Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Posterior Mediastinal Neuroblastoma in Children: An Unusual Location

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DOI: https://doi.org/10.36347/sjmcr.2025.v13i05.092 | **Received:** 11.04.2025 | **Accepted:** 18.05.2025 | **Published:** 23.05.2025

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Abstract **Case Report**

Neuroblastoma is the most common neurogenic tumor in children, primarily affecting those under 5 years old. It often arises from immature neural crest cells, commonly in the adrenal medulla, but can also occur in the posterior mediastinum. A 4-year-old child presented with cervical lymphadenopathy and a right apical opacity on chest X-ray. CT revealed a lesion in the right posterior-superior mediastinum, confirmed as neuroblastoma upon pathological examination. The patient received neoadjuvant chemotherapy but passed away before surgery. Neuroblastoma is highly heterogeneous, with varied symptoms based on age and tumor location. Imaging is crucial for diagnosis and assessing tumor extent. Posterior mediastinal neuroblastoma is rare in children, presenting diagnostic challenges. Imaging aids detection, while histological analysis confirms the diagnosis

Key words: Neuroblastoma, CT, MRI, Mediastinal.

INTRODUCTION

Neuroblastoma is the most common neurogenic tumor in children, primarily affecting infants and young children. Most cases are diagnosed before the age of 5, although rare instances occur in older children and adults. These tumors originate from immature neural crest cells and can develop in various locations in the body, with the adrenal medulla being the most frequent site. They may also arise along the sympathetic chain and nerve ganglia, including the thoracic ganglia, which are a group of 12 paravertebral sympathetic ganglia located in the posterior mediastinum, although this is less common. Localized neuroblastomas in the posterior mediastinum of young children generally have a more favorable prognosis compared to tumors found in other locations [1,2].

PATIENT AND OBSERVATION

A 4-year-old child with no significant medical history presented with cervical lymphadenopathy evolving over the past two years. A standard chest x-ray revealed a right apical opacity. A chest CT scan, performed with and without contrast for better characterization, showed a lesion in the right posterior-superior mediastinal lymph node lesion that was well-defined with lobulated contours, spontaneously isodense, and exhibited heterogeneous enhancement after contrast injection, delineating areas of necrosis measuring approximately 64 x 49 x 76 mm (ap x t x cc). This was associated with multiple bilateral cervical lymphadenopathies forming a mass in some areas, which was spontaneously heterogeneous, showed calcifications, and had heterogeneous enhancement after contrast injection. Pathological examination confirmed that it is a mediastinal neuroblastoma. The patient received neoadjuvant chemotherapy but passed away before surgery.

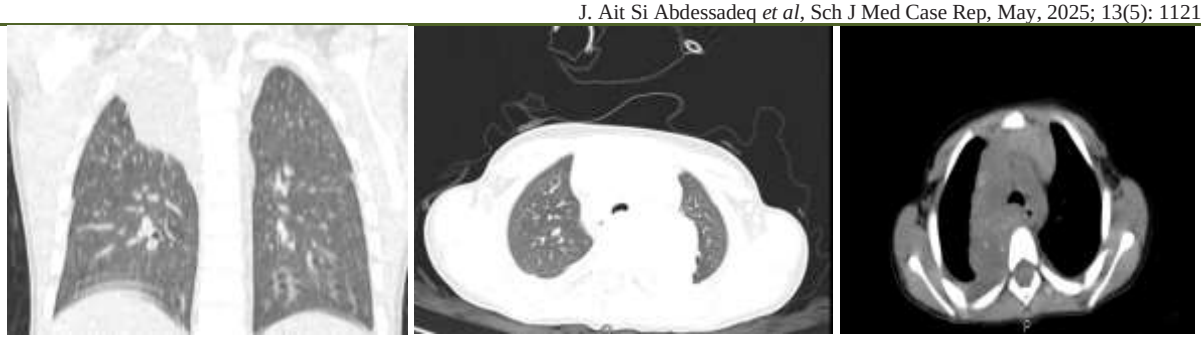

Figure 1: Thoracic CT scan parenchymal and mediastinal window C- shows an isodense process occupying the posterior mediastinum, seat of calcifications

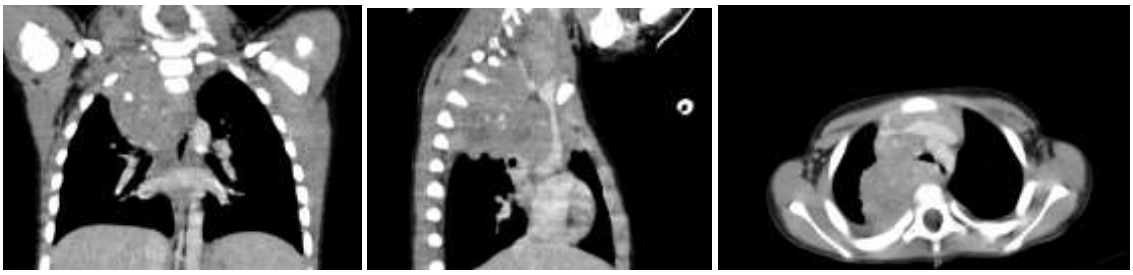

Figure 2: Mediastinal window in coronal, sagittal,and axial sections : shows heterogeneous enhacement of the process

DISCUSSION

Neuroblastoma is a pediatric tumor of the sympathetic nervous system, with initial localization either in the adrenal glands or in the paravertebral or perivascular sympathetic tissue.

Neuroblastoma is characterized by its high heterogeneity; this significant phenotypic variability is based on prognostic factors that determine the distribution of neuroblastomas into different risk groups. These factors primarily include age, stage, and amplification of the n-myc oncogene, along with other parameters such as dna index, histopathological score, serum ldh levels, and the genomic profile of the tumor cells [3,4].

Perinatal localizations in the thoracic, cervical, and pelvic regions are rarer. Although thoracic neuroblastomas account for a minority of all neuroblastoma cases (11% to 26%), they remain the most common mediastinal mass in patients under the age of 2 [2].

The symptoms of mediastinal neuroblastoma in children are varied. This is explained by the variability in the child's age, the tumor's location, size, and whether it is malignant or benign. Symptoms often manifest as compressive, infiltrative, or irritative effects on various anatomical structures in the mediastinum. Depending on the location of the mass, the following syndromes can be distinguished:

- Anterior mediastinal syndrome, which presents with retrosternal pain, superior vena cava syndrome, respiratory signs, or anterior sternal costal anomalies.
- Middle mediastinal syndrome, where the patient primarily experiences cough, dyspnea, or dysphonia.
- Posterior mediastinal syndrome, characterized by dysphagia, radicular pain, sympathetic manifestations, and signs of spinal cord compression.

The presentation of mediastinal neuroblastoma can be variable. It may be asymptomatic, discovered incidentally during a routine radiological examination or while evaluating another often malignant condition, or it may cause compressive symptoms [3,4].

Imaging plays a crucial role in the diagnostic process for mediastinal tumors in children. Chest x-ray is the first-line examination. A mediastinal widening can be seen in 90% of cases on the frontal view, while the lateral view helps localize the mass in the anteroposterior plane. Standard x-rays also assess compressive effects and associated signs such as pleural effusion, calcifications, and bone lesions.

Computed tomography is the preferred examination. It allows for localization and characterization of the mediastinal mass, including its size, consistency (cystic, fatty, solid, calcified), and relationships with surrounding structures. Additionally, CT is useful for detecting secondary locations, guiding biopsies, and assessing the operability of the tumor.

Ultrasound is particularly indicated for young children and can assist in guiding biopsies when the

tumor is accessible. Magnetic resonance imaging is increasingly utilized for mediastinal masses, providing better characterization of lesions, improved assessment of relationships with nearby vascular and cardiac structures, and enhanced evaluation of spinal extension in cases of posterior mediastinal tumors [4,5].

Imaging is essential for localizing neuroblastoma, studying its relationships with adjacent structures, and assessing disease extension. A frontal and lateral chest x-ray is the initial examination requested, which helps clarify a mediastinal opacity that may sometimes show signs of malignancy, such as costal lysis or associated pleural effusion [6].

Contrast-enhanced chest CT provides a better characterization of this opacity, which may appear isodense or hypodense and enhances heterogeneously after contrast injection. It allows for measurement, highlights the presence of characteristic calcifications, and assesses its relationships with nearby organs and vessels. It also checks for lymphadenopathy. While CT can suggest spinal extension, MRI remains the preferred examination. MRI typically shows a relative hypointensity on t1 and hyperintensity on t2 in the bone marrow, often with the classic "hourglass" appearance.

This MRI can also help rule out other differential diagnoses for posterior mediastinal masses. The combination of CT and MRI offers a comprehensive assessment of the tumor and effectively evaluates its extent [2,6].

The extension assessment included a brain CT, abdominal and pelvic CT a spinal MRI and a bone scan [7].

The biological assessment plays an important role and is indicated based on the clinical and radiological findings. A non-specific biological profile is often requested to evaluate the impact of the mass. Measuring urinary catecholamines and performing a bone marrow biopsy can be highly helpful in guiding the etiological diagnosis. The macro and microscopic examination of the biopsy, supplemented by immunohistochemical and cytogenetic studies, is essential for achieving an accurate and complete diagnosis of the cell type [8].

Three non-exclusive methods are available to treat and often cure neuroblastoma:[9]

- Surgery
- Radiotherapy, as the tumor is radiosensitive. The main indications for radiotherapy are high-risk tumors (nmyc amplification and/or stage 4 tumors in children over 12 months).
- Mibg i131 is a metabolic radiotherapy agent for children aged 12 to 18 months with metastatic disease, as well as for patients whose tumors show amplification of the nmyc gene.
- Chemotherapy, both neoadjuvant and adjuvant, is used because neuroblastoma is highly chemiosensitive, and it is also employed to treat metastases.

CONCLUSION

Posterior mediastinal neuroblastoma is a rare location in the pediatric population and often presents an etiological diagnostic challenge. Imaging is the cornerstone for detecting this lesion; however, histological examination provides a definitive etiological diagnosis.

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