

Case report of a neuroendocrine neoplasm: findings on ultrasound and angiotomography of a carotid body tumor

Neoplasia neuroendocrina: hallazgos en ecografía y angiotomografía de un tumor del cuerpo carotídeo. Presentación de caso

Andrés Felipe Mejía León[1]
Carlos Eduardo Lucio Cortés[1]
Laura Manuela Olarte Bermúdez[2]
José Chaustre[1]
José Luis Roa Benavides[3]

https://doi.org/10.53903/01212095.305

Key words (MeSH)
Carotid body
Neuroendocrine neoplasms
Ultrasonography

Palabras clave (DeCS)
Cuerpo carotídeo
Neoplasias neuroendocrinas
Ultrasonografía

Summary

The carotid paraganglioma is an infrequent neuroendocrine tumor originating from the paraganglia of the autonomic system and presents challenges in differential diagnosis due to its low incidence and diversity of clinical presentations. In this article, we will present a case of an 82-year-old woman with varied symptoms and highlight the importance of multidisciplinary management in the diagnosis and treatment of this pathology. Emphasis is placed on radiological findings, such as ultrasound and computed tomography angiography, which are crucial to confirm the diagnosis. Additonaly, the importance of surgical resection as the treatment of choice is pointed out. In conclusion, a comprehensive and early approach based on clinical evaluation and imaging tools can facilitate the diagnosis and timely management of carotid paraganglioma.

Resumen

El paraganglioma carotídeo es un tumor neuroendocrino infrecuente que se origina en los paraganglios del sistema autónomo. Plantea desafíos en el diagnóstico diferencial debido a su baja incidencia y a la diversidad de presentaciones clínicas. En este artículo se documenta el caso de una mujer de 82 años de edad con síntomas variados y se destaca la importancia del manejo multidisciplinario en el diagnóstico y tratamiento de esta patología. Se enfatiza en los hallazgos por imágenes de ecografía, angiografía por tomografía computarizada y resonancia magnética, herramientas cruciales para confirmar el diagnóstico. Además, se señala la importancia de la resección quirúrgica como tratamiento de elección. En conclusión, un abordaje integral y precoz basado en la evaluación clínica y las herramientas de imagen puede facilitar el diagnóstico y el manejo oportuno del paraganglioma carotídeo.

Introduction

Carotid paraganglioma, belonging to the glomangioma or paraganglioma group, is a rare neuroendocrine tumor that originates in the paraganglia of the autonomic nervous system (1). It has an incidence of 1 to 2 per 100,000 people, tends to be prevalent in women, with an approximate ratio of 1:3, and occurs most frequently in individuals aged 40 to 60 years (2,3). Most of these tumors arise in the head and neck, and many of them have a hereditary component (1). Fifty-seven percent of these tumors occur on the right side, 25% on the left side (4), and 5% of cases may be bilateral (3).

The diagnosis of paragangliomas is based on medical history, physical examination, and radiological imaging, such as ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI). Currently, the importance of multidisciplinary management is emphasized, as they present challenges in differential diagnosis. This study will review a case and emphasize the radiological findings and differential diagnoses of carotid body paraganglioma.

Case description

An 82-year-old woman with a history of smoking presented with 20 days of fatigue, weakness, palpitations accompanied by oppressive chest pain, generalized myalgia, and respiratory symptoms such as dyspnea and dry cough. The patient denied any recent trauma. Physical examination revealed lower limb edema, pulmonary rales on auscultation, desaturation, and a pulsatile, firm, painless mass on the right side of the neck. Given these findings, additional tests were performed to rule

[1]Radiology Resident, Hospital Universitario Fundación Cardioinfantil, Institute of Cardiology, Department of Radiology and Diagnostic Imaging, Universidad del Rosario. Bogotá, Colombia.

[2]Student, University of La Sabana. Department of Radiology and Diagnostic Imaging, Hospital Universitario Cardioinfantil. Bogotá, Colombia.

[3]Radiologist, Department of Radiology and Diagnostic Imaging, Sabana. Department of Radiology and Diagnostic Imaging, Hospital Universitario Cardioinfantil. Bogotá, Colombia.

out decompensated heart failure, and imaging tests were performed to characterize the neck lesion.

Initially, a diagnosis of carotid paraganglioma was suspected after a detailed analysis of the patient's medical history and a thorough physical examination. Additional studies included ultrasound and neck CT angiography. The neck ultrasound (Figure 1) revealed lesions in both carotid bifurcations, which were considered initial differential diagnoses of bilateral carotid paragangliomas, cervical adenopathies, and schwannomas. However, Doppler examination showed increased predominantly central flow in the lesion, confirming the diagnosis of carotid body tumor: carotid paraganglioma. A neck CT angiogram was performed to better characterize the findings (Figure 2).

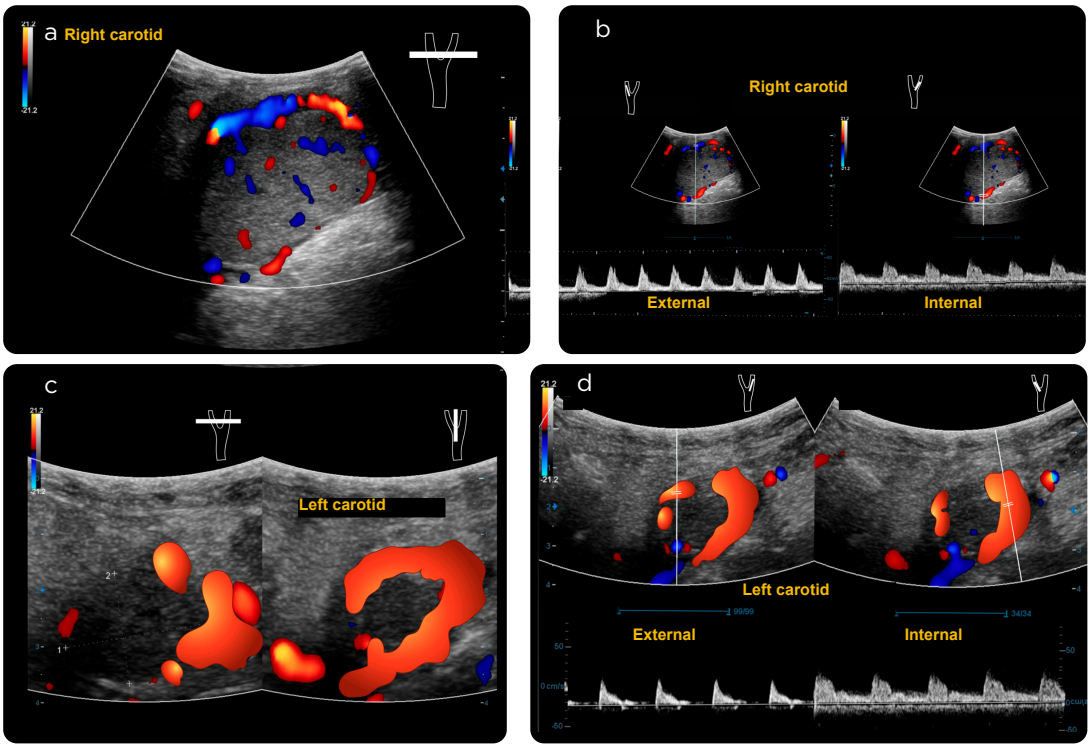


Figure 1. Doppler ultrasound of the neck. In the right half of the neck, centered on the carotid bifurcation and separating both carotid arteries, a solid, oval, circumscribed lesion is visible, with lobulated contours and heterogeneous echogenicity, predominantly hypoechoic and with small cystic areas inside. a) Color Doppler: marked increase in central flow. b) Separates both carotid arteries, with preservation of their flow. c and d) In the left carotid bifurcation: lesion with similar characteristics and smaller size.

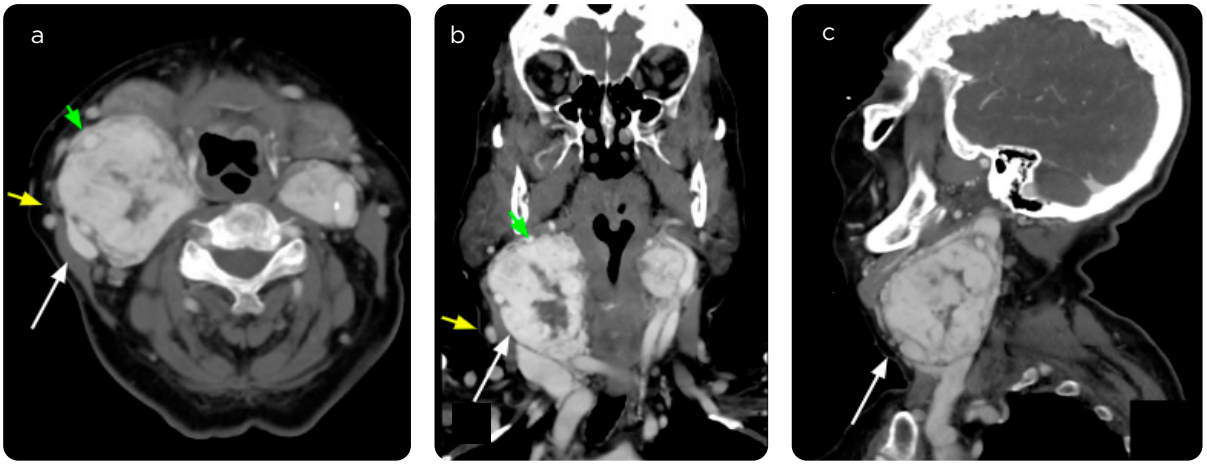


Figure 2. Contrast-enhanced CT angiography of the head and neck. a) Axial acquisition, b) coronal reconstruction, and c) right parasagittal reconstruction. Well-defined, lobulated lesions are visible in both carotid bifurcations, showing intense enhancement with contrast medium. These exert moderate compression on the internal jugular veins, displacing the internal carotid arteries posteriorly and the external carotid arteries anteriorly; they contact less than 180° of the circumferences of the internal carotid arteries.

Discussion

The carotid body was first described by Von Haller in 1743 (2). Its cells arise from embryonic neural crest cells surrounding Jacobsen's nerve (*paraganglioma tympanicum*) or the adventitia of the jugular bulb (*paraganglioma jugulare*) (3). Paragangliomas are characterized by slow growth, benign appearance, and high vascularity (5). The most common locations are: the bifurcation of the common carotid artery (carotid paraganglioma) in 35% of cases, around the vagus nerve in the carotid space (*vagal paraganglioma*) in 11%, in the jugular foramen (*jugular paraganglioma*), and in the middle ear (*tympanic paraganglioma*) (6). Additionally, they can be adrenal (the most common is pheochromocytoma) and extra-adrenal (abdomen, Zuckerkandl organ and thorax, paravertebral and cardiac) (7). However, those in the head and neck, which are associated with the parasympathetic system, account for up to 70% of extra-adrenal paragangliomas (7).

Clinically, they manifest as a painless, slow-growing mass in the neck, some with symptoms such as dysphagia, hoarseness, or breathing difficulties, as in the case presented (8); on several occasions, different types of studies are required to assess the extent of the tumor in detail (3). As in this case, the predominant histological pattern is that of epithelioid cells arranged in distinct groups separated by a prominent capillary network (2).

The differential diagnoses of carotid paraganglioma include other neck masses, both congenital and inflammatory, infections, benign and malignant lesions, and vascular lesions (1). In some cases, they can be confused with lymph nodes, but they are distinguished by their high vascularity, which is one of the main characteristics in imaging studies (7).

In imaging methods, the typical finding is a hypervascular soft tissue mass located at the bifurcation of the common carotid artery, causing separation of the carotid arteries (2). Doppler ultrasound will show a well-defined, heterogeneous, hypoechoic, highly vascularized mass with a pattern of carotid artery displacement; this is the method of choice for diagnosis and follow-up in cases of non-surgical management (1). CT angiography will reveal anatomical and vascular details and determine the local extent (1). On MRI, they typically have a signal similar to that of muscle on T1-weighted images and increased signal on T2-weighted images, with classic signs of well-defined flow voids appearing as a "salt and pepper sign" on T2-weighted images, representing the internal vessels.

Imaging studies show restriction due to their high cellularity (6). Treatment requires a multidisciplinary approach. In most cases, the treatment of choice is surgical resection, with the aim of performing a complete excision to preserve the vessels (7). For lesions larger than 2 cm, preoperative embolization is recommended to reduce the risk of bleeding and morbidity (4). However, in elderly patients or those with multiple comorbidities, radiotherapy is an excellent alternative.

Conclusion

Carotid body tumors are rare, but their clinical and imaging presentations are quite characteristic. With an appropriate approach, considering the differential diagnoses of neck masses and using the appropriate imaging studies, an accurate diagnosis can be made in most cases, allowing for early and timely multidisciplinary management.

References

1. Lin EP, Chin BB, Fishbein L, Moritani T, Montoya SP, Ellika S, Newlands S. Head and neck paragangliomas: Am updated on the molecular classification, state of the art imaging, and management recommendations. Radiol Imaging Cancer. 2022;4(3):e210088. https://doi.org/10.1148/rycan.210088
2. Hoang VT, Trinh CT, Lai TAK, Doan DT, Tran TTT. Carotid body tumor: a case report and literature review. J Radiol Case Rep. 2019;13(8):19-30. https://doi.org/10.3941/jrcr.v13i8.3681
3. Cárdenas K, Zamora G. Radiología en el glomus carotídeo. Presentación de un caso. Imaging in carotid body paraganglioma. A case report. Metro Ciencia. 2021:29(2):63-9. https://doi.org/10.47464/MetroCiencia/vol29/3/2021/63-69
4. González-Urquijo M, Hinojosa-González D, Viteri-Pérez VH, Llausas-Villarreal A, Becerril-Gaitan A, González-González M, et al. An analysis from the CAPACITY database of outcomes of preoperative embolization before carotid body tumor surgery compared with resection alone. J Vasc Surg. 2023;77(5):1447-52. https://doi.org/10.1016/j.jvs.2023.01.012
5. Delantoni A, Raftopoulos A, Pol Anagnostaki A, Orhan K. B-mode and color Doppler imaging of carotid paragangliomas in different neck regions. J Ultrason. 2020;20(82):e218-e221. https://doi.org/10.15557/JoU.2020.0037
6. Alqhtani AJ, Al Qahtani SM, Alharbi MK, Aljehani MH, Alharbi A. Carotid body tumor: a case report of a rare neuroendocrine neoplasm. Cureus. 2023;15(7):e42224. https://doi.org/10.7759/cureus.42224.
7. Avgerinos N, Avgerinos I, Troupis T, et al. Carotid Body tumor in a 26-year-old male patient managed with preoperative embolization. Curaeus. 2023;15(12):e 49917. https://doi.org/10.7759/cureus.49917
8. Kihara C, Patel S, Moss R. A rapidly progressing carotid body tumor: a case report. Cureus.2023;15(8): e43654. https://doi.org/10.7759/cureus.43654

Correspondence

Laura Manuela Olarte Bermúdez
Universidad de La Sabana
Bogotá, Colombia
lauraolbe@unisabana.edu.co

Received for evaluation: January 16, 2024
Accepted for publication: February 23, 2024