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Double trouble at the carotid bifurcation: A rare case of bilateral carotid body tumors

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Abstract

Carotid body tumors (CBTs) are rare, highly vascular paragangliomas arising from chemoreceptor tissue at the carotid bifurcation. Bilateral tumors are particularly uncommon in patients without a known familial or genetic predisposition. We present a 54-year-old man with bilateral, long-standing, asymptomatic neck masses diagnosed as bilateral CBTs based on clinical findings and computed tomography angiography. After multidisciplinary discussion of staged surgery, radiotherapy, and active surveillance, the patient chose surveillance. The tumors remained stable at the 6-month follow-up. This case highlights the importance of careful clinical assessment, appropriate imaging, and individualized management to balance tumor control against the potential morbidity of bilateral intervention.

Keywords: carotid body tumor, bilateral, paraganglioma, active surveillance, sporadic

Introduction

Carotid body tumors (CBTs), also known as glomus caroticum tumors or chemodectomas, are uncommon neuroendocrine neoplasms arising from paraganglionic cells at the carotid bifurcation. They account for approximately 0.6% of all head and neck tumors, and up to 10% are bilateral [1]. These indolent tumors constitute approximately 60%-70% of cervical paragangliomas, with an estimated incidence of one to two cases per 100,000 individuals [2]. Although CBTs are usually benign and slow-growing, approximately 5% may metastasize [3]. Advances in genetic testing have improved the understanding of paragangliomas, and pathogenic variants in genes encoding the succinate dehydrogenase (SDH) complex are frequently implicated [3]. Surgical excision remains the mainstay of treatment, although it is technically challenging because of the tumors' rich vascular supply and proximity to major neurovascular structures. This report describes bilateral CBTs in a patient with no reported family history or known genetic predisposition, highlighting the rarity and clinical complexity of this presentation and the importance of individualized management.

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Informed Consent
Written informed consent was obtained from the patient for publication of this case report and the accompanying images. All patient identifiers were removed to protect confidentiality.

Conflict of Interest
No conflict of interest was declared by the authors.

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Case presentation

A 54-year-old man with no known comorbidities presented to the outpatient clinic with a 10-year history of bilateral neck swellings that had remained stable in size. He reported no pain, dysphagia, hoarseness, dyspnea, weight loss, or other constitutional symptoms. He also had no history of trauma, previous neck irradiation, or a family history of similar swellings.

On examination, firm, nontender, pulsatile swellings were present bilaterally in the upper neck, immediately below the angles of the mandible (Figures 1 and 2). The right swelling measured approximately 3 cm in diameter, and the left measured approximately 4 cm. Both swellings were mobile horizontally but fixed vertically, consistent with Fontaine's sign. Cranial nerve examination findings were normal, and no cervical lymphadenopathy was detected.

Figure 1: Right-sided neck swelling below the angle of the mandible.

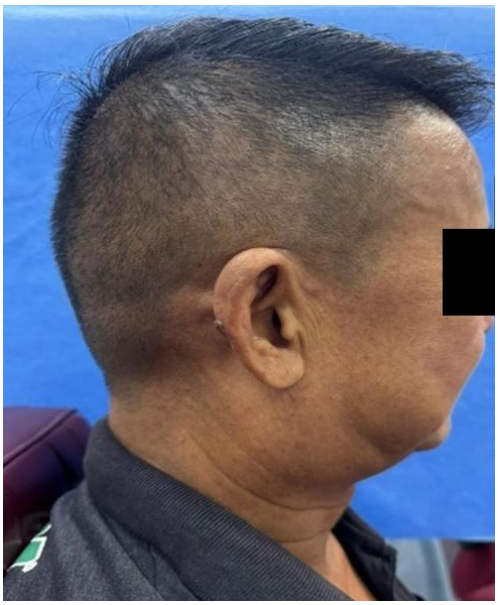
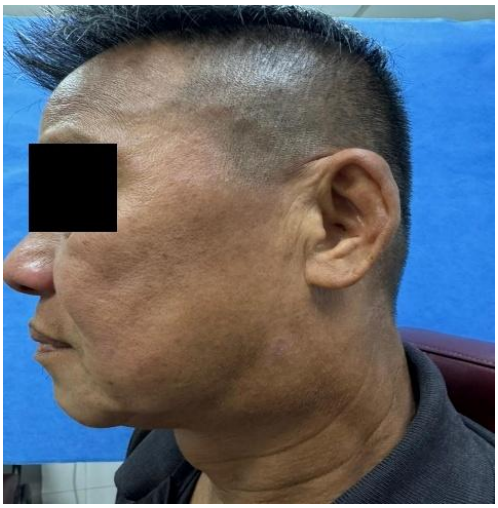


Figure 2: Left-sided neck swelling below the angle of the mandible.



Flexible endoscopy showed slight medialization of the left lateral pharyngeal wall at the oropharyngeal level. Both vocal cords were symmetrically mobile. Contrast-enhanced computed tomography angiography demonstrated intensely enhancing, highly vascular tumors at both carotid bifurcations. The tumors splayed the internal and external carotid arteries, producing the characteristic lyre sign (Figures 3 and 4). These findings were consistent with bilateral carotid body paragangliomas.

Figure 3: Coronal contrast-enhanced computed tomography image showing bilateral masses at the carotid bifurcations (arrows).

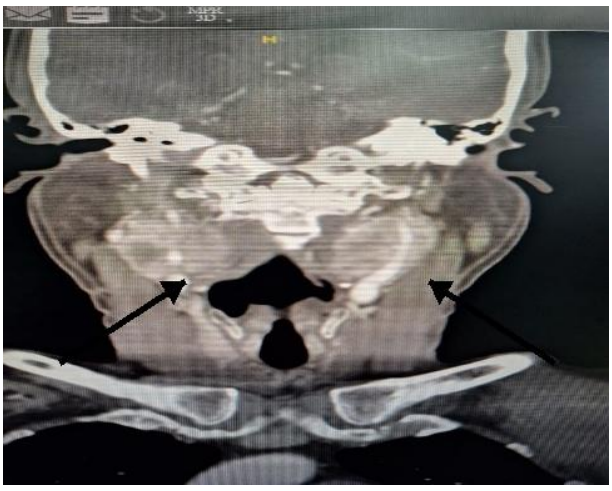
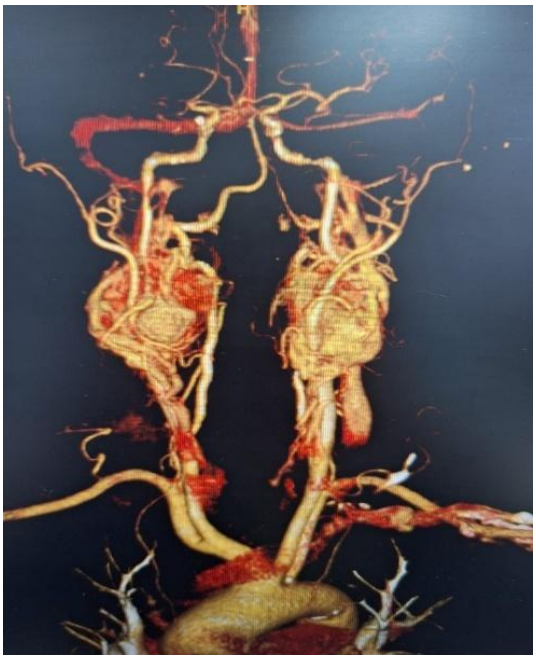


Figure 4: Computed tomography angiogram showing bilateral masses at the carotid bifurcations with splaying of the internal and external carotid arteries.



Given the long-standing, indolent course and absence of symptoms, a multidisciplinary team discussed several management options with the patient, including staged surgical resection, radiotherapy, and active surveillance. The potential risks of surgery, particularly the cumulative complications associated with bilateral intervention, were explained in detail. Considering the stable clinical course and absence of functional deficits, the patient chose conservative management with active surveillance. At the most recent six-month follow-up, magnetic resonance imaging (MRI) demonstrated no significant change in tumor size, and the patient remained asymptomatic.

Discussion

Bilateral CBTs are an uncommon subset of paragangliomas, accounting for approximately 3%-5% of cases and posing distinct diagnostic and management challenges [4, 5]. The reported incidence of CBTs varies, with earlier epidemiologic studies estimating approximately one case per 30,000 individuals [4]. Bilateral involvement occurs more frequently in familial disease, affecting up to 31.8% of patients with familial tumors compared with approximately 4%-5% of patients with sporadic tumors [6]. In this patient, the absence of a reported family history,

together with his age and prolonged indolent course, supports a presumed sporadic etiology.

Hereditary paraganglioma syndromes are associated with pathogenic variants in genes encoding subunits of the SDH complex, including SDHD, SDHB, SDHC, and SDHA. These variants are particularly associated with multicentric or bilateral disease [7]. Environmental factors, including chronic hypoxia in individuals living at high altitude, have also been implicated in carotid body hyperplasia and neoplasia [8].

Clinically, CBTs typically present as painless, pulsatile neck swellings at the carotid bifurcation that are mobile horizontally but fixed vertically, producing Fontaine's sign [9]. Patients with bilateral tumors may remain asymptomatic or develop symptoms related to mass effect; catecholamine secretion is uncommon. Imaging is central to diagnosis. Doppler ultrasonography, computed tomography angiography, and MRI are commonly used. Splaying of the internal and external carotid arteries produces the characteristic lyre sign on angiographic imaging [7, 10]. MRI may show a salt-and-pepper appearance caused by flow voids and hyperintense foci. Biochemical evaluation with plasma-free metanephrines or urinary catecholamine metabolites may help identify functional tumors.

The Shamblin classification assesses CBTs according to the extent of carotid vessel involvement and helps predict operative complexity and risk. Shamblin I tumors are small and minimally attached to the vessels; Shamblin II tumors partially surround the carotid arteries; and Shamblin III tumors extensively encase the vessels and are associated with greater operative difficulty and a higher risk of neurovascular complications [11]. This classification is therefore important for preoperative planning and shared decision-making.

Management of bilateral CBTs is complex and should be individualized. Surgical excision remains the conventional definitive treatment, whereas radiotherapy and active surveillance are appropriate alternatives in selected patients. Bilateral surgery carries substantial risks, including cranial nerve injury, stroke, and baroreflex dysfunction. When staged resection is planned, some clinicians prioritize the larger or more symptomatic tumor, whereas others operate first on the smaller tumor to reduce the immediate surgical risk [4]. Radiotherapy generally provides durable growth control while preserving cranial nerve function, although substantial tumor shrinkage is uncommon. Reported cranial nerve deficits after surgery range from approximately 20%-40%, whereas radiation-related deficits are generally less frequent and milder [12]. For selected complex tumors, a coordinated endovascular and surgical approach may facilitate treatment [13].

In this patient, the long-standing, asymptomatic tumors presented as pulsatile neck swellings with Fontaine's sign. The larger left-sided lesion produced medialization of the lateral pharyngeal wall and was considered a Shamblin III tumor on imaging. Because of the anticipated neurovascular and cranial nerve morbidity, immediate surgery was deferred in favor of active surveillance. Follow-up was planned with clinical assessment and serial MRI at 6-month intervals during the first year, followed by annual imaging if stability was confirmed. Indications for active intervention include documented tumor growth, new symptoms such as pain, dysphagia, or cranial nerve dysfunction, progressive carotid artery encasement, or newly detected biochemical activity.

This individualized strategy balances the morbidity of intervention against the typically indolent behavior of CBTs, particularly in older or asymptomatic patients. Evidence indicates that many head and neck paragangliomas grow slowly and can be monitored safely with structured follow-up [14]. In conclusion, bilateral CBTs should be considered in the differential diagnosis of chronic, slow-growing neck masses. Active surveillance is a reasonable evidence-based option for carefully selected patients with long-standing, asymptomatic tumors and substantial operative risk. Multidisciplinary assessment and shared decision-making are essential, and longer-term studies are needed to refine criteria for surveillance and intervention in bilateral disease.

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