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Radiologic Features of an Aggressive Metastatic Cervical Paraganglioma: A Case Report

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Abstract **Case Report**

Head and neck paragangliomas are rare neuroendocrine tumors with potential for metastasis, which defines malignancy according to current WHO classification. A 37-year-old woman presented with altered general condition and dysphonia. Imaging revealed a hypervascular mass at the left carotid bifurcation with vascular encasement, necrosis, and multifocal cervical lesions. Nodal involvement and compression of the internal jugular vein were observed. Thoracoabdominal imaging demonstrated pulmonary and hepatic metastases. Histopathology confirmed metastatic paraganglioma. Aggressive imaging features such as large size, necrosis, vascular encasement, and multifocality should raise suspicion for malignancy. Imaging is essential for staging and detection of metastatic disease.

Keywords: Paraganglioma; Carotid body tumor; MRI; Metastasis.

INTRODUCTION

Paragangliomas (PGLs) are rare neuroendocrine neoplasms arising from extra-adrenal paraganglia derived from neural crest cells. In the head and neck region, they most commonly originate from the carotid body, jugulotympanic region, or along the vagus nerve pathway. These tumors are typically slow-growing, highly vascular, and often clinically indolent [1,2].

According to the 2022 World Health Organization (WHO) Classification of Endocrine and Neuroendocrine Tumours, all paragangliomas possess metastatic potential. Importantly, malignancy is defined exclusively by the presence of metastases in anatomical sites devoid of normal paraganglionic tissue, rather than by histopathological features [1].

Imaging plays a pivotal role not only in tumor detection and characterization but also in locoregional staging and identification of distant metastatic disease. We report a case of an aggressive cervical paraganglioma with widespread thoraco-abdominal metastases, emphasizing radiological features suggestive of malignant behavior.

CASE PRESENTATION

A 37-year-old female patient initially presented with constitutional symptoms characterized by altered general condition and persistent dysphonia, evolving over approximately one year.

A cervico-thoraco-abdomino-pelvic computed tomography (CT) scan was performed as first-line imaging, revealing two left cervical masses associated with multiple secondary lesions, raising suspicion for either metastatic lymphadenopathy or a primary tumor of the carotid space.

Subsequent magnetic resonance imaging (MRI) of the neck demonstrated a large hypervascular mass centered at the left carotid bifurcation. The lesion caused splaying of the internal and external carotid arteries (classic “lyre sign”), exhibited heterogeneous intermediate T2 signal intensity, diffusion restriction, and intense peripheral enhancement with central necrosis. There was circumferential vascular encasement with luminal narrowing but no complete arterial occlusion. The ipsilateral internal jugular vein was compressed with absence of detectable flow.

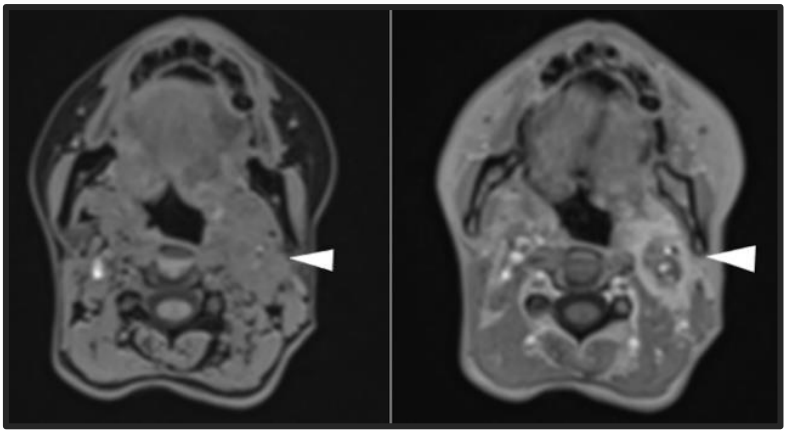

Figure 1. Cervical MRI—Axial T1-weighted images before (right) and after (left) gadolinium administration
Left carotid body paraganglioma demonstrating circumferential vascular encasement and central necrosis.

Tumoral extension into adjacent parapharyngeal and paralaryngeal spaces was observed. A second lesion with similar imaging characteristics was identified in the ipsilateral infrahyoid carotid space, suggesting multifocal disease. Multiple ipsilateral cervical lymph nodes (levels IB, III, and supraclavicular) showed necrosis and restricted diffusion, consistent with nodal involvement. No intracranial extension was detected.

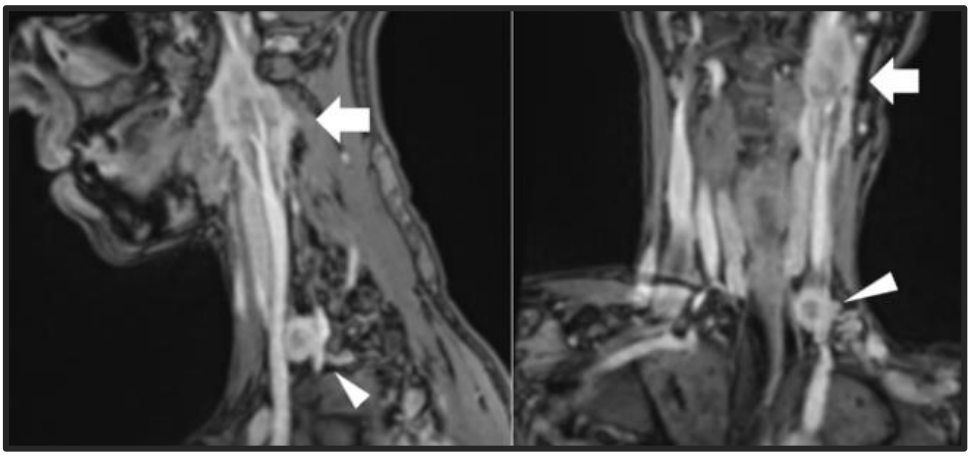

Figure 2. Cervical MRI—Sagittal (right) and coronal (left) post-contrast T1-weighted images
Tumor (arrow) extension into the parapharyngeal and paralaryngeal spaces, with a second enhancing lesion (arrowhead) in the left infrahyoid carotid space, consistent with multifocal disease.

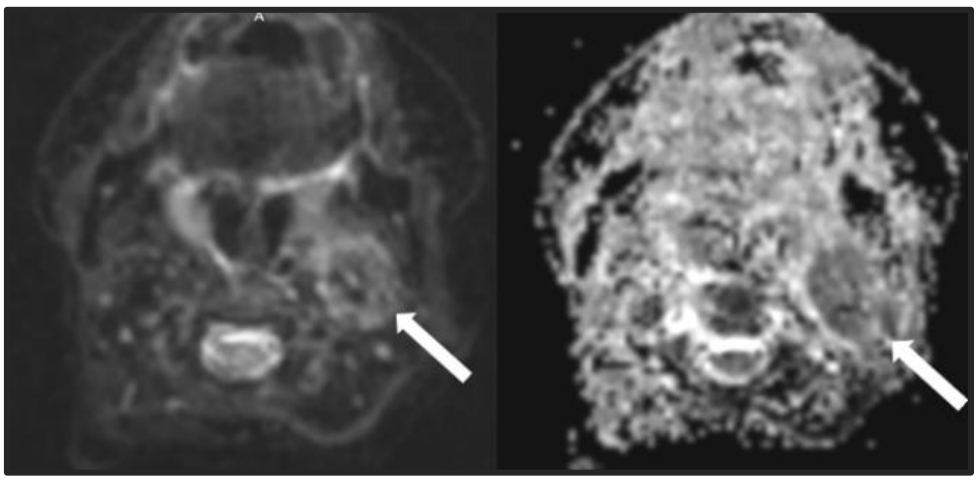

Figure 3. Cervical MRI—Diffusion-weighted imaging (DWI)
DWI shows hyperintensity of the primary lesion (arrow) with corresponding low ADC signal, consistent with diffusion restriction and high cellularity.

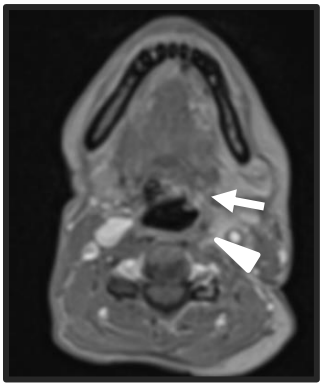

Figure 4. Cervical MRI—Axial T1 post-contrast image (lymph node)
Left carotid body paraganglioma (arrowhead) associated with ipsilateral cervical lymphadenopathy (arrow), consistent with metastatic nodal involvement.

Thoracoabdominal CT revealed multiple bilateral pulmonary nodules and masses, along with numerous hepatic lesions consistent with metastatic dissemination.

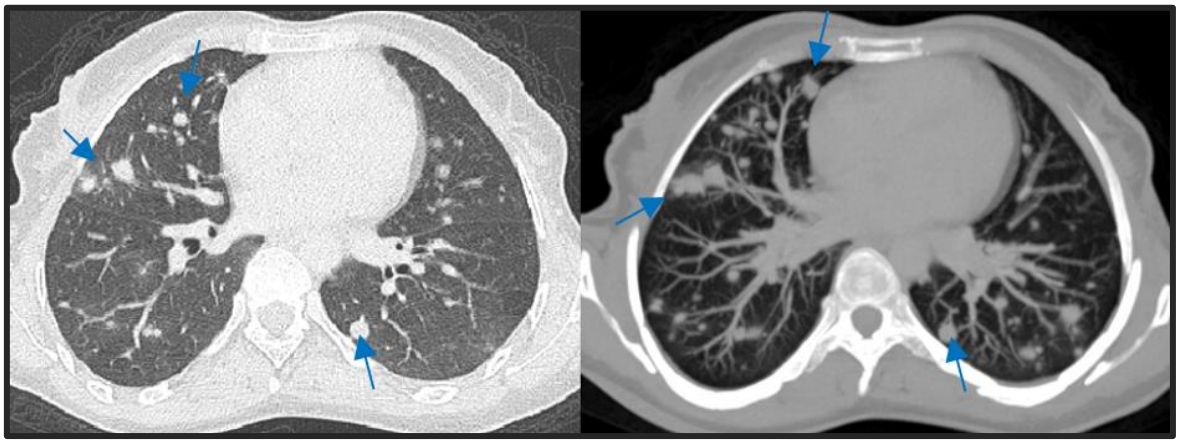

Figure 5. Thoracic CT—Axial parenchymal window images (right) and MIP reconstructions
Multiple bilateral pulmonary nodules and micronodules (arrows) of varying sizes, randomly distributed, consistent with hematogenous metastatic dissemination

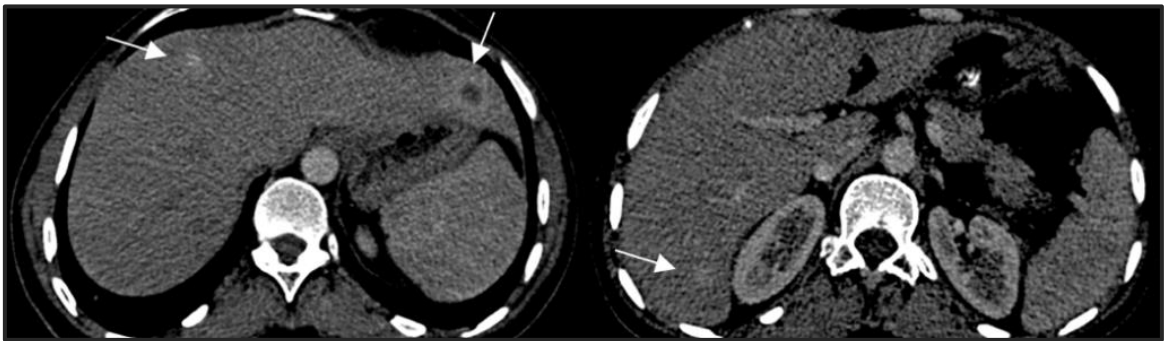

Figure 6. Abdominal CT (Portal venous phase)
Multiple hepatic lesions with heterogeneous enhancement (arrows), consistent with metastases

Histopathological examination demonstrated the characteristic *zellballen* architecture. Immunohistochemistry was positive for chromogranin A and synaptophysin, confirming the diagnosis of metastatic malignant paraganglioma.

DISCUSSION

Carotid body paragangliomas represent the most common subtype of head and neck paragangliomas. They arise from paraganglionic chemoreceptor cells located at the carotid bifurcation. Although generally indolent, these tumors are highly vascular and may exhibit locally aggressive growth patterns [1–3]. The estimated annual incidence ranges from 0.03 to 0.1 per 100,000 individuals [2].

MRI is the imaging modality of choice for diagnosis and locoregional assessment. The hallmark feature is a well-defined mass centered at the carotid bifurcation, producing separation of the internal and external carotid arteries, known as the "lyre sign," which strongly suggests carotid body origin [3].

On T2-weighted imaging, paragangliomas typically demonstrate hyperintense or heterogeneous intermediate signal intensity. Larger lesions may exhibit the characteristic "salt-and-pepper" appearance due to the presence of multiple intratumoral flow voids and areas of hemorrhage [3,4]. After gadolinium administration, intense enhancement is typically observed, reflecting marked tumor vascularity, although necrotic areas may result in heterogeneous enhancement patterns.

Diffusion-weighted imaging may show restricted diffusion due to high cellularity; however, these finding lacks specificity [4]. Vascular imaging using MR angiography or CT angiography is essential for evaluating arterial encasement, luminal narrowing, and venous compression, all of which are critical for surgical planning and assessment of tumor aggressiveness [3,5].

Functional imaging has emerged as a highly sensitive modality for detecting multifocal and metastatic disease, especially in patients with hereditary syndromes [2,5].

Importantly, no reliable imaging or histopathological criteria can definitively distinguish benign from malignant paragangliomas. The diagnosis of malignancy relies solely on the identification of regional or distant metastases [1,6]. Metastatic spread occurs in approximately 5–10% of cases but is significantly more frequent in patients with SDHB gene mutations [2,5]. Common metastatic sites include lymph nodes, lungs, liver, and bone.

In the present case, several imaging features were suggestive of aggressive behavior, including large tumor size, central necrosis, vascular encasement, multifocality, nodal involvement, and the presence of distant metastases at diagnosis.

CONCLUSION

Although head and neck paragangliomas are typically benign and slow-growing, a subset may demonstrate aggressive behavior with metastatic dissemination. Imaging plays a crucial role not only in tumor detection and characterization but also in identifying features suggestive of malignancy, such as large size, necrosis, vascular encasement, multifocality, and nodal involvement.

Comprehensive staging, including cross-sectional and functional imaging, is essential for detecting metastatic disease, which remains the only definitive criterion of malignancy according to current WHO classification. Early recognition of aggressive imaging features is critical for guiding management, prognostic assessment, and long-term surveillance.

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