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RESEARCH ARTICLE
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A RARE CASE OF PAEDIATRIC THORACIC INLET GANGLIONEUROMA PRESENTING AS A CERVICOTHORACIC MASS

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Introduction

Posterior mediastinal tumours in children represent a unique subgroup of thoracic neoplasms, with the majority being of neurogenic origin. These lesions arise from sympathetic chain elements, peripheral nerve sheaths, or paravertebral ganglia. Although neuroblastomas account for most posterior mediastinal tumours in early childhood, the ganglioneuromas are considerably less common and typically present in adolescents. [1]

Ganglioneuromas grow slowly and may extend through anatomical conduits such as the thoracic inlet or intervertebral foramina because of their indolent behaviour. As a result, they often reach considerable size before detection. Their location adjacent to the airway, great vessels, vertebral column, and cervical neurovascular bundle makes surgical management challenging despite their benign nature. [2]

Cross-sectional imaging is essential to delineate the tumour's relationship to critical mediastinal structures and to evaluate potential intraspinal extension. Although complete excision is the preferred treatment, the approach must be individualised based on the cranio-caudal extent, proximity to the arch vessels, and degree of displacement of the airway and oesophagus. Large tumours involving the cervicothoracic junction may require combined cervical and intrathoracic exposures to achieve complete resection while avoiding damage to neurovascular structures.

This report describes a rare paediatric ganglioneuroma extending from the posterior mediastinum through the thoracic inlet into the neck and highlights the value of an extended median sternotomy approach for safe and complete resection.

Case Presentation

An 8-year-old boy was evaluated for a progressively enlarging, left supraclavicular swelling first noticed three years back. The child had no prior medical or surgical history. Physical examination revealed a firm, non-pulsatile, well-defined, vertically ovoid mass measuring approximately 6 × 4 cm. Its inferior border was not palpable, associated with visible dilation of the superficial left anterior chest wall veins and rightward tracheal deviation.

Routine laboratory tests were within normal limits.

Chest radiography demonstrated a left paratracheal opacity, which is compressing and deviating the trachea to the right.

Contrast-enhanced computed tomography (CT) of the chest and neck revealed a heterogeneously enhancing soft tissue density posterior mediastinal mass of size 8.6 × 10.8 × 7.7 cm, which is extending superiorly into the neck through the thoracic inlet and reaching the lower pole of the thyroid on the left side and pushing the thyroid gland towards the right side. The mass is displacing the trachea and oesophagus to the right and compressing the left lung upper lobe. It is abutting the left common carotid and subclavian arteries without any evidence of invasion.

Image-guided core needle biopsy showed spindle-shaped, bland oval, elongated nuclei; a few foci showed ganglion cells with eccentrically placed nuclei and eosinophilic to amphophilic cytoplasm, favouring a diagnosis of ganglioneuroma.

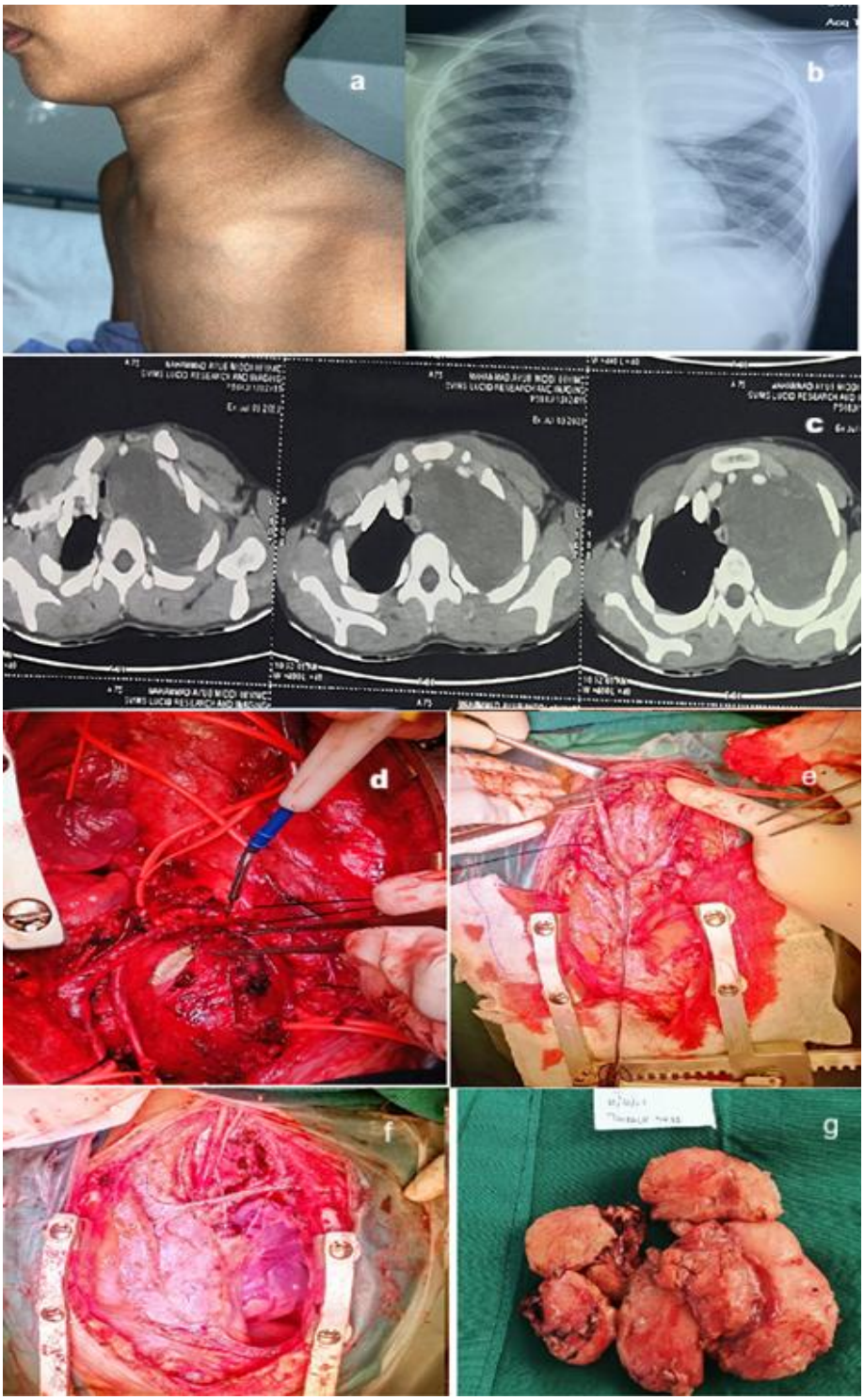

Figure 1: Image showing a. preoperative image of the child, b. Chest X-ray PA view of the child, c. CECT axial images of the child, d-f. intraoperative images of the child, g. Postoperative specimen

Operative Technique

Because of the cranial extension and close relationship with the great vessels, an extended median sternotomy with a left cervical extension along the anterior border of the sternocleidomastoid was performed. The left pleural cavity was entered. A 10 × 8 cm firm mass with irregular borders was identified in the posterior mediastinum with continuity into the cervical region through the superior mediastinum.

The tumour is located posterior to the arch vessels and the innominate vein. Careful dissection was performed to separate the mass from the innominate vein, carotid and subclavian arteries, vagus and phrenic nerves, trachea, and oesophagus. Each structure was looped and protected. Due to restricted working space at the thoracic inlet, the mass was removed completely in a piecemeal fashion, dividing the cervical and thoracic components, while ensuring complete R0 excision.

The patient was extubated in the operating room and shifted to the recovery room. The postoperative recovery was uneventful, and the patient was discharged on postoperative day seven.

Pathologic Findings

Gross examination revealed a well-circumscribed tumour composed of Schwannian stroma with mature ganglion cells of varying size, eccentric hyperchromatic to vesicular nuclei and bright eosinophilic to granular cytoplasm, focally forming clusters. Cystic degeneration, focal myxoid areas, lymphoid aggregates, and dystrophic calcifications were present. No immature neuroblastic elements were identified. These features confirmed the diagnosis of ganglioneuroma.

Postoperative Course

He had regular follow-ups every 3-6 months. On follow-up at 18 months after surgery, he was observed to have no radiological signs of recurrence.

Discussion

Mediastinal neurogenic tumours originate from the neural crest cells and may arise from any sympathetic or peripheral neural structure within the thorax. They occur most commonly in the posterior mediastinum, where they encompass both nerve sheath neoplasms and sympathetic chain–derived tumours. The latter group ranges from benign ganglioneuromas to highly malignant neuroblastomas. [3]

Ganglioneuroma is an infrequent, well-differentiated neurogenic tumour that develops from sympathetic ganglia and is characteristically benign. The posterior mediastinum represents the predominant site, accounting for approximately 60%–80% of cases. The other recognised sites include the adrenal medulla and retroperitoneum, pelvic and sacrococcygeal sympathetic ganglia, the organ of Zuckerkandl, accounting for 10% -15% and, less commonly, the cervical sympathetic chain (5%). Because of their slow growth rate, these tumours typically remain asymptomatic until they attain a considerable size, with symptoms largely determined by compression of adjacent structures. [3][4]

Histologically, ganglioneuromas consist of mature Schwannian stroma interspersed with fully differentiated ganglion cells, without immature neuroblastic components, confirming their benign nature. Their age distribution varies according to tumour subtype; sympathetic ganglia–derived lesions are more often encountered during childhood, while other neurogenic tumours may present later in adulthood. [3]

Diagnostic evaluation typically begins with a chest radiograph, where these tumours appear as a posterior mediastinal opacity. Contrast-enhanced CT serves as the principal imaging modality and delineates the lesion's size, morphology, and relationship to vital mediastinal structures. Magnetic resonance imaging (MRI) plays an important adjunctive role in assessing lesions abutting the spinal canal or when neural foraminal or intraspinal extension is suspected. [5][6]

Definitive management consists of complete surgical excision, which is usually curative owing to the tumour's benign biology and negligible metastatic potential. Resection may be accomplished through single-stage or staged procedures, depending on the anatomic complexity. Particular care is required when dense adhesions are present to avoid injury to major vessels, nerve roots, or adjacent vital structures. [3]

The surgical approach must be tailored to optimise exposure. While posterolateral thoracotomy or thoracoscopic approaches may suffice for small posterior mediastinal lesions, they are inadequate for tumours projecting through the thoracic inlet. Similarly, a cervical incision alone does not allow full control of the intrathoracic portion of the tumour. Therefore, making an extended median sternotomy with cervical extension the most appropriate route. This approach offers superior exposure to the thoracic inlet and allows safe dissection of the carotid arteries, subclavian artery, innominate vein, trachea, and oesophagus.

Recurrence is extremely rare and usually associated with incomplete resections. Our patient remained recurrence-free at 18-month follow-up, consistent with favourable outcomes reported in the literature.

Conclusion

The present case describes a substantially enlarged posterior mediastinal mass with superior extension into the cervical region, a configuration that is particularly challenging to manage in paediatric patients due to the confined anatomy of the thoracic inlet. Lesions traversing this region lie in proximity to vital structures, including the branches of the aortic arch, brachial plexus, phrenic and vagus nerves, trachea, oesophagus, and sympathetic chain, making surgical dissection technically demanding. An extended median sternotomy with cervical extension provides wide and controlled access to the cervicothoracic junction, permitting meticulous mobilisation of the tumour and safeguarding adjacent neurovascular and airway structures. Because these tumours are typically benign with minimal risk of malignant transformation or recurrence, complete surgical excision is usually curative and associated with excellent long-term outcomes.

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