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International Journal of Clinical Case Reports and Reviews
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Case Report

Tongue Deviation as a Manifestation of Paraganglioma

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Received Date: December 18, 2024 | **Accepted Date:** January 23, 2025 | **Published Date:** February 01, 2025

Citation: Leite L. Marques, Silva Sara, Ribeiro Elisabete, Bessa I. Mendes, (2025), Tongue Deviation as a Manifestation of Paraganglioma, *International Journal of Clinical Case Reports and Reviews*, 23(1); **DOI:10.31579/2690-4861/687**

Abstract:

Cervical paragangliomas are typically of parasympathetic origin, asymptomatic until they cause compression of surrounding tissues and nerves. The presented clinical case represents a paraganglioma diagnosed after hypoglossal nerve palsy.

Early diagnosis and genetic testing allow for earlier treatment and individualized follow-up, as well as enabling the screening of family members and integrating them into an appropriate screening program.

Key words: neuroendocrine tumor; hypoglossal

Introduction

Paraganglioma is a neuroendocrine tumor originating from the paravertebral ganglia of the sympathetic nervous system or from the parasympathetic ganglia, most commonly in the vagus and glossopharyngeal nerves present in the neck and skull base. Paragangliomas are classified as catecholamine-secreting and non-catecholamine-secreting. Catecholamine-secreting paragangliomas have nonspecific symptoms, including paroxysmal hypertension, headache, tachycardia, diaphoresis, and anxiety. Parasympathetic paragangliomas, which are frequently non-secreting, present symptoms resulting from the mass effect on surrounding tissues and nerves.

Case report:

We present the clinical case of a 43-year-old woman with no relevant personal medical history, who had a progressive increase in a right lateral cervical swelling over the course of one year. She also presented with ipsilateral tongue deviation over the last 15 days. On physical examination, a mass was noted below the mandibular angle and right carotid triangle, measuring 5 cm x 3.5 cm, with irregular contours, firm consistency, adherent to the underlying tissues, non-tender on palpation, without warmth or redness. The tongue protruded with deviation to the right and fasciculations on the right side (figure 1). There was difficulty in tongue mobility, such as curling it and laterally moving it to the left.

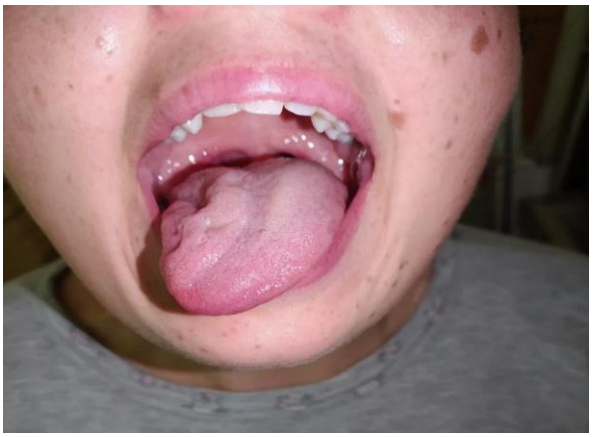

Figure 1: Tong deviation to the right and fasciculations on the right side (hypoglossal nerve palsy)

A computed tomography (CT) scan of the neck was performed, which showed the presence of an expansive, enhancing mass measuring 4.5 x 3 cm, centered in the carotid space, without a cleavage plane with the carotid artery or internal jugular vein. The mass had superior internal extension, partially occupying the parapharyngeal space and was inseparable from the right pharyngeal wall (figure 2).

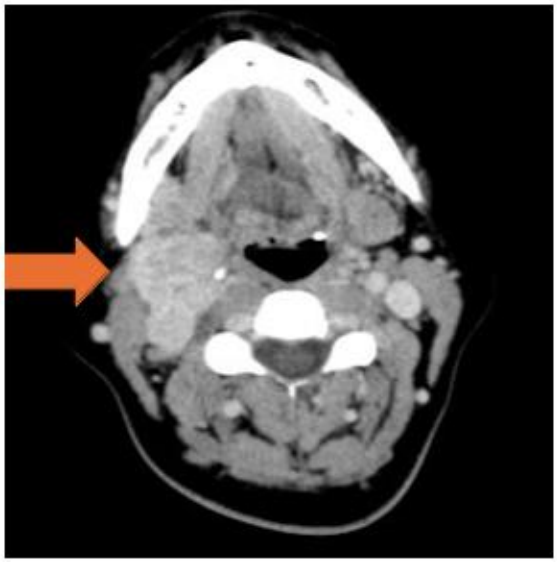

Figure 2: Mass centered in the right carotid space (orange arrow).

The cervical magnetic resonance imaging (MRI) revealed a mass measuring 4.5 x 5.6 cm, with a "salt-and-pepper" signal on T1 and T2, hypervascular, with probable origin from the carotid body. The internal jugular vein was obliterated by the lesion, and both the internal and external carotid arteries were surrounded by the mass.

Urinary and plasma metanephrines and catecholamines were negative, as well as vanillylmandelic acid and chromogranin A.

As a complementary study, a thoraco-abdominal-pelvic CT scan was performed, which showed multiple bilateral pulmonary nodular lesions, with a maximum diameter of 6 mm, suggestive of metastases. The Gallium-68-DOTA-TOC positron emission tomography (PET) scan revealed abnormal uptake with abnormal overexpression of somatostatin receptors in the right lateral cervical mass (image 3), in the pulmonary nodules, and in bilateral mediastinal-hilar lymph nodes.

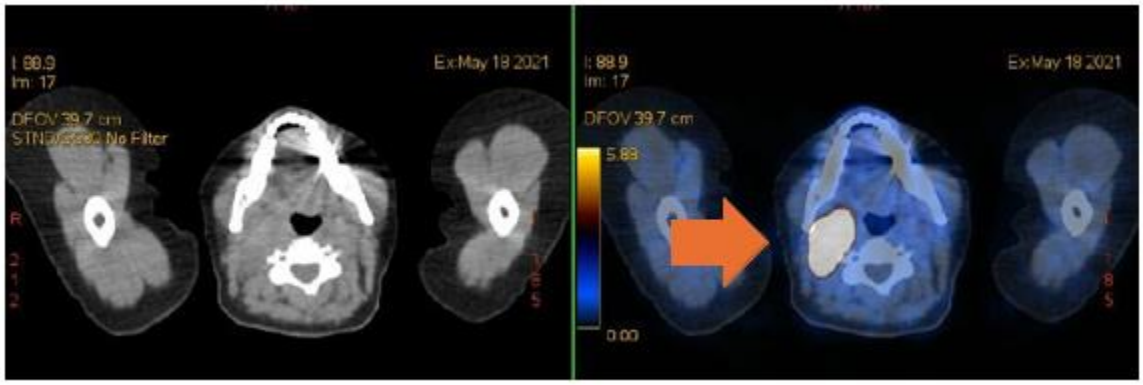

Figure 3: Abnormal uptake with abnormal overexpression of somatostatin receptors in the right lateral cervical mass in the Gallium-68-DOTA-TOC PET (orange arrow).

The patient was referred to the Oncology consultation with a diagnosis of metastatic non- secretory paraganglioma, with a positive genetic test for the SDHB mutation. Initially, she underwent partial embolization of the paraganglioma. She also started therapy with somatostatin analogs. After 4 years, she maintains clinical and radiological stability. During the follow-up period, a family genetic study was performed, where a brother tested positive for the SDHB mutation.

Discussion and conclusion:

A thorough clinical, biochemical, radiological, and morphological evaluation is essential in the diagnosis of paraganglioma. Genetic analysis has become particularly important in recent years due to its prognostic and therapeutic value. The SDHB mutation is associated with 40% of metastatic paragangliomas, indicating a worse prognosis and a higher risk of neoplasms. When this mutation is detected or a hereditary syndrome is established with more cases in the family, more regular screenings can be performed, leading to earlier diagnoses and treatments. [1]

References

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DOI:10.31579/2690-4861/687