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Carotid paragangliomas: case report and imaging review

Running head: Carotid paragangliomas: cases and review

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Abstract

Background: Presentation of case reviews depicting the imaging characteristics of carotid paragangliomas, associated with a thorough analysis of the anatomical morphological features and the current therapeutic strategies.

Methods: We present the cases of three patients diagnosed with carotid paragangliomas in our clinic, illustrating diagnostic imaging elements by computer tomography (CT) and magnetic resonance imaging (MRI), but also the postoperative aspect of the carotid system, with respective anatomical, clinical and surgical considerations.

Results: The imaging aspect of the carotid paragangliomas is characterized by a mass of soft tissue with intense contrast enhancement and with “salt and pepper” MRI appearance on conventional spin-echo sequences. The postoperative evolution of the patients included in the article was favorable, without any perioperative complications or signs of local tumor recurrence.

Conclusions: Carotid paragangliomas are rare, often asymptomatic tumors, but with potential for increased malignancy, which raises the need for good knowledge of the cervical region pathology as well as the features of neuroendocrine tumors. CT and MRI examinations are essential for diagnosis, staging and, implicitly, for establishing the therapeutic strategy.

Key words: paraganglioma, carotid body tumor, carotid arteries, diagnostic tools, imaging, therapeutic strategy

Introduction

Carotid paragangliomas (CPs), also known as carotid body tumors, are neuroendocrine tumors originating in the parenchymal cells of the neuroectoderm and neural crests, but also in the mesodermal elements of the third branchial arch [1, 17, 19, 28, 30]. Thus, the cells that make up paragangliomas are similar to the cells in the APUD (amine precursor uptake and decarboxylation) system and can release catecholamines, cholecystokinin, serotonin, somatostatin, and vasoactive intestinal peptide [20].

Paragangliomas have variable localization and may develop in the head, neck, thorax, or abdomen. Head and neck paragangliomas can show the following topography: carotid (carotid glomus), tympanic (glomus tympani), jugular (glomus jugulare) or vagal (glomus vagale) [22]. These tumors are rare [1, 26, 30], can be single or multifocal tumors, and are considered benign but with malignant potential in 6-12% of cases [14, 19, 28, 34]. CPs represent 0.6% of head and neck neoplasms, accounting for 60-70% of paragangliomas of the head and neck region, and make up 0.5% of tumors affecting the entire body [1, 14, 34].

The majority of paragangliomas are sporadic, however, in about 40% of cases, family history is demonstrated [5]. Hereditary tumors are more often multicentric and tend to occur earlier than sporadic forms, with a peak incidence at 30-35 years of age [18]. Familial paragangliomas exhibit autosomal dominant transmission and are commonly seen in patients with von Hippel-Lindau disease, type I neurofibromatosis, and type II multiple endocrine neoplasias [23].

The scope of this paper is to mark the essential diagnostic elements of carotid paragangliomas useful to the clinicians and surgeons in planning the therapeutic strategy. Starting from the presentation of three illustrative cases investigated in our clinic, we analyzed the specific literature data and detailed the most important elements involved in the management of these tumor types.

Case reports

Case 1. A 42-year-old woman was admitted to the hospital for a painless left cervical tumor measuring close to 3 cm, with a soft consistency, and mobile in the transverse plane. The ultrasound showed a submandibular hypoechogenic mass that displaced the internal carotid artery. The MRI examination revealed a hypervascular lesion with a "salt and pepper" appearance located in the left carotid space, at the terminal bifurcation of the common carotid artery, with suggestive characteristics for CP (Figure 1). The patient refused surgery, but 90 days later she returned determined to receive treatment. CT angiography confirmed a bright and rapidly enhancing mass located at the level of the left carotid bifurcation compatible with a diagnosis of a type II Shamblin CP (Figure 2). The chosen treatment course implied the surgical cure of the tumor with a favorable postoperative evolution, without incidents, and with no detectable tumor recurrence on imaging follow-up at 3 months and, subsequently, at 6 and 12 months.

Case 2. A 55-year-old woman with no particular pathological background was admitted with a right cervical mass showing slow dimensional progression within a year, with no other associated symptoms. The clinical examination and ultrasound confirmed the presence of a tumor with soft consistency adjacent to the right internal carotid artery. The

MRI and CT examinations revealed a type IIIa Shamblin CP, with a characteristic "salt and pepper" appearance at the level of the right carotid bifurcation, with a small extension to the submandibular space and the right postero-lateral wall of the pharynx (Figure 3). The patient showed favorable postoperative evolution with a good evolution and is periodically evaluated in our clinic, without complications or tumor recurrence at 3.5 years after surgery.

Case 3. A diabetic 46-year-old woman presented with a left-cervical mass manifested in the last two years, with a gradual increase in size and associating dysphonia at the time of hospitalization. The ultrasound examination suspected a CP by highlighting a hypoechogenic mass at the carotid bifurcation, which splays the external and internal carotid arteries, extending medially to the retropharyngeal space, anteriorly to the submandibular branch, and posteriorly through the soft prevertebral tissues. CT angiography established the diagnosis of a type IIIb Shamblin CP (Figure 4). No incidents occurred during the surgical procedure and intraoperative observations confirmed the inclusion and infiltration of the internal carotid artery. The follow-up examinations performed at 3 and 12 months did not reveal any complications or local tumor recurrence.

Imaging review of paragangliomas

Imaging is an essential part of the diagnostic protocol in the management of carotid glomus tumors. CT and MRI examinations using contrast mediums are useful in lesion detection, thus obtaining valuable information regarding the lesion size, the relationship with the adjacent anatomical structures, and the degree of vascularization. Additionally, blood flow downstream of the lesion can be ascertained, and the presence of collateral circulation, anatomic variants, and other synchronous paragangliomas in other regions of the body can also be demonstrated [33].

Ultrasonography (US) is a first-line imaging method for determining the location and features of lateral-cervical masses. CP appears as a well-delimited round-oval hypoechogenic mass located at the level of the carotid sinus, causing the carotid bifurcation to flare. Color Doppler or Doppler Duplex highlights the hypervascular character of the

lesion and allows differentiation from other tumors, but with limited possibilities in detecting local invasion [15].

Computed Tomography (CT) shows CP as a soft tissue mass located in the carotid space, with intense and homogeneous early enhancement due to its highly vascular nature. Large tumors may exhibit a heterogeneous structure due to the presence of thrombi or focal hemorrhages. Flaring (splaying) of the external and internal carotid arteries, a hallmark of PC, can also be easily visualized on CT (Figure 5). Irradiation and the allergic risk to the iodinated contrast substance are the main disadvantages of CT [32].

Magnetic Resonance Imaging (MRI) has proven its superiority to other imaging techniques in offering a more accurate appreciation of lesion margins and invasion of adjacent structures [24]. Using native and contrast-enhanced spin-echo sequences with fat suppression, MRI examination can determine vascular invasion more accurately compared to CT, and can also provide more detailed vascular morphometry measurements [9]; additionally, MRI can reveal lesions smaller than 5 mm, while CT usually depicts lesions larger than 8 mm [31]. The typical MRI aspect of CP is given by the presence of multiple punctiform or serpentine shaped intralesional foci with signal void. These are caused by the higher flow velocity in the intratumoral vessels and give the whole lesion a "salt and pepper" appearance (Figure 6). The "salt" represents areas of hyperintensities due to slow vascular flow or microhemorrhages, while the "pepper" corresponds to the above-mentioned areas with markedly decreased T1 and T2 signals (signal void) [15]. This imaging pattern is rarely found in lesions larger than 1 cm, and can also be observed in other hypervascular tumors such as renal cell carcinoma metastases and thyroid carcinoma metastases [12].

Conventional digital subtraction angiography is the gold standard for evaluating the vascular architecture of these tumors. The typical angiographic aspect of paragangliomas is that of a hypervascular mass which splays the internal and external carotid arteries and presents large feeding arteries, intense arterial flash, and quick contrast washout. The tributary arteries of CPs are the ascending pharyngeal artery and the ascending cervical artery. As the tumor increases in size, contributions from other arterial sources such as the

facial, lingual, or thyroid arteries may add up. It is mandatory to note the patency of the internal jugular vein, which may be thrombosed in large CPs [6].

Differential imaging diagnosis of paragangliomas should include nerve sheath tumors that displace the carotid arteries medially and anteriorly while the internal jugular vein is pushed posteriorly, as compared to CPs that splay the carotids; other diagnoses include vagal glomus tumors which display similar morphological characteristics but are more rostrally located, and also hypervascular metastatic adenopathies from renal or thyroid malignancies, which are difficult to distinguish from CPs [6, 32]. Histopathology is not necessary for the diagnosis and performing a biopsy may even be very dangerous due to the high degree of tumor vascularization [21, 29].

Discussions

Carotid paragangliomas generally affect women [1, 19, 28, 30, 34], are bilateral in about 10% of cases [6], and the involvement of hereditary factors is described in 4-9% of cases [28, 30, 34].

From a clinical standpoint, CP are slow-evolving tumors which may remain painless and generally asymptomatic and for a long period of time, and are located anteriorly to the lateral sternocleidomastoid muscle, showing mobility in the lateral plane, but fixed in the cephalocaudal plane (Fontaine's sign) [31]. CPs significantly alter the loco-regional anatomy, both through their dimensions and their position. Therefore, any lesion that develops adjacent to the terminal bifurcation of the common carotid artery will induce changes in the muscular, vascular, and nervous anatomical relations. Taking into account the vascular nature of the lesion in question, the most apparent altered index could be the angle between the origins of the internal and external carotid arteries. As carotid body tumors grow in size they can invade the parapharyngeal space causing a bulging of the lateral oropharyngeal wall inducing dysphagia, odynophagia, or syndromes associated with cranial nerves IX-XII disorders [35].

Carotid body tumors are paragangliomas that show high malignant capacity, a feature that occurs in 6-12% of cases [14, 19, 28, 34]. In the absence of specific

histopathological criteria, malignancy is marked by the anatomical presence of metastases [10], with a predilection for regional lymph nodes, and very rare distant metastases [3]. The therapeutic method of choice for carotid body tumors is surgical removal, which in select cases may be preceded by angiographic embolization [8, 25]. Embolization may reduce the size of the tumor and promote its disconnection from the vascular system, thereby reducing intraoperative bleeding [13, 36]. Among the significant complications of surgical treatment are secondary bleeding, cranial nerve deficiency, and perioperative stroke; therefore, the intervention requires caution in aged patients, especially in patients with cardio-ischemic pathology despite the general addressability of the surgery [7, 8, 13, 25, 36].

The key element in choosing the right therapeutic conduit is the assessment of the extent of tumoral vascular involvement through medical imaging, a method that also provides insight regarding the prognosis and possible complications. As such, a classification system for CP was proposed by Shamblin and collaborators in 1971 and is currently in use despite its shortcomings regarding the degree of tumor infiltration into the carotid wall, a critical element to be factored when the preservation of the arteries is considered. According to Shamblin, type I tumors are located at the bifurcation of the common carotid artery, with minimal contact area with blood vessels, type II encompasses tumors that have a diameter generally less than 5 cm, and include about 50% of the circumference of the main arterial axis, entailing difficulties for the surgical cure, while type III is reserved for generally large tumors with a diameter greater than 5 cm, that completely enclose the main arterial axis [27].

Luna-Ortiz emphasizes that imaging by axial sections does not accurately approximate the carotid wall tumor infiltration preoperatively, but it highlights the circumferential ratio of the tumor to the adjacent vascular structures. Thus, in 2006, Luna-Ortiz and collaborators proposed the introduction of a grade IIIb to the carotid paragangliomas, which, regardless of their size, infiltrate the carotid wall. According to Luna-Ortiz, type IIIa is superimposed on the old (Shamblin) type III, respectively CPs that include the vessels without infiltrating them [16].

A study by Arya et al. sustains a good correlation between the radiological aspects of the Shamblin classification and surgical results, and concludes that without taking into account the tumor size, the maximum degree of circumferential tumor/artery contact should be the only criterion for the Shamblin classification, therefore predicting the degree of vascular viability [2].

Radiation therapy, alone or associated with surgery, is another therapeutic strategy that can be chosen in the case of carotid glomus tumors. The treatment is based on the induction of local fibrosis, which can stop tumor development and is indicated in inoperable cases or postoperative relapses [4, 11].

Conclusions

Carotid paragangliomas are hypervascular tumors, with imaging features of a soft tissue mass with intense enhancement and “salt and pepper” appearance on MRI in conventional spin-echo sequences.

The radiological examination is essential for diagnosis and the establishing of the therapeutic strategy. Firstly, the tumor must be detected and characterized, and secondly, the lesion extension must be appreciated in regard to the surrounding vascular structures. Last but not least, the presence of concomitant tumors should be verified, considering that paragangliomas are multifocal lesions in 30% of patients.

Considering all the information presented, the medical team managing a CP case should benefit from a mindful preoperative planning and a correct selection of patients, in order to obtain a successful therapeutic result.

Compliance with ethical standards: The study was performed in compliance with the local institutional research ethics committee and was carried out in accordance with the ethical standards of the Declaration of Helsinki and its later amendments.

Informed consent: Informed consent was obtained from all participants included in the study.

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Figure 1. MRI investigation consisting of (a) coronal Short Tau Inversion Recovery (STIR), (b) axial T2 fat-saturated WI, (c) sagittal T1 WI, and (d) gadolinium-enhanced axial T1 fat-saturated images depicting a inhomogeneous oval solid mass (star) with intermediate signal on T1WI and high signal on T2WI, intralesional foci in signal void that create an overall "salt and pepper" appearance, developed at the level of left carotid bifurcation; the lesion shows intense early enhancement, widens the angle of bifurcation of the common carotid artery and spreads the left external (ECA) and internal (ICA) carotid arteries, without altering their permeability and rapid blood flow signal.

Figure 2. CT examination: (a) axial CT angiography, (b) coronal-oblique maximum-intensity-projection (MIP) reformatted image, (c) curved-planar reformatted image of the left internal carotid artery, and (d) 3-D volume-rendered reformatted image. Large solid mass (star), with intense and slightly inhomogeneous enhancement, located at the level of the left carotid bifurcation, separating the external carotid (ECA) and internal carotid (ICA) arteries. The described lesion encases about 50% of the circumference of the internal carotid artery, without modifying its lumen (type II Shamblin) and is adjacent to the internal jugular vein (IJV). Macroscopic appearance (e) of the carotid body tumor after complete excision with a pseudo capsule (black arrow) and multiple feeding vessels (orange arrow). Microscopic specimen (f): uniform polygonal cells with abundant eosinophilic granular cytoplasm and large, regular, central nuclei.

Figure 3. MRI saturated T2 axial fat image (a) and gadolinium-enhanced T1 fat axial image (b), and axial CT postcontrast-arterial phase (c) showing a solid mass (star) with well-defined contour, inhomogeneous signal, "salt and pepper" aspect, and intense early contrast enhancement, located at the level of the right carotid bifurcation, separating the right internal (ICA) and external (ECA) carotid arteries. The lesion splays the carotid arteries, encloses the ICA and ECA without infiltrating them. Intraoperative view of the carotid body tumor before complete excision (d).

Figure 4. CT-angiography coronal-oblique volume-rendered images pre (a), (b), and postoperatively (c) highlighting a large hypervascular mass (star) at the level of the left carotid bifurcation that splays the carotid arteries and infiltrates the internal carotid wall, correspondingly to a type IIIb Shamblin tumor. Imaging performed one year after surgery demonstrates the absence of recurrence and normal appearance of the left carotid arteries. Intraoperative appearance of the carotid body tumor (d). Gross pathology specimen with a meaty appearance with a bulging surface (e). Photomicrograph of the histologic specimen (HE, x400) showing highly vascularized fibrous septa surrounding the chief neoplastic cells (f).

Figure 5. Contrast-enhanced CT: axial images (a) and coronal-oblique MIP (b) showing a widening of the distance between the left carotid arteries (black arrow) as a result of the development of a carotid body tumor (star). Left side vessels are marked: vertebral (VA), common carotid (CCA), internal carotid (ICA) and external carotid (ECA) arteries.

Figure 6. MRI aspect of a carotid body tumor with “salt and pepper” sign generated by signal void areas due to numerous intratumoral vessels (arrowheads); (a) T1 WI, (b) T2FS WI, (c) short tau inversion recovery (STIR) images.











