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# International Journal of Surgery Case Reports

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Case report

# Retropharyngeal carotid body tumor: A challenge in terms of surgical technicality

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#### ARTICLE INFO

# Keywords: Retropharyngeal Carotid body tumor Surgical resection Head and neck neoplasm Paraganglioma Anatomic location Case report

#### ABSTRACT

*Introduction and importance:* Carotid body tumors are rare neoplasms originating from neural crest cells, commonly presenting as a painless, slow-growing mass in the lateral neck. The retropharyngeal variant of these tumors is particularly uncommon, with an incidence of approximately 2.6 %.

Case presentation: A 50-year-old man presented to the Otolaryngology Department at Rasoul Akram Hospital with a painless  $3 \times 4$  cm mass on the right side of his neck, which had gradually increased in size. Clinical examination revealed a pulsatile, nontender mass exerting pressure on the right lateral aspect of the pharynx. A CT scan showed a well-defined, enhancing retropharyngeal mass located between the internal and external carotid arteries on the right side, suggestive of a carotid body tumor. The patient underwent surgical resection, achieving complete tumor removal without complications. A four-year follow-up revealed no signs of tumor recurrence. Clinical discussion: Carotid body tumors are indolent masses. Diagnosis typically involves clinical evaluation, supplemented by imaging techniques such as ultrasound, CT scan, and MRI, which aid in delineating the tumor's morphology and dimensions. Surgical resection remains the preferred treatment, aiming to achieve complete tumor excision while preserving blood flow, avoiding damage to critical organs and nerves, and safeguarding brain function.

Conclusion: The retropharyngeal location of carotid body tumors represents an exceptionally rare variant, and their surgical resection presents a significant challenge for surgeons. This case report offers crucial insights into the surgical management of a retropharyngeal carotid body tumor, serving as a valuable resource for future surgeons who may face similar cases.

#### 1. Introduction

Carotid body tumors are infrequent neoplasms of the head and neck that originate from neural crest cells. These tumors were first documented by von Haller in 1743 [1]. They generally present as a painless, slow-growing mass in the lateral neck region. Based on their relationship with surrounding vessels, they are classified into three Shamblin groups: Group I consists of tumors located between the internal and external carotid arteries without attachment to the vessels; Group II includes tumors that partially attach to the vessels; and Group III tumors completely encase the carotid vessels [1].

The preferred treatment for these tumors is surgical resection. Given their critical and complex anatomic location, surgery poses significant challenges. Although preoperative embolization is sometimes used due to the tumors' extensive vascularity, it has been associated with

neurological complications and increased inpatient mortality due to blood loss. Consequently, surgical resection remains the gold standard [2,3].

These tumors typically occur at the carotid artery bifurcation, around the C3 and C4 levels, superior to the thyroid cartilage, within the carotid sheath, and lateral to the foramen transversarium. However, anatomical variations exist; for instance, the retropharyngeal location of these vessels, identified since 1920 [2,3], is one such variant with a reported incidence of about 2.6 % [4]. While carotid body tumors are rare in themselves, the occurrence of such a tumor in a retropharyngeal carotid artery is exceedingly rare [1–4]. This article presents a documented case of a retropharyngeal carotid body tumor. We ensure that this study was reported in compliance with the SCARE 2023 criteria [5].

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#### 2. Case presentation

The patient is a 50-year-old man referred to the otorhinolaryngology clinic with a complaint of a painless mass on the right side of the neck. The mass has been present for an extended period and has gradually enlarged. Upon detailed examination, a mass measuring  $3\times 4$  cm was noted in the neck region below the mandible. Palpation revealed that the tumor was pulsatile, nontender, and mobile only in the horizontal plane. The patient reported a sensation of globus, but did not experience dysphagia, odynophagia, or dysphonia. Auscultation revealed a bruit over the mass.

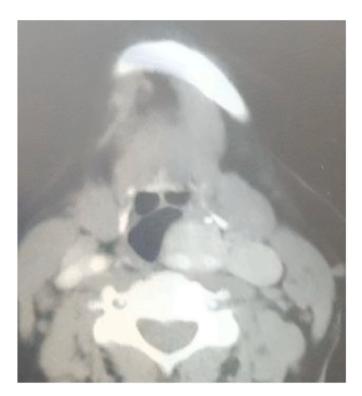
A clinical diagnosis of carotid body tumor led to the request for a cervical CT scan with intravenous contrast. The scan revealed a well-defined, enhancing retropharyngeal mass between the internal and external carotid arteries on the right side. This mass exerts pressure on the right lateral pharynx, consistent with a carotid body tumor. Notably, the common carotid artery, along with its bifurcation site and both the internal and external branches, was found to be positioned retropharyngeal (Figs. 1, 2).

## 3. Surgical technique

Following the diagnostic tests, the patient was diagnosed with a retropharyngeal carotid body tumor and was prepared for surgical excision. Informed consent was obtained prior to the procedure.

Given the novel surgical approach for this type of tumor, the initial stages of the surgery presented significant challenges. We opted to commence with a traditional surgical approach, while remaining prepared to modify the strategy if necessary.

The patient was placed under general anesthesia. During intubation, a bulge on the right side of the pharynx, which narrowed the airway, was observed. However, intubation proceeded smoothly to avoid disturbing the tumor. Cerebral and cardiac monitoring were implemented throughout the operation. The patient was positioned supine with the



**Fig. 1.** Computed tomography angiography (CTA) of the neck with contrast, axial section, shows an enhancing mass in the right side of the neck with mass effect on the right airway.

head rotated to the contralateral side. A transverse incision was made along the standard line of the sternocleidomastoid (SCM) muscle on the lateral neck. The flap was elevated superiorly to the hyoid bone within the subplatysmal plane, and the strap muscles along with the SCM were dissected to reveal the major vascular structures. The vagus and hypoglossal nerves were identified and marked. We then carefully dissected the common carotid artery in the subadventitial plane, exposing the bifurcation and the retropharyngeal carotid body tumor. The tumor, situated between the internal and external carotid arteries on the right side, exerted pressure on the right lateral pharynx. It was free from extension or adherence to surrounding tissues and was meticulously resected (Figs. 3, 4). To minimize intraoperative bleeding, hypotensive anesthesia was employed. Postoperatively, the patient experienced no complications and was discharged in stable condition. A four-year follow-up revealed no evidence of tumor recurrence.

#### 4. Discussion

Carotid body tumors are neoplasms originating from the paraganglia at the carotid arteries' bifurcation [6]. These tumors are typically slow-growing masses and are classified as head and necck paragangliomas. Due to their propensity for causing significant neurovascular and vascular complications, the surgical resection of these tumors presents a formidable challenge to otolaryngologists [6]. The growth of these tumors is generally slow and asymptomatic, with diagnosis often occurring within the initial years following onset. While primarily growing laterally, these tumors may also extend slightly in the vertical plane. Additionally, vascular pulsations and audible sounds are characteristic features of these tumors. In advanced cases, symptoms such as dysphagia, odynophagia, and dysphonia may arise due to pressure effects or nerve involvement [6].

The differential diagnosis for any masses identified in the lateral neck compartment should encompass a broad range of potential neck masses [7]. In younger patients, differential considerations should include bronchial cysts, hygromas, and lymphadenopathy, whereas in adults, differential diagnoses should also consider carotid artery metastases and glomus jugular tumors [7]. Clinical examination remains the cornerstone of diagnosis, with palpation revealing a pendular or transverse motion of the mass, although this may be transient. The presence of a pulse is possible, but murmurs are typically absent [7,8].

Imaging modalities such as ultrasound and contrast-enhanced CT scans are instrumental in delineating the vascular mass's proximity to the carotid artery. High-resolution MRI can identify lesions as small as 0.8 mm. Larger masses exhibit a distinct "salt and pepper" appearance on T1-weighted MRI, whereas smaller lesions may pose a challenge for radiological detection [9].

The carotid body shows limited responsiveness to radiotherapy; although radiation does not reduce the tumor's size, it effectively inhibits further growth. For patients ineligible for surgical intervention, radiotherapy or vigilant monitoring is recommended, though prompt surgical excision remains the preferred approach upon diagnosis [10].

Surgical management of carotid body tumors aims to achieve complete tumor resection while preserving vascular integrity, avoiding damage to critical organs and nerves, and maintaining cerebral function [11,12].

Typically, these tumors develop at the carotid artery bifurcation in the neck, often located below the jaw angle. The bifurcation commonly occurs at the C4 and C5 vertebral levels, at the superior line of the thyroid cartilage, within the carotid sheath, and lateral to the foramen transversarium. However, a retropharyngeal location is a rare variant, with an incidence of approximately 2.6 % [2–4]. Given the rarity of carotid body tumors and the improbability of their occurrence in a retropharyngeal carotid artery [1–4], this article reports an exceptionally rare case of a retropharyngeal carotid body tumor in a 50-year-old man. As this case represents a unique occurrence, it may provide valuable insights for surgeons encountering similar rare presentations.

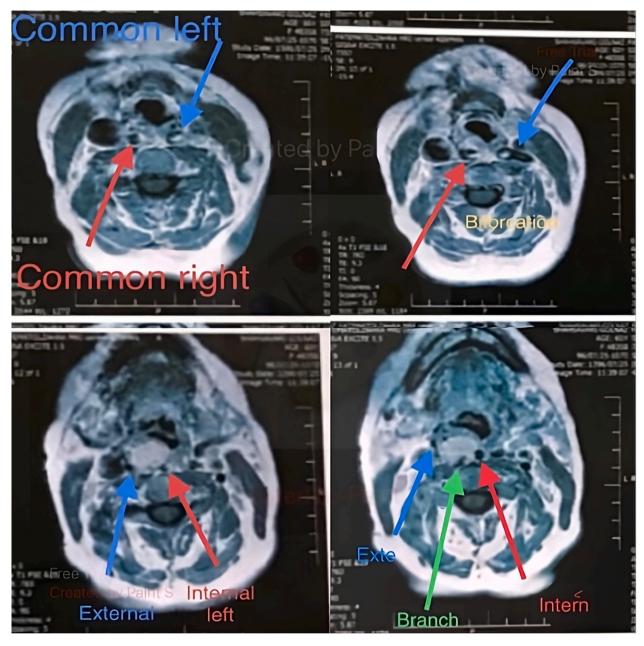


Fig. 2. Location of common, internal and external carotid artery demonstrated with arrows.

# 5. Conclusion

Retropharyngeal carotid body tumors are uncommon. They can be safely removed through this case report's surgical approach with acceptable morbidity.

# **Author contribution**

A.A., A.S. and F.J. wrote the manuscript and conducted the literature review. A.S., F.J. and D.N. provided critical feedback and revised the manuscript. F.J. collected and analyzed the patient data. A.A. and A.S. and F.J. provided additional clinical input and helped to interpret the findings. All authors have read and approved the final version of the manuscript for publication.

# Consent

Written informed consent was obtained from the patient for

publication of this case report and any accompanying images.

# **Ethical approval**

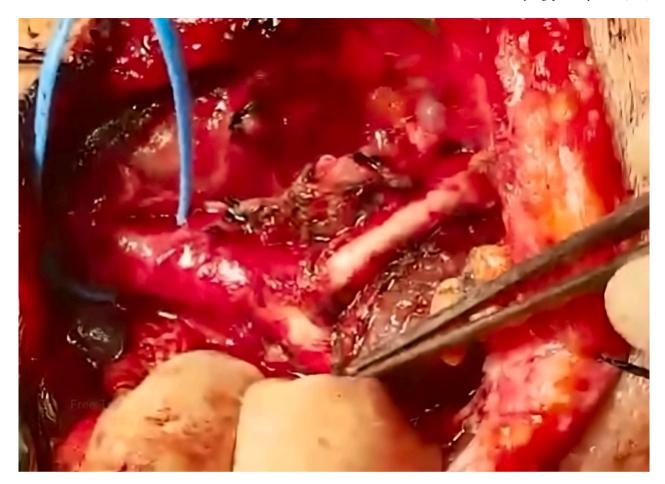
Institutional review board approval for the case report is not required at our institution. To keeping ethical principles, names of the patients were not pointed in the paper and the rights of the subject were protected.

### Guarantor

Fatemeh Jahanshahi.

# **Funding**

There was no funding for this paper.



 $\textbf{Fig. 3.} \ \ \textbf{Intraoperative figure showed the carotid artery.}$ 

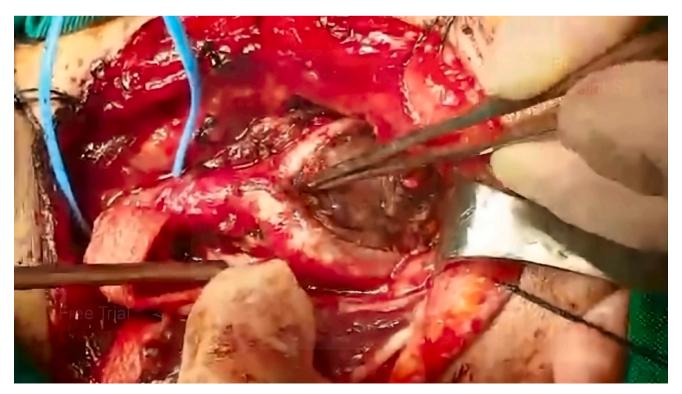


Fig. 4. Intraoperative figure showed the intact carotid bifurcation after the excision of a carotid body tumor.

#### Conflict of interest statement

The authors report no conflicts of interest in this work.

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