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Case Report

The Vagus in Disguise: Plexiform Schwannoma of Left Cervical Vagus Nerve - A Rare Case Report

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Abstract: ***Background*:** Schwannomas are benign peripheral nerve sheath tumors originating from Schwann cells. While they commonly present as solitary, slow-growing masses in the head and neck, the plexiform or multinodular variant involving the vagus nerve is exceedingly rare, particularly in pediatric patients. ***Case Description*:** We report the case of an 11-year-old male who presented with a painless, gradually progressive left-sided neck mass measuring 12 cm x 10 cms over a 6-month period, associated with progressive speech disturbances and dysphagia. Computed tomography (CT) and Magnetic Resonance Imaging (MRI) suggested a vagal nerve neurogenic tumor situated within the left parapharyngeal space. Fine-needle aspiration cytology (FNAC) was inconclusive. The patient underwent an en bloc surgical resection of the 10 x 9 cms tumor, which was intraoperatively noted to be continuous with the left vagus nerve. Histopathological examination confirmed a plexiform schwannoma, demonstrating spindle cells arranged in small fascicles with prominent Verocay bodies and nuclear palisading. Transient postoperative left vocal cord paresis and dysphagia occurred but subsequently resolved. ***Conclusion*:** Vagal nerve plexiform schwannomas pose significant diagnostic challenges due to their rarity and atypical presentations. Surgical excision remains the mainstay of treatment, requiring a delicate balance between complete oncological resection and the preservation of lower cranial nerve function.

Keywords: Plexiform Schwannoma, Vagus Nerve, Parapharyngeal Space, Pediatric Neck Mass, Case Report.

1. INTRODUCTION

Schwannomas, also known as neurilemomas, are benign, encapsulated, slow-growing peripheral nerve sheath tumors derived entirely from Schwann cells. Approximately 25% to 45% of all schwannomas occur in the head and neck region, with the parapharyngeal space being the most frequent site. However, the vagus nerve is rarely affected, accounting for a minimal fraction of these cases.

While classic schwannomas present as a solitary localized mass, the plexiform or multinodular growth pattern is a rare morphological variant. Plexiform schwannomas usually affect the superficial soft tissues and are generally sporadic, though they can occasionally be associated with Neurofibromatosis Type 2 (NF-2). Diagnosing a vagal nerve schwannoma preoperatively is highly challenging because patients often present with an asymptomatic lateral neck mass, and conventional cytological investigations like fine-needle aspiration cytology (FNAC) frequently yield inconclusive results.

We present a rare case of a massive left vagal nerve plexiform schwannoma in an 11-year-old male child, highlighting the clinical presentation, radiological features, surgical management, and subsequent functional outcomes.

2. CASE PRESENTATION

2.1 Clinical History and Presentation

An 11-year-old male child presented to our department with a major complaint of a swelling on the left side of his neck lasting 6 months. The swelling was initially noticed by the patient as a small 3 x 3 cms nodule that progressively increased in size to attain its current dimensions. The mass was completely painless. However, the patient's parents noted recent onset of

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speech disturbances and mild difficulty during swallowing (dysphagia). The patient had an unremarkable past medical history with no features suggestive of Neurofibromatosis.

2.2 Physical and Local Examination

On localized physical examination, a large, oval-shaped swelling measuring approximately 12 x 10 cm was observed on the left side of the neck.

Anatomical Extension:

The mass extended superiorly to the tragus, inferiorly to about 6 cm above the clavicle, anteriorly to the mandibular ramus, and posteriorly to the mastoid process.

Characteristics:

The overlying skin was normal and freely pinchable. The mass had a smooth surface, regular margins, but an ill-defined deep border. On palpation, it was firm in consistency.

Mobility:

Notably, the swelling became significantly less prominent upon contraction of the ipsilateral sternocleidomastoid muscle, suggesting its deep submuscular/parapharyngeal location.

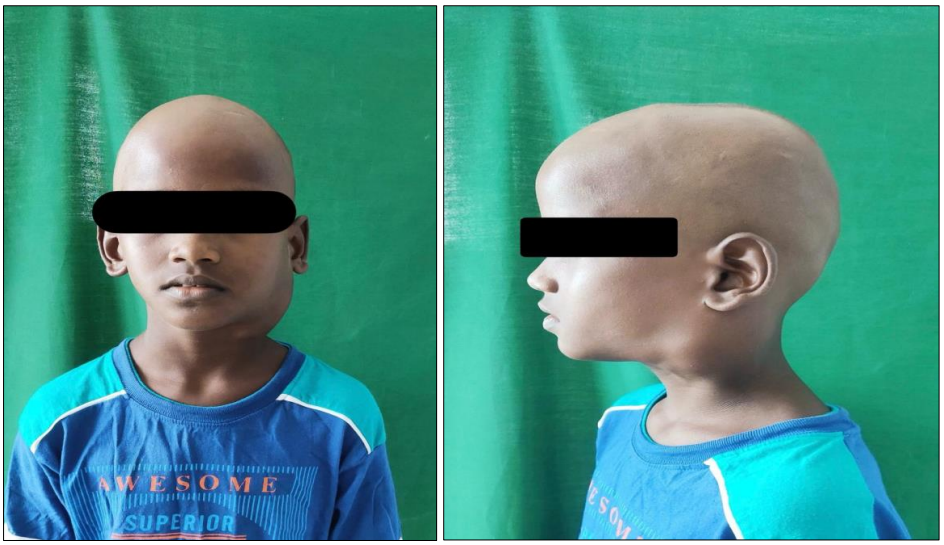

Fig. 1

2.3 Diagnostic Evaluation

Ultrasonography (USG): Indicated a well-defined, solid neurogenic tumor in the left cervical region.

Fine-Needle Aspiration Cytology (FNAC): Attempted preoperatively but yielded an inconclusive diagnosis.

Contrast-Enhanced Computed Tomography (CECT) & MRI:

Advanced imaging demonstrated a large, well-circumscribed retrostyloid parapharyngeal space mass pushing adjacent structures anteriorly. The imaging features strongly raised a high suspicion of a vagal nerve schwannoma.

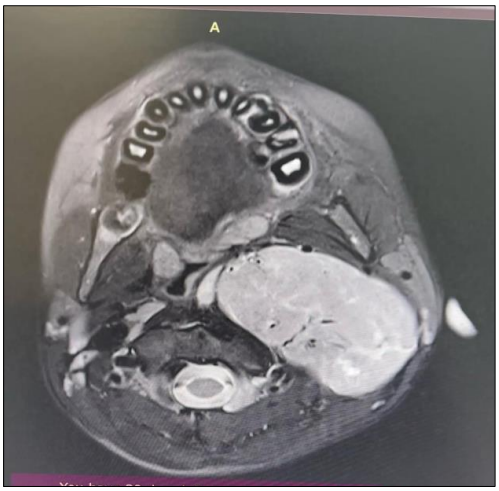

Fig. 2

2.4 Surgical Management

The patient was scheduled for surgical excision under general anesthesia via a transcervical approach.

Intraoperative Findings:

A yellowish-red, well-encapsulated tumor measuring 10 cm x 9 cm was encountered within the left parapharyngeal space. The carotid bifurcation and its branches were displaced anteriorly by the tumor. Both the superior and inferior poles of the mass were found to be directly continuous with the trunk of the left vagus nerve.

Procedure:

Utilizing meticulous dissection, the tumor was resected en bloc. Every effort was made to minimize direct traction injuries to the surrounding neurovascular bundle.

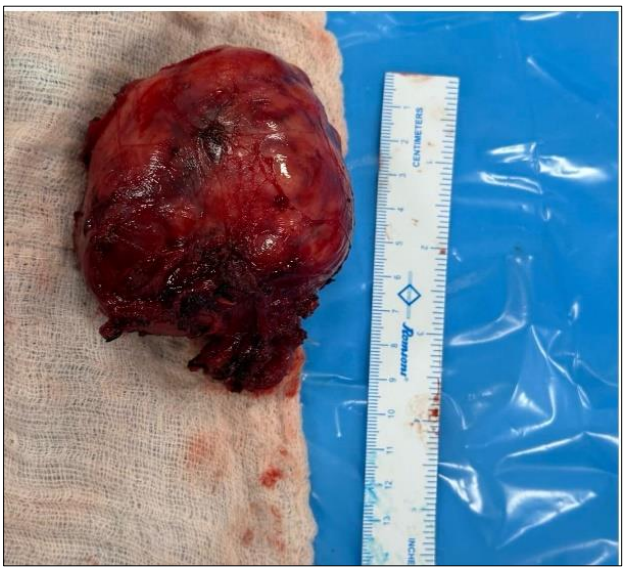

Fig. 3

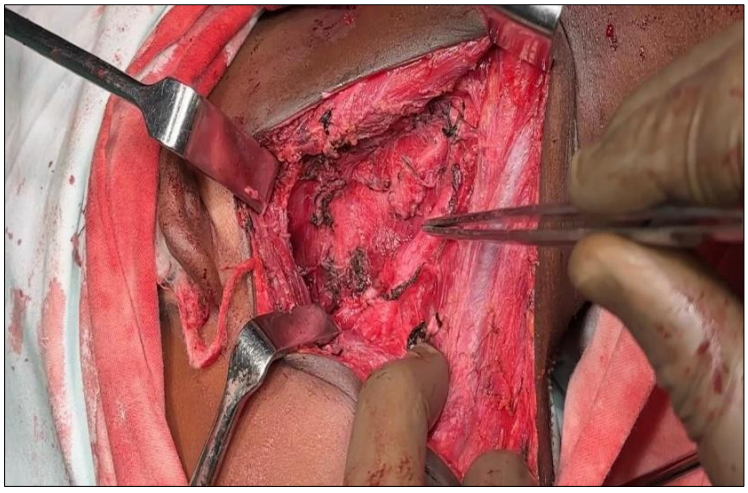

Fig. 4

3. HISTOPATHOLOGICAL EXAMINATION (HPE)

Gross examination confirmed a well-encapsulated, multinodular mass. Microscopic evaluation of the resected tissue sections stained with Hematoxylin and Eosin (H&E) revealed characteristic elements of a plexiform schwannoma:

Cellular Architecture: Spindle-shaped cells arranged in small, interlacing fascicles.

Palisading and Verocay Bodies: Prominent nuclear palisading around acellular eosinophilic regions (Verocay bodies) characteristic of Antoni a areas.

Growth Pattern:

A multinodular/plexiform architecture confined within the nerve sheath. There were no signs of mitotic atypia or malignancy, confirming a benign plexiform schwannoma of the vagal nerve.

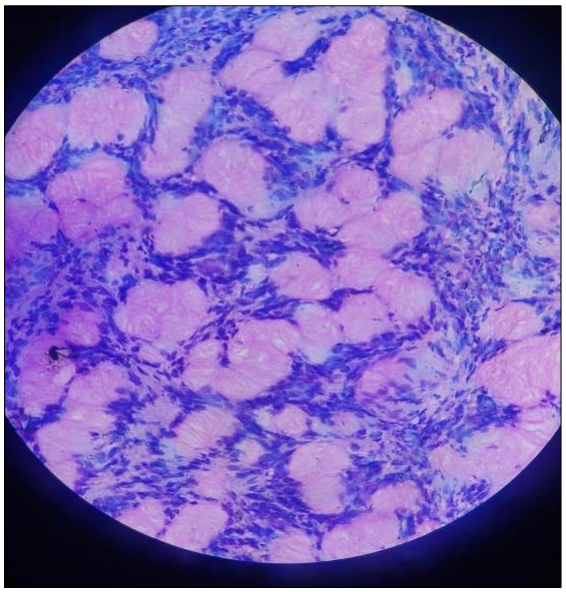

Fig. 5

4. DISCUSSION

Vagal nerve schwannomas are uncommon benign tumors that create a diagnostic dilemma for head and neck surgeons. They frequently present as a slow-growing, asymptomatic lateral neck mass, mimicking branchial cleft cysts, lymphadenopathy, or carotid body tumors. In this patient, the enormous size 12 cm x 10 cm led to compressive symptoms, presenting as speech disturbances and dysphagia, which are unusual presentation features in young pediatric cohorts.

The plexiform variant is rarer still in the vagus nerve trunk. Unlike plexiform neurofibromas, which carry a significant risk of malignant transformation and are pathognomonic for Neurofibromatosis Type 1 (NF-1), plexiform schwannomas are benign, rarely undergo malignant degeneration, and are more frequently associated with NF-2 or occur sporadically.

Preoperative cytological assessment via FNAC is notorious for being inconclusive in deep-seated neurogenic tumors due to dry taps or inadequate cellular sampling from dense stromal components. Consequently, high-resolution imaging modalities like CECT and MRI play a critical role. On cross-sectional imaging, vagal schwannomas characteristically displace the internal jugular vein laterally and the internal carotid artery medially/anteriorly, which perfectly matches our intraoperative findings where the carotid bifurcation was pushed anteriorly.

Postoperative Course Note:

Postoperatively, the patient experienced left vocal cord paresis alongside temporary worsening of deglutition/dysphagia. This is a common and anticipated complication following the resection of large vagal nerve masses due to surgical manipulation or direct vagal nerve discontinuity. Fortunately, with conservative management and structured voice therapy, these neurological deficits completely subsided.

5. CONCLUSION

Schwannomas of the vagus nerve are rare clinical entities in the head and neck, especially when presenting with a plexiform growth pattern in pediatric patients. Achieving a definitive diagnosis preoperatively is challenging, given that FNAC is frequently inconclusive. Cross-sectional imaging remains indispensable for surgical planning.

The primary modality of treatment is complete surgical resection. Clinicians must maintain a high index of suspicion and perform meticulous dissection to preserve lower cranial nerve function whenever possible. Postoperative vocal cord palsy is a known complication that can be effectively managed via conservative approaches and dedicated postoperative rehabilitative therapy.

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