International Journal of Contemporary Pediatrics
Hasvitha G et al. Int J Contemp Pediatr. 2026 May;13(5):800-803
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20261152

Case Report

Thoracic Ewing sarcoma (Askin tumor) presenting with severe respiratory distress in a four-year-old child: a case report

Gurram Hasvitha[1], Mithila D. Mazumder[1], E. Shruthy[1], Rakshitha S. Prasad[1]*, Tanusree Paul[2], Mary V. Jehendran[3]

[1]Department of Pediatrics, Vydehi Institute of Medical Sciences and Research Centre, Bengaluru, Karnataka, India
[2]Department of Oncology, Vydehi Institute of Medical Sciences and Research Centre, Bengaluru, Karnataka, India
[3]Department of Radiology, Vydehi Institute of Medical Sciences and Research Centre, Bengaluru, Karnataka, India

Received: 13 March 2026
Accepted: 09 April 2026

***Correspondence:**
Dr. Rakshitha S. Prasad,
E-mail: rakshithaprasad05@gmail.com

ABSTRACT

Ewing sarcoma is an aggressive malignant small round cell tumor that primarily affects children and adolescents. Thoracic involvement, known as Askin tumor, represents a rare variant arising from the chest wall and may mimic infectious or mediastinal pathologies, often leading to delayed diagnosis. We report the case of a four-year-old female child who presented with progressive left-sided chest wall swelling and severe respiratory distress. Imaging revealed a large soft-tissue mass occupying the left hemithorax with rib destruction and mediastinal shift. Histopathological confirmation was challenging due to inadequate core biopsy sampling; however, fine-needle aspiration cytology suggested a small round cell tumor consistent with Ewing sarcoma. Given the patient's critical clinical condition, neoadjuvant chemotherapy was initiated based on clinicoradiological and cytological findings. Following initiation of chemotherapy with vincristine, doxorubicin, and cyclophosphamide, the child demonstrated rapid clinical stabilization and significant improvement in respiratory distress. This case highlights the diagnostic challenges of thoracic Ewing sarcoma in critically ill pediatric patients and underscores the importance of early imaging, pragmatic diagnostic strategies, and prompt initiation of multimodal therapy.

Keywords: Ewing sarcoma, Askin tumor, Thoracic PNET, Pediatric chest wall tumor, Small round cell tumor, Neoadjuvant chemotherapy

INTRODUCTION

Ewing sarcoma is a highly aggressive malignant small round cell tumor and represents the second most common primary malignant bone tumor in children and adolescents. It accounts for approximately 10–15% of all bone sarcomas.[1]

The Ewing sarcoma family of tumors includes skeletal Ewing sarcoma, extraosseous Ewing sarcoma, primitive neuroectodermal tumor (PNET), and Askin tumor. The disease most commonly occurs during the second decade of life, with an estimated incidence of approximately 2.9 cases per million population.

Thoracic Ewing sarcoma, also known as the Askin tumor, arises from the chest wall and may involve the ribs, pleura, and adjacent lung parenchyma. It is uncommon in early childhood and often presents with nonspecific respiratory symptoms such as cough, dyspnea, and chest pain, which may lead to misdiagnosis.

The current standard treatment involves a multimodal approach consisting of systemic chemotherapy. The Children's Oncology Group protocol includes alternating vincristine, doxorubicin, and cyclophosphamide (VDC) with ifosfamide and etoposide (IE). Five-year survival rates for localized disease range from 75% to 80%, whereas metastatic disease carries a poorer prognosis.

We report a rare case of thoracic Ewing sarcoma in a four-year-old child presenting with life-threatening respiratory distress.

CASE REPORT

A four-year-old female child presented to the pediatric outpatient department with a history of progressive swelling over the left upper chest wall for one year, which had gradually increased in size. The swelling was with worsening chest pain over the preceding one month. In the weeks preceding presentation, the child developed persistent cough and increasing shortness of breath.

On examination, the child was tachycardic (heart rate 120 beats/min), markedly tachypneic (respiratory rate 72 breaths/min), and hypoxic with an oxygen saturation of 84% on room air. A prominent precordial bulge was noted, and the apex beat was shifted to the right side. Chest auscultation revealed markedly reduced air entry over the left hemithorax.

Chest radiography demonstrated complete opacification of the left hemithorax with contralateral mediastinal shift (Figure 1). Contrast-enhanced computed tomography (CECT) of the chest revealed a large heterogeneously enhancing soft-tissue mass arising from the left costal pleura with expansive lytic destruction of the left sixth rib and periosteal reaction. The mass caused compressive atelectasis of the left lung with rightward mediastinal displacement and mild left pleural effusion (Figures 2 and 3).

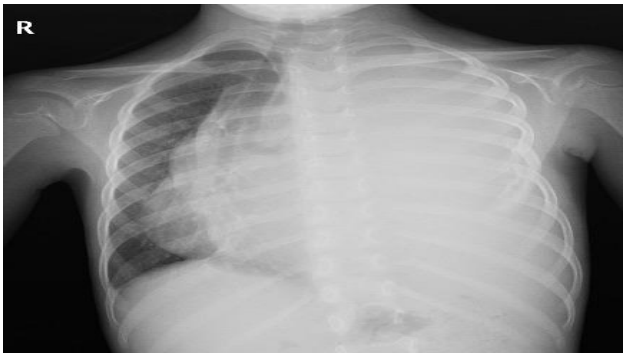

Figure 1: Chest radiograph showing complete opacification of the left hemithorax with contralateral mediastinal shift.

These findings suggested an aggressive malignant chest wall tumor, with Ewing sarcoma being the leading differential diagnosis.

An ultrasound-guided core biopsy was attempted; however, the sample obtained was inadequate and consisted of friable tissue. Considering the patient's unstable clinical condition, a repeat biopsy under general anesthesia was deemed unsafe. Fine-needle aspiration cytology (FNAC) was therefore performed.

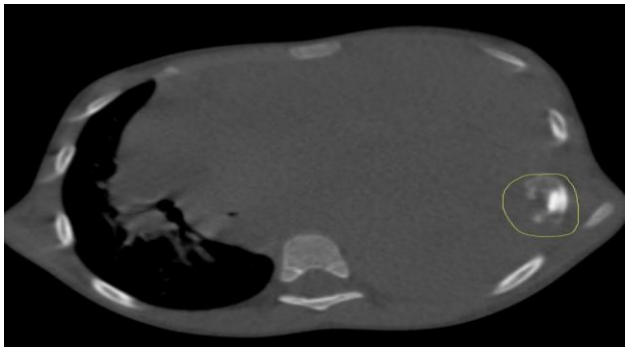

Figure 2: Chest CT (axial bone window) showing a soft-tissue mass arising from the left sixth rib (yellow circle) causing compressive collapse of the left lung.

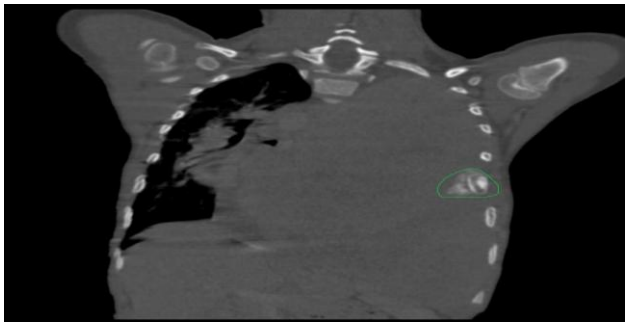

Figure 3: Coronal CT bone window showing a soft-tissue mass arising from the left sixth rib (green circle).

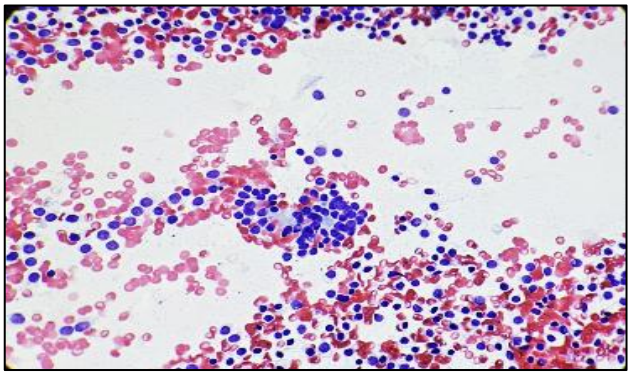

Figure 4: Fine-needle aspiration cytology smear (Papanicolaou stain, ×40 magnification) showing small-to-medium-sized tumour cells with round to oval nuclei, fine chromatin, and scant to poorly defined cytoplasm. Occasional cells show cytoplasmic vacuolations. The cells are dispersed singly and in small clusters with occasional rosette-like formations.

Cytological examination revealed sheets and dispersed small-to-medium-sized round cells with round to oval nuclei, fine chromatin, and scant cytoplasm, with occasional cytoplasmic vacuolations. Rosette-like formations were also observed. These findings were consistent with a small round cell tumor suggestive of Ewing sarcoma (Figures 4 and 5).

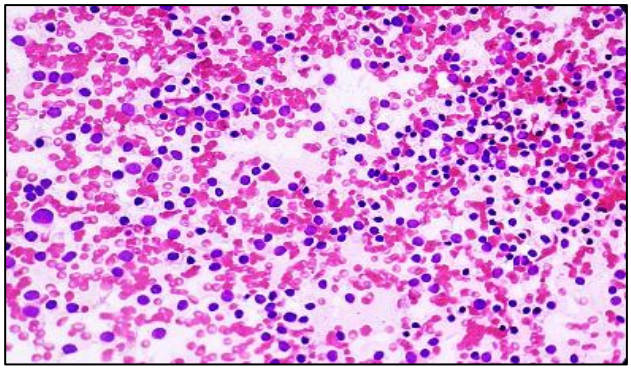

Figure 5: Cytology smear (Hematoxylin and eosin stain, ×40 magnification) showing a cellular smear composed of monotonous round tumor cells, predominantly dispersed singly.

Based on the anatomical location and cytomorphological features, a diagnosis of thoracic Ewing sarcoma (Askin tumor) was established.

Positron emission tomography–computed tomography (PET-CT) demonstrated a metabolically active mass in the left hemithorax with rib involvement and no evidence of distant metastasis (Figure 6).

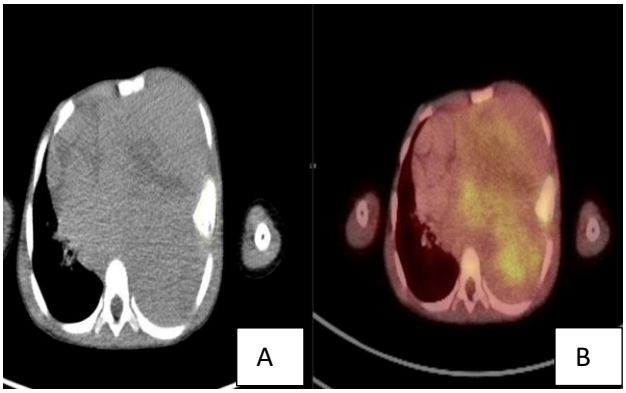

Figure 6 (A and B): PET-CT scan showing a metabolically active mass in the left hemithorax with involvement of the adjacent rib and no evidence of distant metastasis.

During hospitalization, the child developed worsening respiratory distress and was started on high-flow nasal cannula oxygen therapy for respiratory support.

Given the high procedural risk associated with repeat biopsy and the urgent need for treatment, empirical neoadjuvant chemotherapy using vincristine, doxorubicin, and cyclophosphamide (VDC regimen) was initiated.

The patient received two cycles of chemotherapy and demonstrated marked clinical improvement, including stabilization of vital parameters and significant reduction in respiratory distress.

DISCUSSION

Askin tumor is a rare malignant small-cell tumor of the thoracopulmonary region seen mainly in children and adolescents, with a female predominance. It usually arises from the chest wall or peripheral lung, shows frequent local recurrence, and carries a poor prognosis with a median survival of about 8 months. Its exact histogenesis remains uncertain, although a neuroepithelial origin has been suggested by Askin et al.[2]

Askin tumor is considered a rare subtype of primitive neuroectodermal tumor arising from the soft tissues of the chest wall and predominantly affecting children and adolescents. It belongs to the Ewing sarcoma family and is characterized by aggressive behavior. Devi et al emphasized the importance of considering PNETs in the differential diagnosis of chest wall tumors in children for early diagnosis and appropriate management.[3]

Ghewade et al reported that, PNETs are rare aggressive small round-cell tumors arising from primitive nerve cells and belonging to the Ewing sarcoma family. When occurring in the thoracopulmonary region, they are termed Askin tumors and typically arise from the chest wall in children and young adults. They usually present with a chest wall mass and respiratory symptoms, while imaging typically shows a heterogeneous chest wall mass with possible necrosis or pleural involvement. Diagnosis is supported by histopathology and immunohistochemistry, and treatment generally involves multimodal therapy including chemotherapy, surgical resection, and radiotherapy.[4]

Similarly, Abidi et al described Askin tumor as an aggressive tumor of the Ewing sarcoma group that commonly presents as a painful chest wall mass with possible rib or pleural involvement. Diagnosis is confirmed by histopathology and immunohistochemistry, and treatment involves multimodal therapy including chemotherapy, surgery, and radiotherapy, although prognosis remains guarded.[5]

Radiologically, Winer-Muram et al noted that Askin tumors typically appear as large heterogeneous chest wall masses with pleural involvement and possible rib destruction due to areas of hemorrhage and necrosis. CT and MRI are complementary in evaluating disease extent, with MRI better for assessing chest wall muscle invasion and CT more useful for detecting pulmonary metastases.[6]

Our case is notable because it describes thoracic Ewing sarcoma (Askin tumor) occurring in a much younger patient (four years old), whereas most reported cases involve older children and adolescents. In addition, our patient presented with severe, life-threatening respiratory compromise due to a massive chest wall tumor causing near-complete opacification of the hemithorax and significant mediastinal shift. Unlike many reported cases in which diagnosis is confirmed through core biopsy and

immunohistochemistry, the unstable clinical condition in our patient limited invasive procedures, and the diagnosis was primarily established using FNAC cytomorphology and radiological findings. Furthermore, early empirical neoadjuvant chemotherapy was initiated because of respiratory distress, resulting in rapid clinical improvement after two cycles. This highlights the importance of prompt therapeutic intervention even when complete tissue diagnosis is challenging.

CONCLUSION

Thoracic Ewing sarcoma (Askin tumor) is a rare but aggressive pediatric malignancy that may initially mimic common respiratory conditions. This case underscores the importance of maintaining a high index of suspicion in children presenting with persistent chest wall swelling and respiratory distress.

When conventional tissue biopsy is contraindicated due to clinical instability, FNAC in conjunction with imaging findings may provide adequate diagnostic confidence to initiate timely chemotherapy. Early multidisciplinary management is essential for improving survival and preventing rapid clinical deterioration.

ACKNOWLEDGEMENTS

The authors thank Dr. Shilpa L., Professor, Department of Pathology, for her assistance with pathological evaluation, and Dr. Dinakar Prithviraj, Professor and Head, Department of Pediatrics, Vydehi Institute of Medical Sciences and Research Centre, Bengaluru, for their support in the management of this case.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Esiashvili N, Goodman M, Marcus RB Jr. Changes in incidence and survival of Ewing sarcoma patients over the past 3 decades: Surveillance Epidemiology and End Results data. J Pediatr Hematol Oncol. 2008;30(6):425-30.
2. Askin FB, Rosai J, Sibley RK, Dehner LP, McAlister WH. Malignant small cell tumor of the thoracopulmonary region in childhood: a distinctive clinicopathologic entity of uncertain histogenesis. Cancer. 1979;43(6):2438-51.
3. Devi LP, Kumar R, Kalita JP, Khonglah Y, Handique A. Locally advanced Askin tumor in a child: a rare case report and review of the literature. Indian J Surg Oncol. 2015;6(3):288-91.
4. Ghewade P, Shukla S, Vagha S, Ghewade B, Gadkari P. Askin tumor: a report of a rare case. Cureus. 2024;16(6):e63345.
5. Abidi K, El Baz M, El Houdzi J. Askin tumor in children: two case reports. SAS J Surg. 2023;9(5):469-71.
6. Winer-Muram HT, Kauffman WM, Gronemeyer SA, Jennings SG. Primitive neuroectodermal tumors of the chest wall (Askin tumors): CT and MR findings. AJR Am J Roentgenol. 1993;161(2):265-8.

Cite this article as: Hasvitha G, Mazumder MD, Shruthy E, Prasad RS, Paul T, Jehendran MV. Thoracic Ewing sarcoma (Askin tumor) presenting with severe respiratory distress in a four-year-old child: a case report. Int J Contemp Pediatr 2026;13:800-3.