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CASE REPORT

CAROTID BODY PARAGANGLIOMA PRESENTING AS A LATERAL NECK MASS: A CASE REPORT

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ABSTRACT

Paragangliomas are rare neuroendocrine tumors arising from paraganglionic tissue, most commonly located at the carotid bifurcation. We report a case of a 33-year-old female presenting with a progressively enlarging left-sided neck swelling associated with intermittent radiating pain. Clinical examination revealed a firm, mobile mass measuring 5 × 3 cm. Imaging studies, including ultrasound and computed tomography, demonstrated a highly vascular lesion at the carotid bifurcation. Fine-needle aspiration cytology was inconclusive. The patient underwent surgical excision under general anesthesia. Histopathological examination confirmed the diagnosis of paraganglioma, characterized by nests of cuboidal cells arranged in a classic Zellballen pattern separated by vascular septa. The tumor was classified as Shamblin Type I and was successfully excised without complications. This case highlights the importance of imaging and surgical management in diagnosing carotid body tumors.

Keywords**:** Paraganglioma, neck swelling, carotid bifurcation, excisional biops

INTRODUCTION

Paragangliomas are rare, highly vascular neuroendocrine tumors that arise from paraganglionic tissue derived from neural crest cells and distributed along the autonomic nervous system. Within the head and neck region, these tumors account for less than 0.5% of all neoplasms and most commonly originate at the carotid bifurcation, followed by the jugular bulb, vagal paraganglia, and tympanic plexus [1,2]. Despite their rarity, their clinical significance lies in their close anatomical relationship with major neurovascular structures and their potential for genetic association and malignant transformation.

Carotid body paragangliomas (CBPs), the most frequent subtype of head and neck paragangliomas, arise from the carotid body-a specialized chemoreceptor organ located at the carotid bifurcation that plays a crucial role in oxygen sensing and regulation of respiratory activity. Chronic hypoxia has been implicated in the pathogenesis of these tumors, with epidemiological studies demonstrating a higher prevalence among individuals living at high altitudes or those with chronic hypoxemic conditions [3]. These tumors are typically slow-growing but exhibit marked hypervascularity, which contributes to both their characteristic imaging features and the technical complexity of surgical management.

Historically, the carotid body was first described by Albrecht von Haller in 1743, who initially misinterpreted it as a neural ganglion. Subsequently, Hubert von Luschka provided a more accurate anatomical and

histological description, referring to it as the "carotid gland" [4]. The evolution of surgical management began with early attempts at tumor excision in the late 19th century, culminating in the identification of the subadventitial dissection plane-commonly known as the "white line"-by Gordon Taylor, which significantly improved surgical safety and outcomes [5].

From a histopathological perspective, paraganglia consist of chief (type I) cells and sustentacular (type II) cells arranged in a characteristic nested pattern known as the "Zellballen" architecture. Chief cells contain abundant granular cytoplasm and are responsible for neuroendocrine activity, while sustentacular cells provide structural support. Although most head and neck paragangliomas are nonfunctional, a minority may secrete catecholamines, leading to systemic symptoms such as hypertension, palpitations, and headaches [6].

Paragangliomas are broadly classified into sympathetic (chromaffin) and parasympathetic (nonchromaffin) types. Sympathetic paragangliomas, typically located in the adrenal medulla and along the sympathetic chain, are more likely to be hormonally active. In contrast, parasympathetic paragangliomas, which predominate in the head and neck region, are generally nonsecretory and present as painless, slowly enlarging masses [1,6]. Approximately 80–85% of paragangliomas arise in the adrenal medulla (pheochromocytomas), whereas 15–20% are extra-adrenal, with only a small proportion occurring in the head and neck region [7].

Recent advances in molecular genetics have significantly enhanced the understanding of paraganglioma pathogenesis. It is now recognized that up to 30–40% of these tumors are associated with germline mutations, particularly involving genes encoding subunits of the succinate dehydrogenase (SDH) complex, including SDHB, SDHC, and SDHD [7,8]. These mutations are associated with increased risks of multifocality, recurrence, and malignancy, especially in SDHB mutation carriers. Consequently, genetic counseling and screening have become essential components of modern diagnostic and management protocols.

Radiological evaluation plays a central role in the diagnosis and preoperative assessment of carotid body paragangliomas. Ultrasound may reveal a hypervascular mass at the carotid bifurcation, while contrast-enhanced computed tomography (CT) provides detailed information regarding tumor size and its relationship to adjacent vascular structures. Magnetic resonance imaging (MRI) is considered the imaging modality of choice due to its superior soft tissue contrast and its characteristic "salt-and-pepper" appearance, reflecting flow voids from high vascularity and slow-flow hemorrhage [2,9]. Digital subtraction angiography remains the gold standard for assessing tumor vascularity and is particularly useful when preoperative embolization is considered to minimize intraoperative blood loss [9].

Clinically, carotid body tumors typically present as painless, slowly enlarging lateral neck masses that are mobile in the horizontal plane but relatively fixed vertically, a feature often referred to as Fontaine's sign. As the tumor enlarges, it may compress adjacent cranial nerves, leading to symptoms such as dysphagia, hoarseness, or tongue deviation. The differential diagnosis includes reactive or metastatic lymphadenopathy, branchial cleft cysts, salivary gland tumors, thyroid neoplasms, neurogenic tumors such as schwannomas, and vascular lesions including carotid artery aneurysms [10].

Although the majority of carotid body paragangliomas are benign, malignant transformation—defined by the presence of regional or distant metastases—occurs in approximately 5–10% of cases (8). The risk of malignancy is higher in patients with SDHB mutations and in larger tumors. Multicentric tumors are observed in approximately 10% of cases, particularly in familial forms, with bilateral carotid body tumors being the most common presentation [3,7].

Surgical excision remains the treatment of choice for most carotid body paragangliomas, particularly for Shamblin type I and II lesions. The Shamblin classification categorizes tumors based on their relationship to the carotid vessels and serves as an important predictor of surgical complexity and morbidity. While complete surgical resection offers definitive treatment, it carries risks including cranial nerve injury, stroke, and significant blood loss. In selected cases, particularly in elderly patients or those with high surgical risk, radiotherapy or a "watchful waiting" approach may be considered [9,10].

In summary, carotid body paragangliomas represent rare but clinically significant tumors requiring a comprehensive diagnostic and therapeutic approach. Advances in imaging, molecular genetics, and surgical techniques have improved diagnostic accuracy and patient outcomes. However, due to their complex anatomical location, potential for genetic predisposition, and risk of morbidity, management of these tumors necessitates a multidisciplinary strategy involving radiologists, surgeons, pathologists, and genetic specialists.

We report a case of carotid body paraganglioma in a young female patient, emphasizing its clinical presentation, diagnostic evaluation, and surgical management.

CASE REPORT

A 33-year-old female patient was referred from the general surgical unit for further evaluation and management of a left-sided neck swelling of approximately two months' duration. The patient reported a gradual increase in the size of the swelling, accompanied by intermittent pain radiating to the left ear lobe and forehead.

Her medical history was significant for bronchial asthma, for which she was on regular inhalational therapy with budesonide (400 µg daily). She was otherwise healthy. Her surgical history included a previous excision of a broad ligament uterine fibroid.

On clinical examination, a diffuse, firm, and mildly tender swelling measuring approximately 5 × 3 cm was palpated on the left side of the neck. The mass extended from the midline to the posterior auricular region. The swelling was mobile, and the overlying skin appeared normal, with no evidence of erythema or increased local temperature. No sensory deficits, including paraesthesia or anaesthesia, were noted.

Fine-needle aspiration cytology (FNAC) was inconclusive. Ultrasonographic evaluation revealed a well-defined soft tissue mass located anteromedial to the left sternocleidomastoid muscle, closely related to the carotid bifurcation. The lesion demonstrated marked internal vascularity, raising suspicion of a vascular neoplasm. A contrast-enhanced computed tomography (CT) scan was subsequently performed to further delineate the lesion and its anatomical relationships. Based on the clinical and radiological findings, surgical excision was planned for definitive diagnosis and treatment.

On admission, the patient's vital signs were within normal limits, with a blood pressure of 100/70 mmHg, pulse rate of 71 beats per minute, and respiratory rate of 20 breaths per minute. Under general anaesthesia, an excisional biopsy of the left-sided neck mass was performed. Intraoperatively, the tumor appeared as a well-circumscribed, highly vascular mass located at the carotid bifurcation. The lesion was carefully dissected and excised in toto, preserving the adjacent neurovascular structures. The excised specimen was brownish in color, irregular in shape, and measured approximately 50 × 35 × 30 mm.

Figure1. Preoperative presentation of the patient with left-sided diffuse firm neck swelling measuring 5 × 3 cm

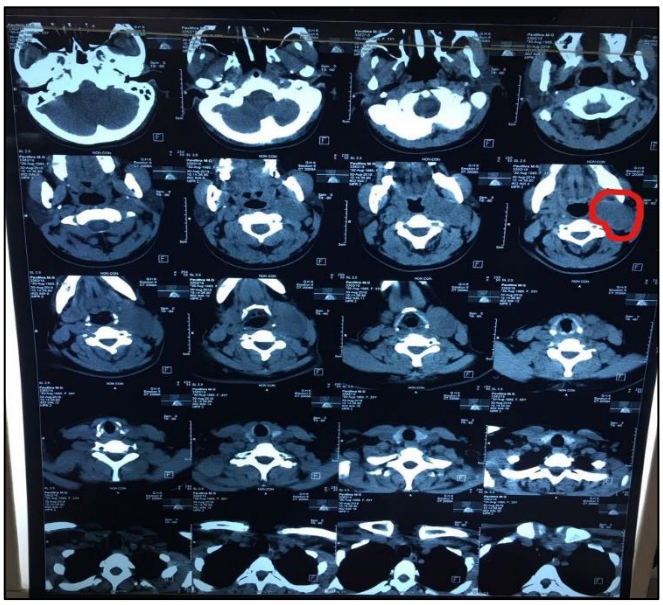

Figure 2. CT scan of the lesion.

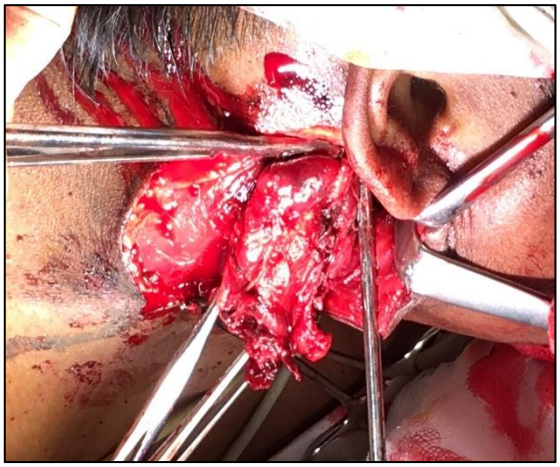

Figure 3. Mass: Irregular and brownish in colour.

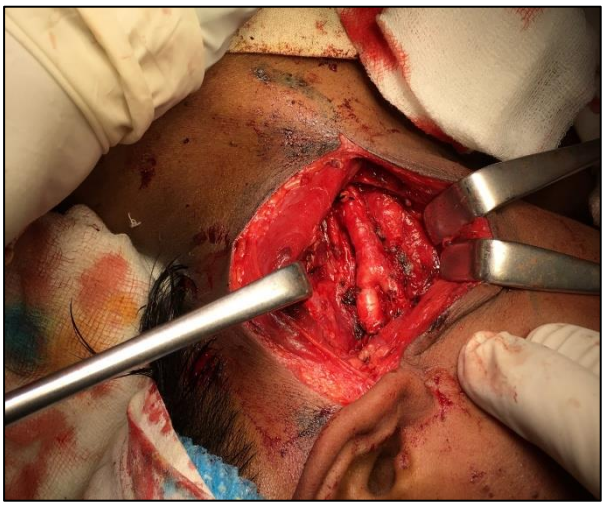

Figure 4. Intraoperative view showing the carotid bifurcation at the site of dissection.

Figure 5. Excised mass measuring 50 × 35 × 30 mm, irregular in shape, and brownish in colour.

Figure 6. Postoperative review of the patient two weeks after excision.

The specimen was submitted for histopathological examination. Microscopic analysis revealed a well-circumscribed tumor surrounded by a thin fibrous capsule. The tumor cells were arranged in characteristic nested patterns (Zellballen architecture), consisting of clusters of polygonal to cuboidal cells separated by delicate, highly vascular fibrous septa. The individual tumor cells exhibited abundant granular eosinophilic cytoplasm and centrally placed round to oval nuclei. These histopathological features were consistent with a diagnosis of paraganglioma.

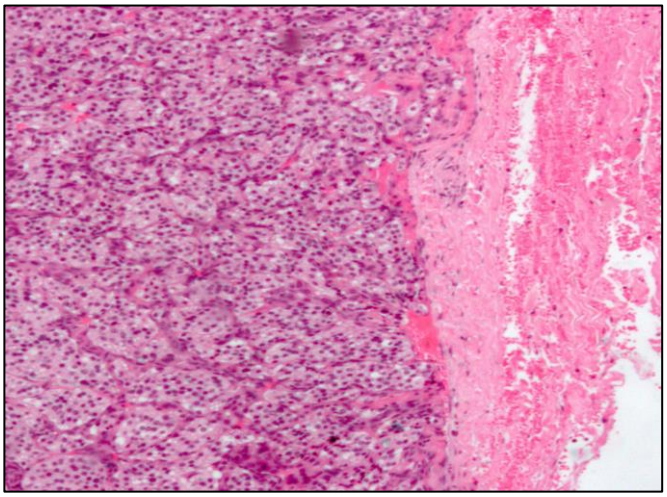

Figure 7. Histopathology: thin fibrous capsule, well-defined nets of cuboidal cells separated by highly vascularized fibroid septa, giving a "zellballen" (cell balloon) appearance. Individual cells had abundant granular basophilic cytoplasm and round nuclei.

DISCUSSION

Carotid body paragangliomas (CBPs) are the most common subtype of head and neck paragangliomas, arising from paraganglionic cells at the carotid bifurcation. Despite their rarity, they are clinically important due to their marked vascularity, proximity to major neurovascular structures, and potential for hereditary predisposition and malignant behavior [1,2]. The present case demonstrates a typical clinical presentation and highlights key diagnostic and therapeutic considerations.

Clinically, CBPs usually present as slow-growing lateral neck masses, often asymptomatic in early stages. However, pain or referred otalgia may occur due to local mass effect or neural irritation, as observed in our patient. The classical finding of horizontal mobility with limited vertical movement (Fontaine's sign) can aid clinical suspicion but is not consistently present [3]. With progressive enlargement, tumors may involve adjacent cranial nerves, particularly the glossopharyngeal, vagus, and hypoglossal nerves, leading to dysphagia, hoarseness, or tongue deviation [2,4]. Radiological imaging is fundamental for diagnosis and surgical planning. Doppler ultrasonography typically demonstrates a

hypervascular mass causing splaying of the internal and external carotid arteries. Cross-sectional imaging with contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) provides detailed anatomical delineation. MRI is considered the gold standard due to its characteristic "salt-and-pepper" appearance resulting from intratumoral flow voids [5]. Digital subtraction angiography (DSA) remains valuable in selected cases, particularly when preoperative embolization is considered to reduce intraoperative blood loss [6]. In the present case, ultrasound and CT imaging were sufficient to suggest a vascular tumor at the carotid bifurcation. Fine-needle aspiration cytology (FNAC) is generally of limited diagnostic value in paragangliomas due to their vascularity and risk of hemorrhage, often yielding inconclusive results, as seen in this case [7]. Therefore, imaging plays a more decisive role in preoperative diagnosis, while histopathological evaluation remains the gold standard for definitive diagnosis.

Histologically, paragangliomas exhibit the classic Zellballen architecture, characterized by nests of chief cells surrounded by sustentacular cells within a highly vascular stroma. These findings were observed in the present case and are considered pathognomonic[8]. Immunohistochemical markers such as chromogranin, synaptophysin, and S-100 protein can further support the diagnosis, although they were not utilized in this instance. The Shamblin classification remains an essential tool for guiding surgical management by categorizing tumors based on their relationship to the carotid vessels [9]. Type I tumors are small and easily resectable, Type II partially encase the vessels, and Type III completely surround them, often requiring vascular reconstruction. The tumor in this case corresponded to Shamblin Type I, allowing complete excision with preservation of adjacent structures and no postoperative complications. Surgical resection is the treatment of choice for most CBPs, particularly in young and medically fit patients [2,6]. However, surgery carries risks, including cranial nerve injury, stroke, and intraoperative hemorrhage. Reported cranial nerve morbidity ranges from 10% to 40%, while perioperative stroke rates are approximately 2–3% in experienced centers [6,9]. Alternative management strategies, such as radiotherapy or active surveillance, may be considered in elderly patients, high-risk surgical candidates, or those with unresectable tumors [10]. Recent advances in molecular genetics have significantly impacted the understanding and management of paragangliomas. Up to 30–40% of cases are associated with germline mutations, particularly in the succinate dehydrogenase (SDH) gene complex (SDHB, SDHC, SDHD), which are linked to increased risks of malignancy, recurrence, and multifocality [11,12]. Although genetic testing was not performed in this case, current guidelines recommend consideration of genetic screening, especially in younger patients or those with multiple tumors. Malignancy in CBPs is relatively uncommon and is defined by the presence of regional or distant metastasis rather than histological features alone. The reported rate ranges from 5% to 10%, with higher risk in SDHB mutation carriers [11]. Long-term follow-up is essential due to the potential for late recurrence or metastasis.

Carotid body tumours are classified by Shamblin into three types:

- Type I: localized mass
- Type II: partially surrounding the carotid artery
- Type III: completely wrapped around and adherent to the carotid artery

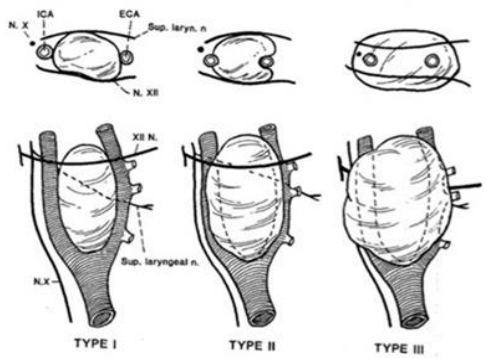

Fig. 8 The classification of Shamblin et al. of the difficulty of surgical resection. Group I tumors are localized and easily resected. Group II includes tumors adherent or partially surrounding vessels. Group III paragangliomas intimately surround or encase the vessels. *ICA* = internal carotid artery; *ECA* = external carotid artery.

Surgical excision is the treatment of choice. For Shamblin Types I and II, careful subadventitial dissection is recommended. The tumour in this case was Shamblin Type I and was excised with clear surgical margins. Incomplete excision leads to a recurrence rate of approximately 10%. In tumour resection, the mean mortality rate is about 2%, perioperative stroke rate 2–3%, and cranial nerve dysfunction rate up to 40%. The vagus and hypoglossal nerves may be involved, affecting function. In this case, all cranial nerves proximal to the tumour remained intact. Although the tumour is benign, malignant transformation occurs in 3–12.5% of cases. Without treatment, mortality may reach 30%. Therefore, regular long-term follow-up, as in this case, is essential.

Future Perspectives

Emerging advances in molecular diagnostics and targeted imaging are expected to further refine the management of paragangliomas. Functional imaging modalities such as positron emission tomography (PET), particularly with 68Ga-DOTATATE, have demonstrated superior sensitivity in detecting multifocal and metastatic disease [13]. Additionally, the integration of genetic profiling into routine clinical practice may enable personalized risk stratification and tailored surveillance strategies. Novel therapeutic approaches,

including targeted molecular therapies and peptide receptor radionuclide therapy (PRRT), are also under investigation and may offer promising alternatives for unresectable or metastatic cases [13,14].

Limitations

This report has several limitations. First, it represents a single case, which limits the generalizability of the findings. Second, advanced imaging modalities such as MRI or angiography were not performed, which could have provided additional diagnostic detail and preoperative planning benefits. Third, immunohistochemical and genetic analyses were not conducted, which may have further supported the diagnosis and provided prognostic information. Finally, long-term follow-up data are not included, limiting assessment of recurrence or late complications.

CONCLUSION

Tumours that are rarely encountered, such as paragangliomas, must be diagnosed early due to their complex relationship with adjacent structures. Surgical treatment is essential, as the potential for malignancy and pressure symptoms may otherwise lead to life-threatening conditions.

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Conflict of Interest
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"Not applicable"

Consent for publication
"Not applicable"

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