. On physical examination, a tumor in the right cervical region of approximately 5 centimeters in diameter was found, with a hard consistency attached to deep planes, for which Eco Doppler and AngioTac of the neck were performed, which reported bilateral carotid paraganglioma. Excision of the glomus was necessary due to its size right carotid artery, surgery using a right cervico-lateral approach, identifying the vascular tumor mass of 4.8 x 2.9 x 2.8 cm in diameter, which was located in the carotid bifurcation. Removal of Shamblin II right carotid body paraganglioma by temporary clamping of the common carotid artery and the digital dissection method, there was no intraoperative massive bleeding or cranial nerve deficits in the postoperative period with favorable evolution, surgical wound in good condition during follow-up in outpatient consultation. ***Conclusion:*** Glomus carotid tumors are generally benign and diagnosed late. Imaging studies are essential for its diagnosis and staging.

Keywords: Paraganglioma, Carotid Glomus, Carotid body.

INTRODUCTION

Carotid body tumors are rare neoplasms that generally present as asymptomatic slow-growing masses in the neck, commonly called cervical paragangliomas, highly vascularized whose cells originate in the embryonic neural crest, 90% of paragangliomas are located in the gland adrenal gland, while 85% of extra-adrenal paragangliomas are located in the abdomen, 12% in the thorax, and only 3% in the head and neck, the glomus carotid tumor is a rare paraganglioma, representing 1 in 30,000 tumors of head and neck. They are located in the adventitia of the vessels and usually grow around the arteries of the carotid bifurcation and the nerves that surround it. They can arise in any area of the body that contains embryonic cellular remnants of the neural crest and are also part of neuroendocrine tumors [2].

The epidemiology in Ecuador has a prevalence greater than 55 years (range, 40-59 years). There being a higher frequency of occurrence in women, with a male to female ratio of 1 to 9, these tumors develop in 57% of cases on the right side of the carotid. While 25% are located on the left side and 17% are usually bilateral [2]. Of all cases, only 1% will have functional capacity and an average of 4.3% may present malignancy, similar to the data presented worldwide. Carotid body tumors are of low frequency, having an

incidence of 1 to 3 in 100,000 people, representing 0.6% of all head and neck tumors. They have a low malignancy rate, their complications are related to compression of neighboring structures during their growth. Malignant paragangliomas are those that present metastasis, vascular invasion, and necrosis. The familial appearance of paragangliomas has been widely documented and represents 10 to 40% of cases [3].

CLINICAL CASE

A 33-year-old female patient with no significant clinical history, presented with a painful hard mass in the right cervical region that had been progressively increasing in size for the past 6 months. On physical examination, a tumor was found in the right cervical region of approximately 5 centimeters in diameter, hard consistency adhered to deep planes, an echo Doppler of the neck was performed, which reported: a solid hypoechoic, hypervascular expansive process with an epicenter in the right carotid space, at the level of the carotid bifurcation, which appears displaced by the finding, measures approximately 4.7 x 30 x 21 mm with a volume of 13cc, a finding suggestive of paraganglioma.

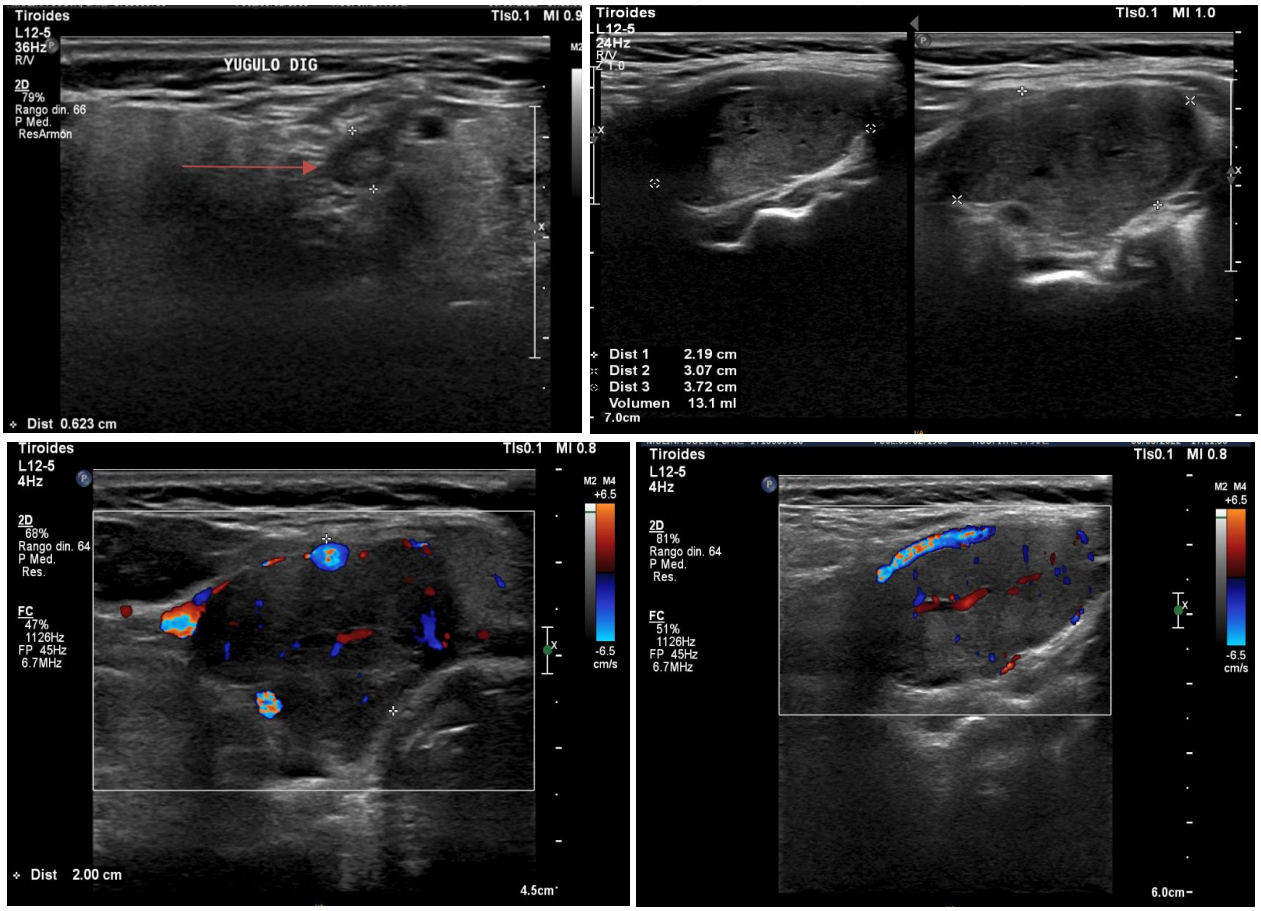

Figure 1: Echo Doppler of the neck

Ultrasound of carotid glomus 1a - b). A partially defined hypervascular hypoechogenic mass with epicenter in the right carotid space is observed, at the level of the carotid bifurcation, which appears displaced by the finding that measures approximately 4.7 x 30 x 21 mm with a volume of 13cc. 3-4a). on color Doppler the mass shows increased flow, which is located between two important structures.

To continue studies, an Angio-Tac of the neck is performed, which reports: at the level of the right carotid bifurcation, a well-defined structure is identified, which includes the proximal segments of the internal and external carotid, which after the injection of intravenous contrast, shows avid uptake, It measures 4.8 x 2.9 x 2.8 cm. Compression and slight displacement of the ipsilateral internal jugular vein are also observed. At the level of the bifurcation of the left carotid vessels, another image with similar characteristics is observed, measuring 8.7x8.9x13.3mm.

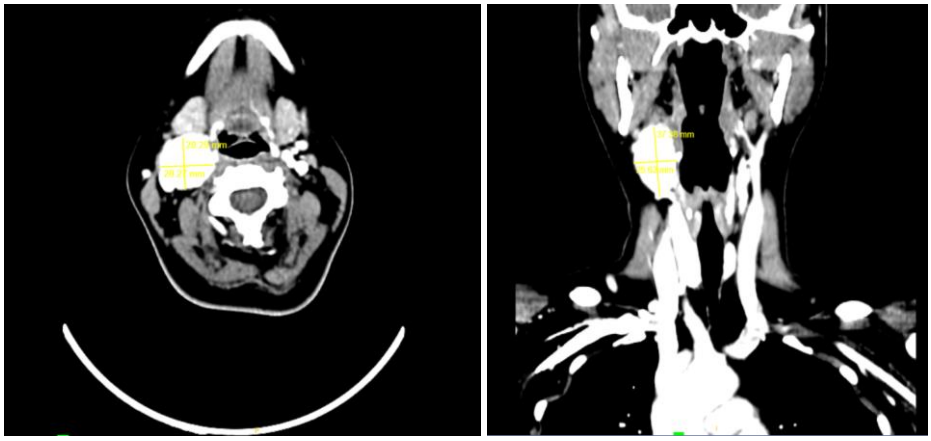

Figure 2: Angio-Tac of the neck

Neck CT angiography 2a-b) with non-ionic contrast shows heterogeneous enhancement of the mass located at the bifurcation level of the common and right carotid artery that encompasses the proximal segments of the internal and external carotid that measures 4.8 x 2.9 x 2.8 cm, compression and slight displacement of the ipsilateral internal jugular is also observed. At the level of the bifurcation of the left carotid vessels, a mass with similar characteristics measured 8.7x8.9x13.3mm.

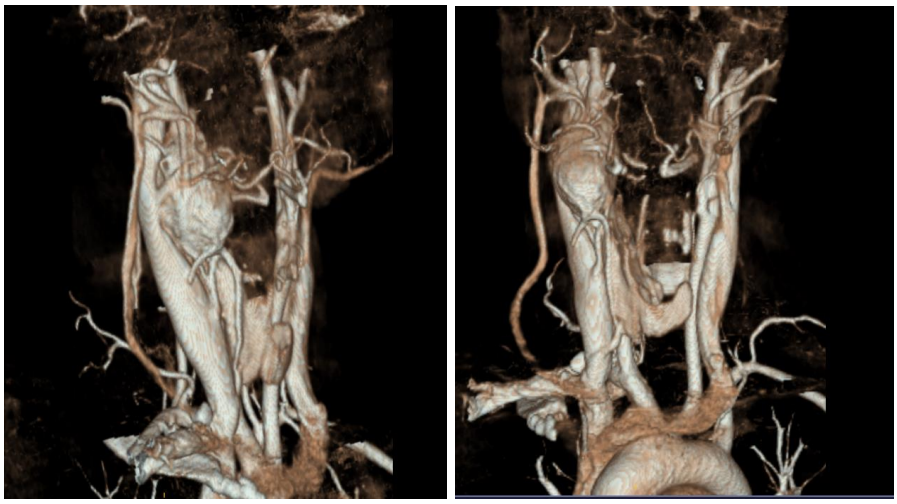

Figure 2 cd): 3D volume rendering reconstruction: the carotid glomus, its location and its hypervascular component can be observed in greater detail

Therefore, it is diagnosed as a STAGE II glomus carotid tumor. Surgical intervention is necessary in order to resolve this anomaly. She underwent surgery through a right cervico-lateral approach, identifying the vascular tumor mass of 4.8 x 2.9 x 2.8 cm in diameter, which was located in the carotid bifurcation.

The postoperative diagnosis was Shamblin II carotid glomus (Figure 3b).

RESULTS AND FOLLOW-UP

Removal of Shamblin II right carotid body paraganglioma by temporary clamping of the common carotid artery and the digital dissection method, there was no intraoperative massive bleeding or cranial nerve deficits in the postoperative period with favorable evolution, surgical wound in good condition during follow-up In outpatient consultation, with respect to left carotid paraganglioma, he is kept under observation.

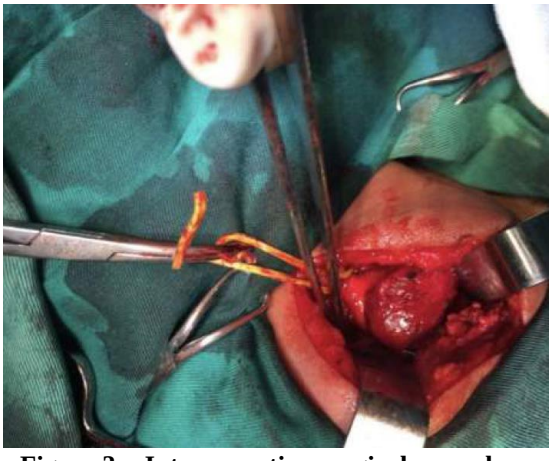

Figure 3a: Intraoperative surgical procedure

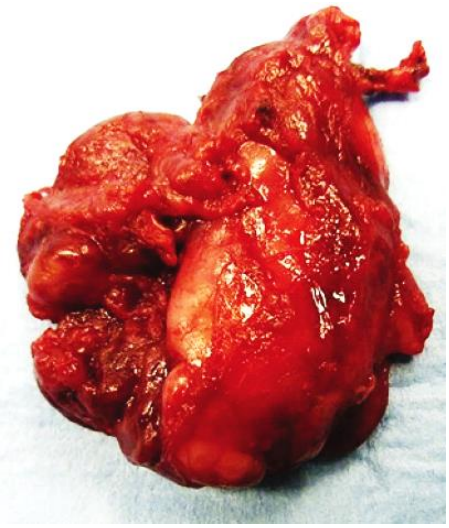

Figure 3b: Right paraganglioma

DISCUSSION

This pathology generally presents in middle adulthood, as asymptomatic non-functioning masses. However, in less than 1% of cases they may present symptoms, due to the secretion of catecholamines, manifesting with constant or paroxysmal arterial hypertension, palpitations, headache, pallor, tachycardia, diaphoresis, weight loss and hyperglycemia. In previous studies, they describe that the first clinical manifestation is usually the increase in size of the mass. While, in other cases there is the presence of a painful mass and slow increase in the size of the lateral face of the neck [4]. In the case studied, the patient presented a mass on the right side of the neck that was increasing in size [5].

On physical examination, a mass is found located under the deep mandibular angle to the anterior edge of the sternocleidomastoid muscle, painless on palpation in certain cases, it is usually mobile laterally, but fixed longitudinally [5], this is a semiological characteristic that is known as Fontaine's sign and is due to its location within the carotid sheath, they are generally unilateral, although 4% of sporadic cases and up to 31% of family members are bilateral or associated with paragangliomas in other anatomical sites, in In these cases, special considerations must be applied. Initially, it is important to know that there is a genetic predisposition, which places these patients at greater risk of developing additional or multicentric lesions. In the exposed case, there was no family history of the pathology [6].

Diagnosis

Computed tomography and magnetic resonance imaging allow diagnostic approximation and initial classification, while angiography allows the use of selective embolization, whose technique in these tumors is controversial. Computed tomography presents a sensitivity of 77–98% and a 29–92% specificity for detecting paragangliomas, while magnetic resonance imaging has a higher sensitivity, being 90–100%, as well as a specificity of 50–100% [7]. The characteristic imaging sign is the lyre sign, which was initially described in the angiographic study, where the tumor located in the carotid bifurcation separates the internal and external carotid arteries, as we can see in the imaging study of the patient [8].

The histopathological diagnosis in most cases can be made only with hematoxylin and eosin, few need immunohistochemical reactions to confirm the diagnosis. In addition, the measurement of serum free metanephrines can be used, this technique is considered the best test to exclude or confirm the presence of a pheochromocytoma or a functional paraganglioma [9].

Treatment

Surgery is the only curative treatment and is considered the treatment of choice in most cases, while radiotherapy is indicated in cases of incomplete resections or when surgery is contraindicated due to unresectability, recurrence, or malignancy. Since the morbidity of surgical treatment of paragangliomas increases with their size, it is suggested that patients at risk should be evaluated by physical examination, urinary catecholamine analysis, and head and neck MRI every 1 to 2 years starting at age 14 to 16 years, in order to detect tumors at an early stage. In the case of the patient, there is no family relationship, a mass of medium size [10].

According to the paraganglioma scale, stage I by Zanareth and did not present other masses in the imaging study; Therefore, a good evolution with a good prognosis is expected. Despite this, an annual control should be followed, after which follow-up will not be necessary unless there is another finding of a mass [10].

Alternatives to surgery

Surgery is simple and free of complications, as there is an early diagnosis. That is, in those cases where a small tumor is present. In the case of older adults, with a short life expectancy, who develop small and asymptomatic tumors, “wait-and-see” management can be opted for [9].

In situations of surgical contraindication, expectant management is advisable according to some authors, although the issue remains controversial. Even so, other possibilities are increasingly being contemplated, such as radiotherapy or follow-up without treatment in specific cases [8].

CONCLUSION

Glomus carotid tumors are generally benign and diagnosed late. Imaging studies are essential for its diagnosis and staging. Early surgical treatment of these tumors is well established.

Conflicts of Interest: The authors declare that there is no conflict of interest regarding the publication of this paper.

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REFERENCES

1. Andraska, E., Haga, L., Reitz, K., Li, X., & Ramos. (2019). *Epidemiological Characterization of Patients with a Diagnosis of Carotid Glomus Undergoing Resection*. UTA Medical Journal.
2. Borroto, A. (2017). Epidemiology of glomus tumors of the head and neck. Cuban Journal of Otolaryngology, 5.
3. Hogan, A. R., Sola, J. E., Jernigan, S. C., Peterson, E. C., & Younis, R. T. (2018a). A pediatric carotid body tumor. *Journal of Pediatric Surgery*, *53*(7), 1432–1436. https://doi.org/10.1016/j.jpedsurg.2018.04.004
4. Hogan, A. R., Sola, J. E., Jernigan, S. C., Peterson, E. C., & Younis, R. T. (2018b). A pediatric carotid body tumor. *Journal of Pediatric Surgery*, *53*(7), 1432–1436. https://doi.org/10.1016/j.jpedsurg.2018.04.004
5. Jansen, TTG, Marres, HAM, Kaanders, JHAM, & Kunst, HPM (2018). A meta-analysis on the surgical management of paraganglioma of the carotid body per Shamblin class. *Clinical Otolaryngology* , *43* (4), 1104–1116. https://doi.org/10.1111/coa.13116
6. Moore, M. G., Netterville, J. L., Mendenhall, W. M., Isaacson, B., & Nussenbaum, B. (2016). Head and Neck Paragangliomas. *Otolaryngology-Head and Neck Surgery (United States)*, *154*(4), 597–605. https://doi.org/10.1177/0194599815627667
7. Nicholas, R.S., Quddus, A., Topham, C., & Baker, D. (2015). Resection of a large carotid paraganglioma in Carney-Stratakis syndrome: A multidisciplinary feat. *BMJ Case Reports*, *2015*, 1–6. https://doi.org/10.1136/bcr-2014-208271
8. Schmid, M., Raithel, D., Hahn, E. G., Daniel, W. G., & Martin, R. (2012a). Glomus caroticum tumor as rare cause of recurrent syncope. In *Clinical Research in Cardiology*, 101(6), 499–501. https://doi.org/10.1007/s00392-012-0429-1
9. Asquel Cadena, V. H., García, C., Vargas Brazales, A. B., & Díaz Piedrahita, M. A. (2021). Diagnosis and treatment of carotid glomus tumor, about a case. *UTA Medicines*, *5*(4), 27. https://doi.org/10.31243/mdc.uta.v5i4.1421.2021
10. Schmid, M., Raithel, D., Hahn, E. G., Daniel, W. G., & Martin, R. (2012b). Glomus caroticum tumor as rare cause of recurrent syncope. In *Clinical Research in Cardiology*, 101(6), 499–501. https://doi.org/10.1007/s00392-012-0429-1