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Dissecting the mystery: A case report of a parapharyngeal ganglioneuroma

Bader Alali, Maha Al-Gilani, Boobalan Samynathan

ABSTRACT

Introduction: Ganglioneuromas, slow-growing non-neoplastic tumors from neural crest cells, are typically asymptomatic but may manifest clinically when reaching a significant size. While rare in the parapharynx, imaging assists in preoperative planning and assessing features of more sinister pathologies.

Case Report: A 45-year-old woman presented with persistent unilateral neck pain and sore throat, computed tomography (CT) imaging parapharyngeal space, ultimately diagnosed as a parapharyngeal ganglioneuroma on tissue biopsy. Surgical excision was performed with an uneventful postoperative course.

Conclusion: Parapharyngeal ganglioneuromas pose diagnostic challenges due to their rarity and diverse presentations. This case underscores the diagnostic challenges posed by rare parapharyngeal lesions and highlights the successful management of ganglioneuromas through appropriate imaging and surgical intervention.

Keywords: Case report, Computed tomography, Ganglioneuroma, Parapharyngeal

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Bader Alali[1], Maha Al-Gilani[1], Boobalan Samynathan[2]

Affiliation: [1]Department of Otorhinolaryngology-Head and Neck Surgery, Ministry of Health, Kuwait City, Kuwait; [2]Department of Pathology, Jaber Alahmed Hospital, Ministry of Health, Kuwait City, Kuwait.

Corresponding Author: Bader Alali, MD, Shuhada Block 5, St 518, House 17, Kuwait City, Kuwait; Email: Mr.badernalali@gmail.com

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INTRODUCTION

Ganglioneuromas, rare benign tumors originating from primordial neural crest cells, are characterized by the presence of ganglion cells, neurites, Schwann cells, and fibrous tissues. While commonly primary in origin, they can also arise through spontaneous or treatment-induced differentiation of a neuroblastoma or ganglioneuroblastoma [1]. Despite their typically slow growth, these tumors may attain significant sizes, posing clinical implications such as the compression of surrounding structures or displaying neuroendocrine activity, including the release of catecholamines [2]. Notably, their often-asymptomatic nature often results in incidental diagnosis [3]. Ganglioneuromas predominantly occur in the posterior mediastinum, retroperitoneal region, or adrenal region, with occurrences in the neck being exceptionally rare [4]. We present a 45-year-old woman who was histologically diagnosed with a parapharyngeal ganglioneuroma, while emphasizing diagnostic and therapeutic implications/challenges.

CASE REPORT

A 45-year-old woman was referred to the otolaryngology head and neck surgery clinic after the discovery of a suspicious retropharyngeal lesion on computed tomography (CT) imaging. She had been experiencing persistent unilateral neck pain and sore throat for several weeks, for which her primary care physician had prescribed oral antibiotics. She requested a CT scan of the neck which incidentally found a large retropharyngeal lesion, leading to her referral.

Upon evaluation at the head and neck surgery clinic, the patient reported no dysphagia, odynophagia, neck stiffness, trismus, systemic symptoms, or recent dental

infections. Her medical history included diabetes mellitus and being a non-smoker. Her surgical history involves an excision of a post-auricular reactive lymph node hyperplasia six months prior to her presentation. A comprehensive head and neck examination demonstrated asymmetric erythematous tonsils, and no palpable suspicious neck nodes.

The initial CT neck indicated a left large retropharyngeal hypodense structure measuring 6.2×3.3×1.3 cm, favoring a differential diagnosis of a retropharyngeal collection with dense content. The patient received a follow-up appointment in two weeks, along with a course of oral antibiotics.

However, her symptoms persisted during the follow-up, prompting a repeat contrast-enhanced CT neck. The imaging revealed a hypoattenuating partly defined non-capsulated lesion at the left parapharyngeal space measuring 6.16×3.14×1.99 cm, displacing the parapharyngeal mucosal space (Figure 1). The lesion extended into the carotid sheath and anterior prevertebral space along the soft tissue planes, as seen in Figure 1A–C.

While the initial radiologist impression suggested a deep neck infection, further discussion led to the decision of progressing to an incision and drainage versus excisional biopsy of an atypical lesion through an external approach. Operative findings revealed a lesion medial to the common carotid artery extending to the parapharyngeal space. Complete excision was performed, and the specimen was submitted for histopathological analysis.

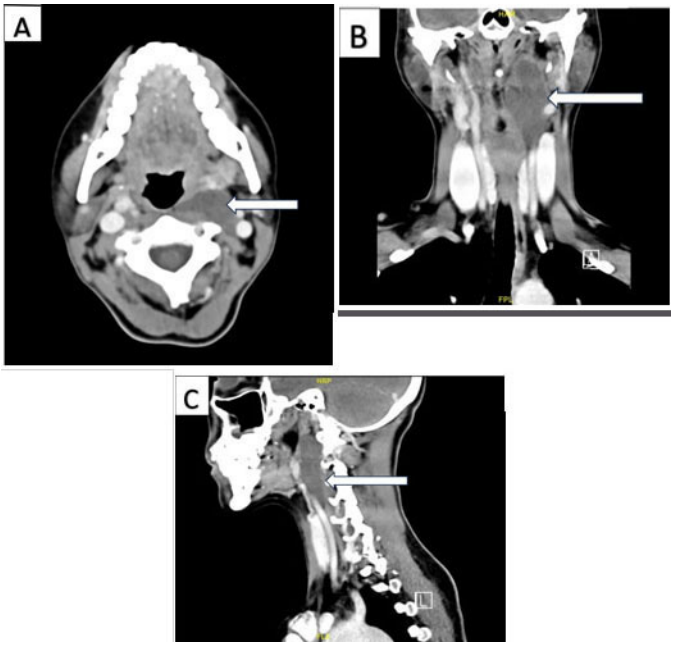

Figure 1: Contrast-enhanced CT scans of the neck. (A) Axial views, (B) Coronal views, and (C) Sagittal views of a contrast-enhanced CT neck show a hypoattenuating partly defined non-capsulated lesion (white arrows) at the left parapharyngeal space measuring 6.16×3.14×1.99 cm.

The histopathologist examined a single, soft, yellow nodular tissue measuring 5×3×1.2 cm and weighing 5.0 grams, displaying a smooth tan-yellow external surface. Upon sectioning, the lesion's cut surface presented a firm, uniform tan-yellow appearance, with a focal gray area measuring 0.8×0.4×0.3 cm. Microscopic analysis revealed a well-circumscribed encapsulated tumor composed of small intersecting fascicles of Schwann cells in a loose myxoid stroma (Figure 2A). Mature ganglion cells were scattered throughout the tumor in small nests and clusters, without evidence of heterogeneous or hemorrhagic areas (Figure 2B). Immunohistochemistry analysis confirmed positive staining for S-100 (stroma, ganglion cells) and synaptophysin (ganglion cells only) as demonstrated in Figure 2C and D. The injury score was 0 with no evidence of cell or extracellular matrix damage. The final diagnosis rendered was a parapharyngeal ganglioneuroma.

The patient's postoperative course was uneventful, she was discharged the following day on oral analgesia. Subsequently, she demonstrated an uncomplicated recovery when seen one-month post-surgery, with assessment for remission of her initial symptoms and for any complications such as dysphagia, dysphonia, or wound infections.

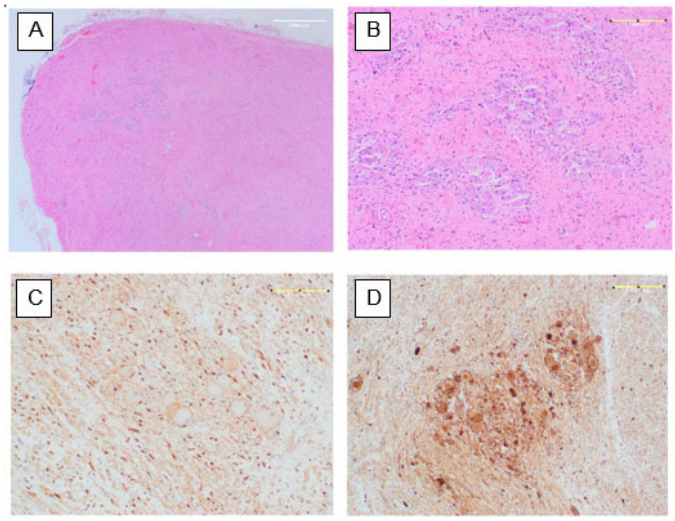

Figure 2: Histopathological characteristics of the parapharyngeal lesion. (A) A well circumscribed lesion with small intersecting fascicles of bland spindle cells in a loose myxoid stroma (H&E stain, ×20). (B) Ganglion cells in small nests and clusters (H&E stain, ×40). (C) Positive-stromal and ganglion cells (S-100 stain, ×40). (D) Positive-ganglion cells (Synaptophysin stain, ×40).

DISCUSSION

In this case report, we present the clinical details of a 45-year-old female diagnosed with a parapharyngeal ganglioneuroma. These rare, benign neurogenic tumors originate from the central or peripheral components of the autonomic nervous system, with common locations being the posterior mediastinum (60–80%), abdomino-pelvic region (10–15%), and cervical region (5%). Less

frequently, they are found in the parapharynx, middle ear, orbital space, or skin. Their typically slow growth often delays clinical manifestation until incidentally discovered or, less commonly, when compression of surrounding structures prompts symptoms such as dysphagia, dyspnea, or a globus sensation. Unfortunately, these nonspecific symptoms may lead to inaccurate diagnoses, as observed in our patient. In rare cases, these tumors can become metabolically active, resulting in symptoms such as diarrhea, hypertension, and virilization [5].

Contrast-enhanced CT imaging plays a crucial role in assessing the size, extension, and composition of the lesion. Ganglioneuromas on CT typically appear well-circumscribed with low to intermediate attenuation, and the presence of discrete calcifications, as opposed to a coarse pattern, aids in distinguishing them from neuroblastomas. T2-weighted magnetic resonance images (MRI) of ganglioneuromas are characterized by inhomogeneity with high-intensity signaling [6].

While radiological diagnosis of parapharyngeal space gangliomas poses a challenge, their utility lies in excluding other pathological entities. The definitive diagnosis of parapharyngeal space tumors is ideally obtained via a tissue biopsy. When lymphoma is suspected, an open neck or transoral approach is preferred for tissue diagnosis [7]. Macroscopically, ganglioneuromas exhibit a mucinous appearance and are encapsulated. Microscopically, features include intersecting bundles of spindle cells, a loose myxoid stroma, and dysplastic ganglion cells, with mature ganglion cells being the most prominent characteristic, as was reflected in our specimen [8].

The preferred treatment for these tumors is surgical excision, usually sufficient if complete resection is achieved. While inadequate resection does not typically lead to regrowth or ongoing symptoms, there are rare, documented cases of malignant transformation into neuroblastomas or a malignant peripheral nerve sheath tumor. Intra-operative risks are a concern, especially when tumors are intimately involved with critical structures such as neurovasculature, which can lead to significant morbidity if injured or sacrificed [8].

CONCLUSION

Parapharyngeal ganglioneuromas (GN) are uncommon benign soft tissue neurogenic tumors recognized for their slow growth and frequently asymptomatic presentation. Although imaging plays a crucial role in the initial assessment, histopathological analysis through surgical biopsy remains the definitive diagnostic method. Surgical excision stands out as the preferred treatment, demonstrating a favorable prognosis with a minimal recurrence rate. This report underscores the intricacies of diagnosis and treatment considerations related to parapharyngeal ganglioneuromas.

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Author Contributions

Bader Alali – Acquisition of data, Analysis of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Maha Al-Gilani – Conception of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Boobalan Samynathan – Design of the work, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Consent Statement
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Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.