CAROTID BODY PARAGANGLIOMA: EARLY DIAGNOSIS

PARAGANGLIOMA CAROTIDEO: UNA DIAGNOSI PRECOCE

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Parole chiave: tumori neuroendocrini, paraganglioma, carotide

Abstract

Background: Paragangliomas (PGLs) are rare neuroendocrine tumors which arise from the extraadrenal chromaffin tissue of the autonomous nervous system, located in the carotid body or glomus (CB). This is a little corpuscle localized at carotid bifurcation; it contributes to the regulation of blood pressure, cardiac and respiratory frequency by giving information to the nervous system bodies located in the brain stem. Histologically, PGLs remind of the CB normal architecture, with clusters cells known as Zellballen, highly vascularized. Rarely secreting vasoactive amines, they come more often to clinical observation as a mass located at the mandibular angle, asymptomatic. Based on the size and connections with the surrounding vascular structures, they are divided into three groups according to the Shamblin classification.

Case report: 57 year old patient, smoker. He reports about a year before, presence of vertigo associated with cold sweat and general malaise that lasts about 20 min resolved spontaneously (blood pressure in association with these episodes is high with maximum values recorded around 180/110 mmHg). He is submitted to a Holter blood pressure investigation, ECD of the neck vessels (TSA), then it was performed a neck MRI. The patient underwent surgery. The final histological examination showed a "paraganglioma composed of nests of chromogranin positive cells with round nuclei and finely dispersed chromatin with abundant eosinophilic, granular cytoplasmic portion".
**Discussion and Conclusions:** The PGLs are rare neoplasms, with an incidence of 0.012%. Very rarely (5% of cases) the patients come to medical attention for the appearance of a "pressor amine syndrome" with tachycardia, palpitations, flushing, hypertensive crisis, dizziness, malaise, diarrhea, amino-linked activity consequences of PGLs secreting. According to the Shamblin classification are divided into three groups based on the size and relationships contracted with the surrounding vascular structures (group III belong to the larger and it forms persistently adhesions to the carotid bifurcation). In most cases, the patients having a PGLs come to medical attention for the appearance of a mass in the lateral region of the neck, at the mandibular angle, often completely asymptomatic. When the tumor reaches considerable size, it may be responsible for nerve deficits (especially for X, XI and XII nerves), dysphagia, neck pain. Early diagnosis of PLGs asymptomatic and small reduces the risk of perioperative lesions of the cranial nerves and carotid arteries. In our case, early identification was made possible by the biological and functional characteristics of the tumor and the consequent clinical manifestations, allowing to carry out the surgery safely for the patient. Despite the size of PLG taken into account, it was ranked to the Shamblin group I.

**Abstract**

**Introduzione:** I chemodectomi (o paragangliomi) carotidei (CC) sono rare neoplasie che originano dalle cellule chemorecettive del glomo carotideo (GC). Il GC è un corpuscolo localizzato a livello della biforcazione carotide; contribuisce alla regolazione della pressione arteriosa, della frequenza cardiaca e respiratoria inviando informazioni ai centri nervosi regolatori situati nel tronco encefalico. Istitologicamente i CC ricordano la normale architettura del GC, con cellule organizzate in clusters detti Zellballen, altamente vascolarizzati. Raramente secernenti amine vasoattive, giungono più spesso all’osservazione clinica come una massa localizzata a livello dell’angolo mandibolare, asintomatica. Sulla base delle dimensioni e dei rapporti con le strutture vascolari circostanti vengono suddivisi in tre gruppi sec. La classificazione di Shamblin.

**Caso Clinico:** Pz di 57 aa, fumatore. Riferisce da circa un anno la comparsa di vertigini oggettive associate a sudorazione algida e malessere generale della durata di circa 20 min che regrediscono spontaneamente (la pressione arteriosa in concomitanza di tali episodi risulta elevata con valori massimi registrati di 180/110 mmHg). Viene sottoposto ad indagini strumentali tra cui un Holter pressorio, un ECD dei vasi epiaortici (TSA) e Angio-Rm del collo. Il pz viene sottoposto ad intervento chirurgico. L’esame istologico definitivo evidenzia un “paraganglioma costituito da nidi di cellule cromogranina positive con nucleo rotondo e cromatina finemente dispersa, con abbondante quota citoplasmatica eosinofila e granulare”.

**Discussione e Conclusioni:** I CC sono neoplasie rare, con un’incidenza dello 0.012%. Secondo la classificazione di Shamblin vengono suddivisi in tre gruppi sulla base delle dimensioni e dei rapporti che contraggono con le strutture vascolari circostanti. Nella maggioranza dei casi i pz portatori di un CC si presentano all’attenzione del medico per la comparsa di una massa in regione laterale del collo, a livello dell’angolo mandibolare, spesso del tutto asintomatica. Quando la neoplasia raggiunge notevoli dimensioni, può rendersi responsabile di deficit nervosi (soprattutto a carico dei nn X, XI e XII), disfagia, dolore cervicale. Molto più raramente (5% dei casi) i pz giungono all’attenzione del medico per la comparsa di una “pressor amine syndrome” con tachicardia, palpitazioni, flushing, crisi ipertensive, vertigini, malessere generale, diarrea, legata all’attività amino-secerente dei CC. La diagnosi precoce di CC asintomatici e di piccole dimensioni riduce i rischi perioperatori di lesioni a carico dei nervi cranici e delle arterie carotide. Nel nostro caso, la precoce identificazione è stata permessa dalle caratteristiche biologiche e funzionali del tumore e dalle conseguenti manifestazioni cliniche, consentendo di condurre l’intervento chirurgico in tutta sicurezza per il paziente. Nonostante le dimensioni del CC preso in considerazione, lo classificavano al I gruppo di Shamblin.
Background

Paragangliomas (PGLs) are rare neuroendocrine tumors which arise from the extraadrenal chromaffin tissue of the autonomous nervous system, located in the carotid body or glomus (CB). This is a little corpuscle localized at carotid bifurcation; it contributes to regulating blood pressure, cardiac and respiratory frequency by giving information to the nervous system bodies located in the brain stem. Isto logically, PGLs remind of the CB normal architecture, with clusters cells known as Zellballen, highly vascularized. Rarely secreting vasoactive amines, they come more often to clinical observation as a mass located at the mandibular angle, asymptomatic. Even more rarely they behave as malignant lesions giving distant metastases. Based on the size and relationship with the surrounding vascular structures, are divided into three groups according to the Shamblin classification (1) (Tab. 1).

Tab. 1 - Shamblin Classification

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>Type I</td>
<td>small tumors, with poor adhesion to the carotid artery, with cleavage plane</td>
</tr>
<tr>
<td>Type II</td>
<td>larger tumors, with adequate adhesion to the arterial wall and partial involvement of the origin of the internal and external carotid</td>
</tr>
<tr>
<td>Type III</td>
<td>full involvement of the bifurcation, with considerable adhesion to the arterial wall, with difficult cleavage plane</td>
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</tbody>
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Case Report

57 year old patient, smoker. Reports about a year before the presence of vertigo associated with cold sweat and general malaise that lasts about 20 min resolved spontaneously (blood pressure in association with these episodes is high with maximum values recorded around 180/110 mmHg). He was submitted to a Holter blood pressure investigation, which documents "mean daytime systolic and in 24 hours values more than referenced ones", and ECD of the neck vessels (TSA) highlighting "at the right carotid bifurcation presence of nodularity, size 27mm X 17mm". Then a neck MRI was performed which "confirms the presence of nodularity at the carotid bifurcation of the size of 27mm X 20mm (Fig. 1), with moderate post-contrast enhancement and that displaces the external and internal carotid with no stenosis; this nodularity is also not infiltrating the surrounding tissues" (Fig. 2).

The patient underwent surgery. The surgery, conducted in loco-regional anesthesia, consisted of the cleavage of the "nodularity" from the carotid bifurcation after hemostasis with metal clips of glomus vessels and removal of this nodularity, in its entirety, without the need for clamping the carotid arteries. The final histological examination showed a "paraganglioma composed of nests of chromogranin positive cells with round nuclei and finely dispersed chromatin with abundant eosinophilic, granular cytoplasmic portion" (Fig. 3). In the one year follow-up the patient has no longer presented episodes of vertigo or general asthenia, and a new Holter blood pressure investigation documented the normalization of blood pressure both day and night average values, while the control of TSA Doppler ultrasound showed no evidence of disease recurrence.
Fig. 1 - *Right glomus tumor: long and transverse sections without contrast*

![Image of Right glomus tumor](image1)

Fig. 2 - *Right glomus tumor: long and transverse sections without contrast*

![Image of Right glomus tumor with contrast](image2)

Fig. 3 - *Paragangioma*

![Image of Paragangioma](image3)
Discussion
PGLs are neuroendocrine structures located in the adventitia carotid bifurcation, posterior-medial side. They are sensitive to hypoxia, acidosis and hypercapnia; their stimulation results in increased frequency and respiratory rate and a rise in blood pressure through vasoconstriction and the production of circulating catecholamines. The PGLs are rare neoplasms, with an incidence of 0.012% (2).
They generally occur between the fourth and fifth decade of life. The average size is of 4.5 cm x 3.5 cm x 3 cm but can grow up to 15 cm and a weight of 200 g. According to the Shamblin classification they are divided into three groups based on the size and connection with the surrounding vascular structures (group III belong to the larger and forms persistently adherent to the carotid bifurcation).
In most cases, the patients having a PGLs come to medical attention for the appearance of a mass in the lateral region of the neck, at the mandibular angle, often completely asymptomatic. When the tumor reaches considerable size, it may be responsible for nerve deficits (especially for X, XI and XII nerves), dysphagia, neck pain (3). Very rarely (5% of cases) the patients come to medical attention for the appearance of a "pressor amine syndrome" with tachycardia, palpitations, flushing, hypertensive crisis, dizziness, malaise, diarrhea, amino-linked activity consequences of PGLs secreting (4). The biological behavior of PLGs, characterized by their tendency to grow and gradually incorporate the surrounding vascular and nerve structures, justifies their complete removal whenever they are identified (5, 6).

Conclusions
Early diagnosis of PLGs asymptomatic and small, reduces the risk of perioperative lesions of the cranial nerves and carotid arteries (2).
Unfortunately, most of the PLGs, when identified, belong to group II or III Shamblin, making the operation much more difficult (7).
In our case, early identification was made possible by the biological and functional characteristics of the tumor and the consequent clinical manifestations, allowing to carry out the surgery safely for the patient. Despite the size of PLG taken into account, it was ranked to the Shamblin group I.
References

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