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Case Report of a Rare Tumor in the Head and Neck: Carotid Body Paraganglioma

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Abstract **Case Report**

Paragangliomas (PGLs) are tumors that are rarely malignant; the majority of them are benign. Similar to pheochromocytoma, they develop from the autonomic nerve system. This system originates from neural crest cells and can undergo neoplastic transformation. PGLs can arise either inside or outside the adrenal glands. Head and neck PGLs are very scarce [1]. The primary locations where this tumor commonly originates within this region are the carotid body, jugular bulb, and vagal body. In our case report, a 65-year-old man, who presented with a firm, painless, pulsatile neck mass that increased in size over the course of months. The diagnosis was suspected based on the patient's clinical history and physical examination. The diagnosis was confirmed with CT angiography (CTA).

Keywords: Paraganglioma, head neck tumors, carotid artery, CT, MRI.

INTRODUCTION

Paragangliomas of the head and neck are extremely rare tumors. Throughout a radiologist's professional career, they may encounter a few patients with these tumors. Therefore, it is important to be familiar with the main diagnostic signs when encountering such cases, as timely diagnosis and treatment can help reduce surgical risks.

The purpose of this review is to examine the most common localizations of paragangliomas in the head and neck and to describe the key visualization characteristics essential for accurate diagnosis and differential diagnosis.

Paraganglioma (PG) is a predominantly benign, slowly growing tumor that originates from neural crest cells. The paraganglionic system consists of structures dispersed throughout various organs and tissues in the human body in the form of clusters, playing an important role in homeostasis. Based on localization, two main groups of PG are identified. The largest group is found in the head and neck region, where neuroendocrine clusters are densely located. These structures are primarily situated in the adventitial tissue of arteries and veins and are typically closely associated with the parasympathetic nervous system, functioning as chemoreceptors. Most tumors are non-functional in terms of catecholamine secretion. PG located lower in the body most often originate from the adrenal medulla (pheochromocytomas), are generally associated with the sympathetic nervous system, and are capable of producing catecholamines.

CASE PRESENTATION

We present the case of a 65 year-old female with a 15-year history of non-insulin-dependent type II diabetes mellitus being treated with metformin 750 mg twice daily. presented to the clinic with a newly discovered mass on the right side of his neck. The patient first noticed the mass a few months prior and noted that it has been progressively increasing in size since he first noticed it.

The patient denied any recent illness, pain, discomfort, neck stiffness, difficulty hearing or swallowing, tinnitus, fever, weight changes, shaking chills, dizziness, fatigue, diaphoresis, palpitations, nausea, vomiting, diarrhea, or constipation.

On examination, the patient was conscious with normal vital signs, well-oriented in time and space, and demonstrated hemodynamic and respiratory stability.

On physical examination, a mass was palpated on the right side of the patient's neck near the angle of the mandible. The mass was firm, slightly mobile side-to-side, and pulsatile. The remainder of the exam was unremarkable.

CT angiogram of the head and neck found a large, hypervascular, hyperenhancing mass at the right carotid bifurcation which caused splaying of the internal and external carotid arteries. This mass measured 6,4x 7,6x4,4 cm in anterior-posterior, lateral, and craniocaudal measurements and was diagnosed to be a paraganglioma. No adenopathy was noted and the thyroid gland appeared normal.

MRI found a well-defined lesional process centered on the left carotid glomus, displacing and enlarging the angle formed by the internal and external carotid arteries, presenting heterogeneous hyperintensity on T2 and traversed by serpentine structures with hypointense "Flow Void" signal, suggesting vascularity. There is heterogeneous diffusion hyperintensity with areas of ADC restriction and intense heterogeneous enhancement after contrast agent injection, delineating central zones of necrosis.

The patient then had his urine collected to assess for free catecholamines, VMA, and metanephrines all of which were found to be within normal limits.

The patient then underwent a CT scan with IV contrast of his thorax, abdomen, and pelvis to assess for metastatic spread. The CT scan found no discrete metastatic disease.

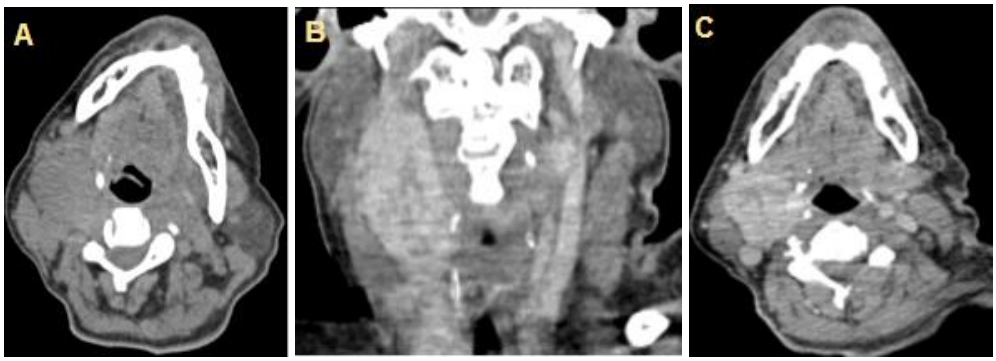

Figure 1: CTA of the neck demonstrating a right neck mass splaying the internal and external carotid arteries known as a Lyre sign. A: Contrast CT. B, C: Non-contrast CT

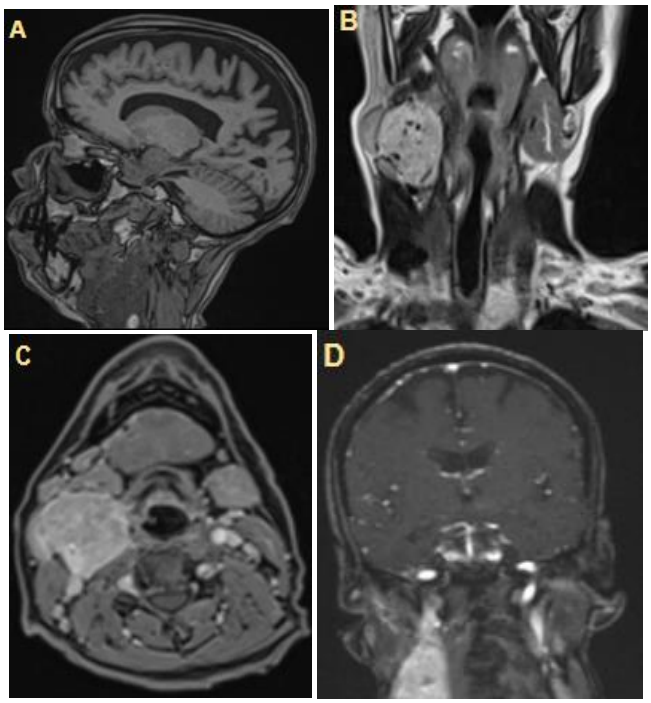

Figure 2: Magnetic resonance imaging (MRI) of the brain showing right glomus carotid lesion in different phases of acquisition T1 and T2 without contrast agent injection (A, B), T1 with contrast agent injection (C), TOF sequence (D). Vascular flow void present within the lesion

DISCUSSION

Paragangliomas are predominantly benign tumors characterized by slow growth, averaging about 0.2 cm per year. However, malignant cases do occur, with such forms reaching a frequency of up to 4%. It is challenging to predict the risk of adverse progression based on radiological or histological data. Reliable signs of malignancy include the presence of invasive growth and metastases, which typically involve the cervical lymph nodes, and less commonly the lungs, liver, and bones. Regional and distant metastases may be diagnosed simultaneously with the primary tumor or may emerge at various times after resection. Research by K. Papaspyrou *et al*., (2012) indicates a strong association between malignant forms and the presence of genetic abnormalities in patients [6].

As mentioned earlier, carotid body paragangliomas are the most common form of paragangliomas in the head and neck, accounting for over 50% [1]. The tumor develops from the carotid body, located in the adventitia at the bifurcation of the common carotid artery (CCA). When large, it may closely contact the lateral wall of the pharynx, trachea, vagus nerve, glossopharyngeal nerve, thyroid gland, and internal jugular vein [4].

The first mention of the carotid body was made by Von Haller in 1743, and in 1962, Heller introduced the concept of "glomus tumor." The term "paraganglioma" was introduced by Kohn in 1903, and due to the work of G.G. Glenner and P.M. Grimley in 1974, this term is now the most preferred for describing tumors of the carotid body, owing to the anatomical and physiological characteristics of the tumor [4, 9].

In any method of investigation, carotid body paragangliomas appear as hypervascular masses located at the bifurcation of the common carotid artery (CCA), displacing the internal carotid artery (ICA) posterior-laterally and the external carotid artery (ECA) anterior-medially, and extending cranially from the bifurcation area [8].

In smaller tumors, there may be no hemodynamically significant impact on the vessel lumen, but larger tumors can significantly deform the vessel's path or narrow its lumen (in cases of cuff-like tumor growth), leading to characteristic ultrasound findings and Doppler changes. Duplex scanning typically shows asymmetry in blood flow velocity, with an increase on the affected side compared to the opposite side, which can exceed a 60% increase [3, 4].

Computed Tomography (CT) allows for the assessment of the size, density, and location of the mass, as well as evaluating the degree of involvement of the carotid arteries and bony structures. The arterial phase is crucial for the differential diagnosis of carotid body paragangliomas from other tumors in the bifurcation of the carotid arteries, characterized by early and pronounced uptake of the contrast agent (ranging from 104 to 250 Hounsfield units, with an average of 174 ± 51 Hounsfield units) [7, 4].

The arterial phase also helps to clearly define the lower pole of the tumor (typically at the level of the CCA bifurcation or 0.5–1.5 cm below) and the distance from the tumor to the base of the skull, as well as the relationship between the tumor and the bifurcation and internal carotid artery. Expansion of the CCA bifurcation is a typical sign of carotid paraganglioma. In larger tumors, contrast enhancement may be heterogeneous due to areas of hyalinization, focal thrombosis, or hemorrhage. Based on CT data, 3D reconstructions can be created, visually illustrating the relationship of the tumor with the carotid arteries [5].

The classic "salt-and-pepper" sign of chemodec tumors on MRI was first described by W.L.G. Olsen *et al*., in 1987. This sign is due to flow voids in the large intratumoral vessels: a hypointense component resembling "pepper" alternates with hyperintense areas resembling "salt" (due to slow blood flow or hemorrhages) and is observed in all magnetic resonance sequences [4].

On T1-weighted images, the tumor typically has an iso-intense signal compared to the muscles, while on T2-weighted images, it is predominantly hyperintense. Post-contrast images show significant accumulation of the contrast agent [4].

G. Gravel *et al*., suggest that using the 3D angio-MR mode during the arterial phase of contrast enhancement is sufficient to detect paragangliomas of any localization, and the addition of post-contrast T1-weighted images provides additional topographic information that is useful for planning surgical treatment or radiotherapy [7].

Modified Shamblin Classification of Carotid Paragangliomas

The modified Shamblin classification categorizes carotid body paragangliomas (CBPs) based on their relationship with the carotid arteries and the degree of infiltration. This classification is important for assessing surgical risk and planning [5].

Type I	The tumor is non-adherent to the carotid artery. It displaces the carotid vessels laterally without invading the vessel wall.
Type II	The tumor is adherent to the carotid artery but does not infiltrate the wall. It may encase the vessel but does not invade it.
Type II	The tumor infiltrates the carotid artery wall. This type poses a higher surgical risk due to the potential for significant vascular involvement.

According to a meta-analysis by Z. D. Guss *et al*., (2011), radiation therapy is recommended as a primary treatment method for jugular paragangliomas (JPGs) due to their more aggressive nature and higher recurrence rates following surgical resection. This approach aims to control tumor growth and minimize symptoms in patients who may not tolerate surgery well [8].

After resection of neck paragangliomas, regular follow-up care is recommended to monitor for potential recurrences and metastases. This proactive approach enables timely detection and management of any new developments related to the tumor [4].

CONCLUSION

Paragangliomas (PGLs) of the head and neck are relatively rare in the population. Carotid body tumors are the most common form in this location, followed by vagal and jugular paragangliomas. Multiple paragangliomas are often associated with a higher frequency of familial forms. These tumors can have a prolonged asymptomatic period, but significant growth may lead to compression of surrounding vascular and nerve structures, the larynx, bones, and other organs in the neck, ultimately diminishing the patient's quality of life [4].

The primary diagnostic sign is the identification of a hypervascular mass in the region where paraganglia accumulate. This is evidenced by changes in duplex ultrasound findings, pronounced contrast enhancement in the arterial phase on CT or MRI, and the classic MRI symptom described as "salt and pepper" [4].

Paragangliomas are considered malignant when local (to lymph nodes) or distant metastases are present [1].

Understanding the key characteristics of paragangliomas enables timely and accurate diagnosis, facilitating effective treatment planning.

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