Case Report

Ganglioneuroblastoma Presenting as a Pleural-Based Mass in a Young Child: A Rare Case Report

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Abstract

Ganglioneuroblastoma (GNB) represents an intermediate entity in the spectrum of neuroblastic tumors derived from neural crest cells, bridging the malignant neuroblastoma and benign ganglioneuroma. Primary pulmonary involvement by GNB is extremely rare, with very few cases reported in the literature. We report a case of a three-year-old female child who presented with cough and rapid breathing for two to three days. Imaging revealed a well-defined pleural-based mass in the right lower lobe of the lung, initially suspected to be an inflammatory myofibroblastic tumor or pulmonary pleuroblastoma. Histopathological examination demonstrated a biphasic pattern comprising mature ganglion cells and immature neuroblasts within Schwannian stroma, consistent with ganglioneuroblastoma. Immunohistochemistry showed positivity for S-100 and neuron-specific enolase (NSE), confirming neural and Schwannian differentiation. This case underscores the diagnostic challenge of identifying GNB in atypical thoracic locations, where clinical and radiologic findings are nonspecific. Recognition of characteristic histological features and appropriate immunohistochemical profiling are essential for accurate diagnosis and classification. Our report adds to the limited literature on primary pulmonary GNB and emphasizes the need for multidisciplinary evaluation in such rare presentations.

Keywords: ganglioneuroblastoma; neuroblastic tumor; pleural-based mass; lung tumor; pediatric oncology; neural crest; immunohistochemistry

Introduction

Neuroblastic tumors represent a spectrum of neoplasms arising from primordial neural crest cells of the sympathetic nervous system, ranging from the undifferentiated and often aggressive *neuroblastoma*, through *ganglioneuroblastoma* (GNB), to the fully differentiated and benign *ganglioneuroma*. The International Neuroblastoma Pathology Classification (INPC) stratifies these tumors based on histological differentiation, Schwannian stromal development, and mitosis–karyorrhexis index into prognostically relevant categories: *neuroblastoma* (undifferentiated, poorly differentiated, differentiating), *ganglioneuroblastoma* (intermixed or nodular), and *ganglioneuroma* (maturing or mature) [1, 2].

GNB is an uncommon entity, most frequently encountered in pediatric populations, and its occurrence in adults or in atypical sites is exceedingly rare [3]. GNB has rarely been described in the lung with the existence of only a few well-documented cases [4].

Clinically, pulmonary GNB may be asymptomatic or present with nonspecific signs such as cough, chest pain, dyspnea, or even paraneoplastic phenomena including chronic diarrhea due to vasoactive intestinal peptide secretion [5, 6].

Histologically, GNB is defined by the presence of both immature neuroblasts and more differentiated ganglion cells, typically admixed with Schwannian stroma. The "intermixed" subtype lacks grossly visible nodules of *neuroblastoma*, whereas the "nodular" variant shows distinct poorly differentiated foci with prognostic implications [1]. Diagnosis hinges on careful histopathological evaluation, often supported by immunohistochemistry for markers such as chromogranin, synaptophysin, NSE (for neuroblasts), and S-100 (for Schwannian elements) [5].

We present a rare case of primary pulmonary *ganglioneuroblastoma*, confirmed on histopathology, adding to the limited but growing body of literature on this unusual tumor. Our report underscores the diagnostic challenges posed by GNB in atypical locations and highlights the importance of integrating clinical, radiologic, and histologic features for accurate classification and management.

Case Presentation

A three-year-old female child presented to the Pediatrics Medicine Department with complaints of cough and rapid breathing for two–three days. There was no history of fever, abdominal pain, or loose stools. The child had a prior admission for pneumonia at one year of age and a history of multiple nebulizations. On examination, the respiratory rate was 68/min with tachypnea and bilateral wheezing. Chest X-ray revealed cardiomegaly.

Contrast-enhanced CT (CECT) of the chest showed a well-defined, broad pleural-based mass lesion in the right lower lobe (measuring 8.7 × 5.6 cm) with calcifications, displacing the right bronchus anteriorly. Differential diagnoses included inflammatory myofibroblastic tumor and pulmonary pleuroblastoma.

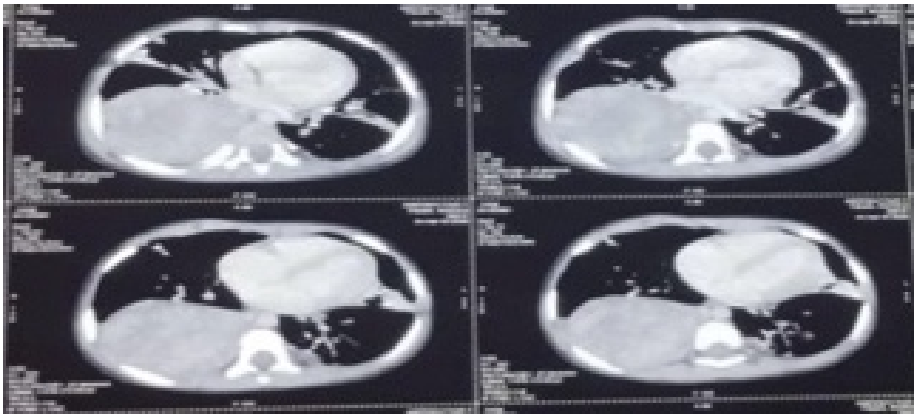

Figure 1: CECT chest shows a pleural based mass lesion.

Gross Examination: Three globular soft tissue pieces were received in the pathology department, measuring 7 cm, 4 cm in diameter, and 2.5 × 2 × 1 cm, with a clinical impression of "?potato tumor."

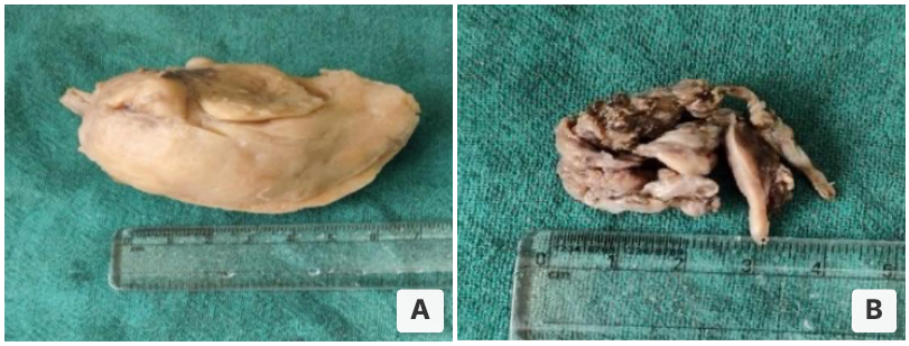

Figure 2: A: Gross specimen received in the pathology department. B: On cut sectioning, homogenous grey yellow areas seen.

Microscopic examination: Histopathology revealed proliferation of oval to spindle-shaped cells with monomorphic, elongated, wavy nuclei and moderate bipolar cytoplasm. Scattered ganglion cells, both singly and in aggregates, were observed. Areas of hemorrhage, nonspecific inflammation, and calcification were also present—suggestive of *ganglioneuroma*. However, the smallest tissue piece showed a monomorphic population of cells with high N:C ratio, round-to-oval nuclei, stippled chromatin, inconspicuous nucleoli, and scant cytoplasm—features consistent with *ganglioneuroblastoma* (this blastemal population was not in other 2 soft pieces). Immunohistochemistry panel was applied using monoclonal antibodies – vimentin with dilution ratio of 1:700, S-100 - 1:200 and NSE- 1:100, showed S-100 positivity and NSE positivity in both ganglion and small round cells.

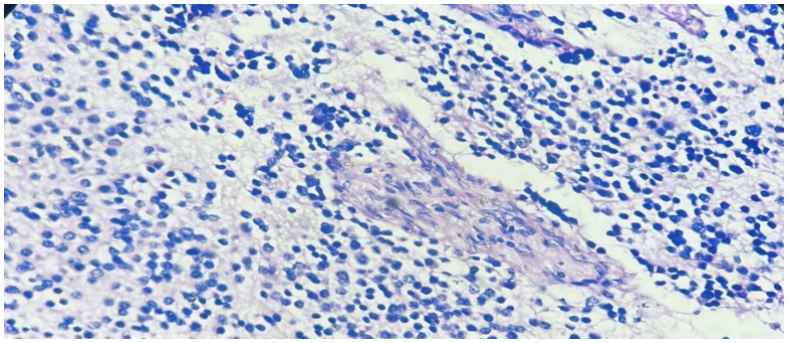

Figure 3: H&E stained slide (at high power) showing monomorphic population of round to oval cells having high N:C ratio, salt and pepper chromatin.

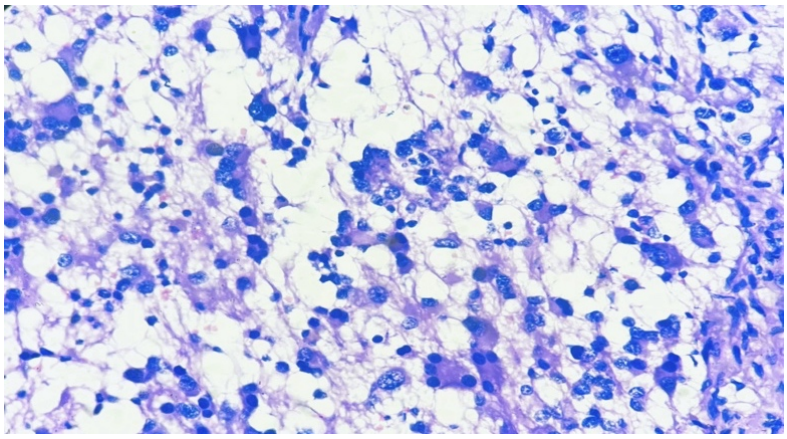

Figure 4: H&E stained slide (at high power) showing singly scattered ganglion cells in a schwannian stroma.

Post operative FISH–*MYCN* AMPLIFICATION was done on formalin fixed paraffin embedded tissue block using Zytolight Spec *MYCN* spectrum green/ CEP 2 (2q11) spectrum orange Dual Color probe. *MYCN* gene:CEP–2 ratio: 1.07 (Positive if ratio is >2.0). Specimen was negative for *MYCN* gene amplification.

Two months postoperative follow up Whole body PET/CT scan was done following intravenous administration of 5.4mCi of F18 FDG and showed post operative changes in posterior mediastinum with mildly FDG avid soft tissue thickening at post operative site along right pleura, right posterolateral chest wall and D6–D7 vertebrae. Few fibrotic bands are seen in bilateral lower lobes. Rest of the lung fields appear normal.

Discussion

Neuroblastic tumors arise from embryonic neural crest cells and are categorized based on maturation into *neuroblastoma*, GNB and *ganglioneuroma*. These represent a spectrum from malignant, immature forms to benign, mature lesions [7]. GNB lies between *neuroblastoma* and *ganglioneuroma* in terms of both differentiation and malignant potential and is classified into four subtypes by the INPC: *neuroblastoma* (Schwannian stroma-poor), *GNB intermixed* (stroma-rich), *GNB nodular* (composite), and *ganglioneuroma* (stroma-dominant) [8, 9].

Histopathologically, GNB is defined by a combination of mature ganglion cells and immature neuroblasts, embedded within Schwannian-rich stroma and neuropil. *GNB-intermixed* features >50% Schwannian stroma and scattered pockets of neuroblastic cells at various maturation stages, differentiating it from *maturing ganglioneuroma*, which has sparse neuroblasts within predominantly mature stroma [10].

Immunohistochemistry supports diagnosis: neurofilament, synaptophysin, chromogranin, and S100 are positive in both GNB and *ganglioneuroma*, highlighting neural and Schwannian differentiation. Importantly, *undifferentiated neuroblastomas* lack Schwannian stroma and background neuropil, possibly explaining their distinct clinical behavior [4].

While GNB usually presents in children as stage I/II tumors, delayed diagnosis in adults due to indolent growth can result in more advanced disease (stage III/IV). Uncommon anatomical presentations, such as in the pleura or lung, can further complicate diagnosis, as seen in our case. CT and MRI assist in evaluating tumor extent, but histopathology remains the gold standard for definitive diagnosis and risk stratification [11].

Conclusion

This case highlights the importance of recognizing histological patterns, Schwannian stroma distribution, and neuronal differentiation in diagnosing GNB, especially in atypical locations and age groups.

Consent for Publication: Written informed consent for publication of the clinical details and images was obtained from the patient's legal guardian, with assurance that the child's identity will remain confidential.

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Competing Interests: None

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