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A Case Report and Literature Analysis of 49-Year-Old Male Patient with Neck Paraganglioma

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Abstract

Paraganglioma are rare neuroendocrine tumors arising from paraganglia derived from neural crest cells. They commonly occur in the head and neck region, with the carotid body being the most frequent site. Here, we present a case report of a 49-year-old male patient with a neck Paraganglioma. The patient presented with a painless neck mass, and further investigations led to the diagnosis of Paraganglioma. We discuss the clinical presentation, radiological findings, histopathological features, management, and prognosis of this rare entity. Additionally, we provide a literature analysis to enhance understanding and management strategies for neck Paraganglioma.

Keywords: Paraganglioma; Neck; Neuroendocrine tumor

Introduction

Paraganglioma are rare neuroendocrine tumors arising from extra-adrenal chromaffin cells derived from neural crest cells. They can occur anywhere along the autonomic nervous system, with the head and neck region being one of the most common sites [1]. Within the head and neck, Paraganglioma often arise from the carotid body, jugular vein, tympanic membrane, and vagus nerve. However, Paraganglioma originating from the neck are relatively uncommon. Here, we present a case report of a 49-year-old male patient with a neck Paraganglioma, highlighting the clinical presentation, diagnostic approach, management strategies, and literature review [2].

Case Report

A 49-year-old male patient presented to our clinic with a painless swelling in the right side of his neck that had been progressively increasing in size over the past six months. The patient denied any history of trauma, pain, dysphagia, dyspnea, or voice changes [3]. He had no significant past medical history and was not on any medications. Physical examination revealed a non-tender, firm, mobile mass measuring approximately 3 cm in diameter in the right lower neck region, just above the clavicle. There were no palpable cervical lymph nodes.

Given the suspicion of a neck mass, further investigations were pursued. Ultrasound examination of the neck revealed a well-defined, hypoechoic mass with internal vascularity, suggestive of a vascular lesion. Subsequent contrast-enhanced computed tomography (CT) scan of the neck demonstrated a homogeneously enhancing mass located poster lateral to the right common carotid artery, consistent with a Paraganglioma [4]. Magnetic resonance imaging (MRI) was performed for better soft tissue delineation and to assess the relationship of the tumor with adjacent structures. MRI confirmed the presence of a well-circumscribed, T2 hyperintense, and T1 hypointense mass, without evidence of invasion into surrounding structures (Figure 1).

Based on the clinical presentation and radiological findings, a fine-needle aspiration biopsy (FNAB) was performed, which yielded cytology consistent with a Paraganglioma. Subsequently, the patient underwent surgical resection of the neck mass [5]. Intraoperatively, the tumor was found to be intimately associated with the carotid artery, but without invasion into the vessel wall. Complete excision of the mass was achieved, and the patient had an uneventful postoperative course.

Histopathological examination of the excised specimen confirmed the diagnosis of a Paraganglioma, showing nests of polygonal cells with eosinophilic cytoplasm arranged in a characteristic "zellballen" pattern. Immunohistochemical staining was positive for chromogranin and synaptophysin, supporting the neuroendocrine nature of the tumor. The surgical margins were negative for tumor involvement [6].

Discussion

Paraganglioma are rare neuroendocrine tumors originating from paraganglia, which are dispersed neuroendocrine cells derived from

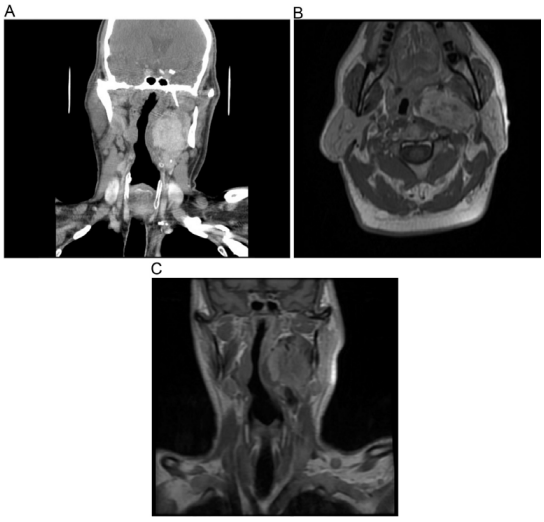

Figure 1: MRI confirmed the presence of a well-circumscribed, T2 hyperintense, and T1 hypointense mass.

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neural crest cells. These tumors can occur sporadically or as part of hereditary syndromes such as multiple endocrine neoplasia type 2 (MEN2) and von Hippel-Lindau (VHL) syndrome [1]. In the head and neck region, paragangliomas most commonly arise from the carotid body, accounting for approximately 60% of cases, followed by the jugular vein (15-20%), tympanic membrane (5-10%), and vagus nerve (5-10%).

The clinical presentation of neck paragangliomas varies depending on the size and location of the tumor. Patients may present with a painless neck mass, as seen in our case, or with symptoms related to compression of adjacent structures such as dysphagia, dyspnea, or hoarseness [7]. The diagnosis of paragangliomas relies on a combination of clinical, radiological, and histopathological findings. Imaging modalities such as ultrasound, CT, and MRI are helpful in delineating the extent of the tumor and its relationship with adjacent structures. Fine-needle aspiration biopsy may be performed to obtain cytological material for diagnosis, although the definitive diagnosis is established by histopathological examination of the surgical specimen. Surgical resection remains the mainstay of treatment for neck paragangliomas [8]. The goal of surgery is complete excision of the tumor while preserving vital structures such as the carotid artery and cranial nerves. Preoperative embolization may be considered in larger tumors to minimize intraoperative bleeding. Adjuvant radiotherapy or chemotherapy may be indicated in cases of unresectable tumors, metastatic disease, or as a palliative measure. Long-term follow-up is essential due to the risk of recurrence and the potential for the development of metachronous or synchronous tumors, particularly in patients with hereditary syndromes [9,10].

Conclusion

Neck paragangliomas are rare neuroendocrine tumors that require a multidisciplinary approach for diagnosis and management. Clinical suspicion, coupled with appropriate imaging studies and histopathological examination, is crucial for accurate diagnosis. Surgical resection remains the cornerstone of treatment, with the goal of complete excision while preserving neurovascular structures. Long-term follow-up is essential to monitor for recurrence and to manage patients with hereditary syndromes predisposing to paraganglioma development. Further studies are needed to elucidate the molecular mechanisms underlying paraganglioma pathogenesis and to optimize treatment strategies for this rare entity.

Acknowledgement

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Conflict of Interest

None

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