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Case Report

A rare presentation of extraosseous ewing sarcoma in a 14-year-old boy with pleural involvement

Rajesh Raju[1], Sulbha M. Arya[2], Ipshita Raju[3], Banuru Prasad[4*], Sanjeev Saini[5], Sharath Burugina Nagaraja[6]

[1]Director General Health Services, Swasthya Bhawan, Haryana, India
[2]Dept. of Radiotherapy, Pandit Bhagwat Dayal Sharma Post Graduate Institute of Medical Sciences or PGIMS, Rohtak, Haryana, India
[3]Independent Researcher, Panchkula, Haryana, India
[4]Independent Consultant, Bengaluru, Karnataka, India
[5]Independent Consultant, New Delhi, India
[6]Dept. of Community Medicine, ESIC Medical College and PGIMSR, Bengaluru, Karnataka, India

Abstract

Extraosseous Ewing Sarcoma (EES) is a rare subtype of Ewing Sarcoma which originates usually in soft tissues when compared to bones. We present the case of a 14-year-old boy presenting with an atypical painless swelling on the left side of the chest and pleural effusion. On evaluation that included clinical, histopathological and radiological led to a diagnosis of EES. We make a case to emphasize the importance of considering EES in differential diagnosis of thoracic masses particularly among paediatric patients and highlights the diagnostic challenges encountered with such rare presentations.

Keywords: Extraosseous ewing sarcoma, Chest, Tumours, Sarcoma

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1. Introduction

The first description of "Ewing Sarcoma" was made by James Ewing during 1921.[1] It was described as a malignant small round blue cell tumour primarily affecting the paediatric and adolescent age group. It commonly involves long bones; however, extraosseous presentations, such as Extraosseous Ewing Sarcoma (EES), are rare. Recent cancer registry data from India (2021) indicate that 3.4% of all pediatric cancers are Ewing Sarcoma.[2] The presentation of EES with pleural involvement quite often is misdiagnosed due to its rarity. We describe a case of 14-year-old boy presenting with an insidiously growing chest wall mass and pleural effusion.

2. Case Presentation

Apparently, healthy 14-year-old boy presented with a progressively enlarging lump on the left side of the chest with four months duration. The lump was initially painless, but, became tender over the past ten days. On physical examination, the boy appeared pale, with a body mass index appropriate for his age. The mass, located on the left anterolateral chest wall, measured approximately twenty centimetre in diameter, was firm, non-reducible, non-tender, and immobile, with normal overlying skin temperature and no visible vascularity. Tracheal deviation was towards the right, along with reduced tactile vocal fremitus, dullness on percussion, and diminished breath sounds were confirmed on the left side. (**Figure 1**)

*Corresponding author: Banuru Prasad
Email: drprasadbm.icf@gmail.com

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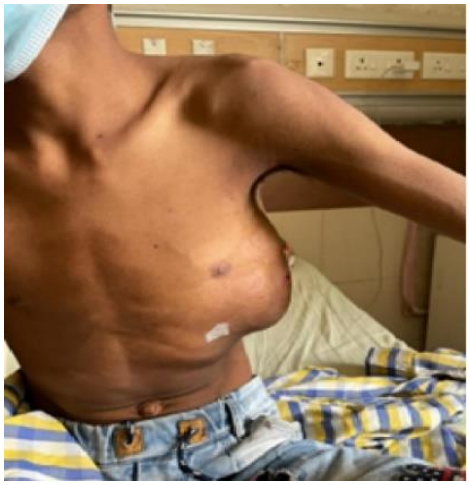

Figure 1: Physical appearance of the sarcoma

3. Laboratory Investigations

The initial laboratory tests, including fine-needle aspiration cytology (FNAC) had ruled out infectious causes such as tuberculosis; while, culture and hematological studies showed no signs of infection.

4. Radiological Findings

A chest radiography revealed a massive pleural effusion on the left side, with mediastinal and cardiac displacement to the contralateral side and soft tissue swelling suggestive of a cold abscess. Contrast-enhanced computed tomography (CECT) of the thorax demonstrated a heterogeneously enhancing soft tissue mass occupying the entire left chest cavity. The mass measured 20 centimetre and 15 centimetres in length and breadth. It also had multiple hypodense areas, and caused near-total collapse of the left lung with significant mediastinal shift. The lesion had a broad base abutting the adjacent pleura. (**Figure 2**- **Figure 4**)

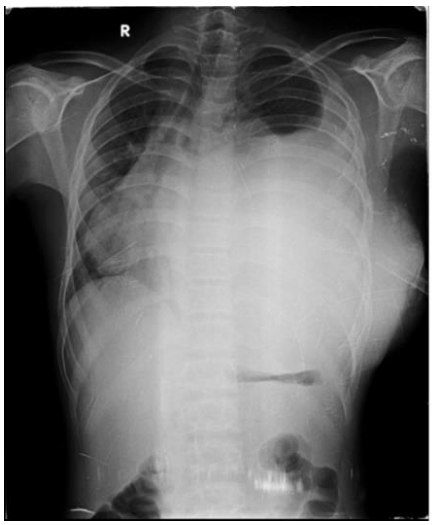

Figure 2: Radiography of the sarcoma

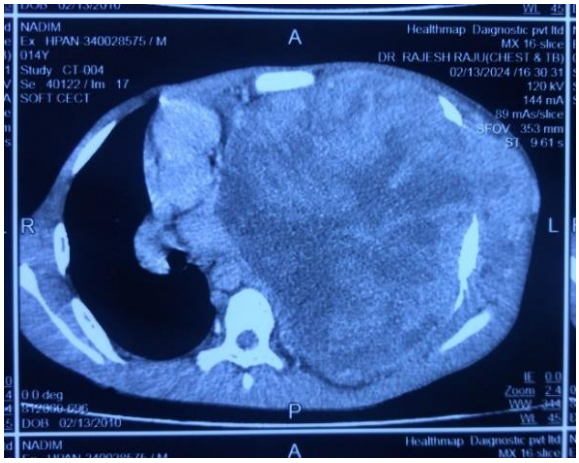

Figure 3: CECT scan of sarcoma – horizontal section

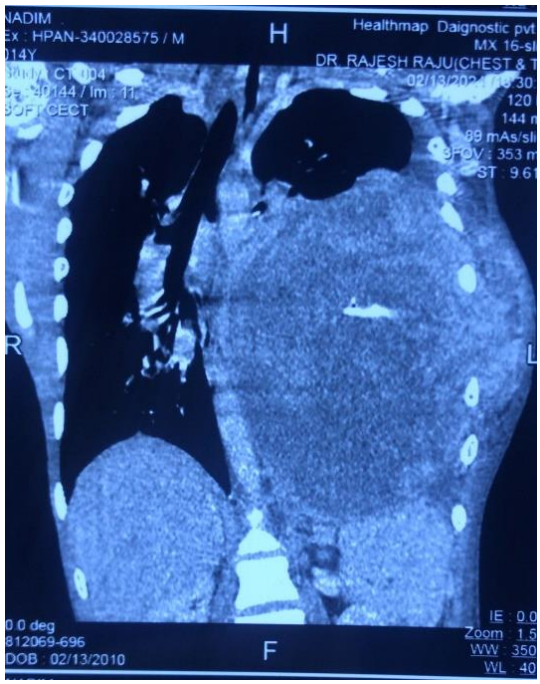

Figure 4: CECT scan of sarcoma – vertical section

5. Histopathology

The FNAC revealed malignant small round cells with round-to-oval nuclei, scant cytoplasm, and fine granular chromatin, consistent with a malignant small round cell tumour. The section in **Figure 5** shows diffuse sheets of small round tumour cells having hyperchromatic nuclei and scanty cytoplasm (H & E, X 400). Similarly in **Figure 6**, the section shows diffuse membranous positivity for CD 99.

Based on radiological and histopathological findings, a provisional diagnosis of EES was made. The patient was referred for immunohistochemical (IHC) analysis, including markers such as CD99 and FLI1, and confirmatory fluorescence in situ hybridization (FISH) testing. (**Figure 5**)

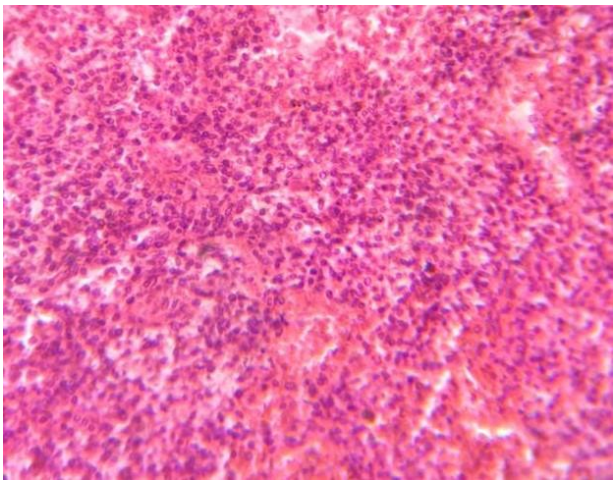

Figure 5: Histopathological section of the sarcoma

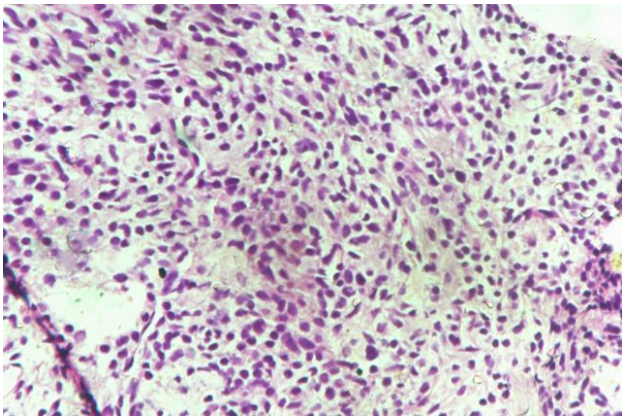

Figure 6: Section shows diffuse sheets of small round tumour cells having hyperchromatic nuclei and scanty cytoplasm (H&E x 400)

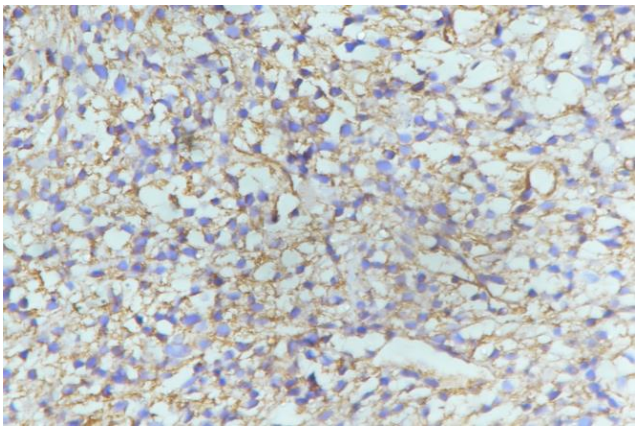

Figure 7: Section shows diffuse membranous positivity for CD 99 (CD 99 x 400)

6. Discussion

Based on the signs and symptoms of painless thoracic mass and massive pleural effusion, raising initial differential diagnoses such as empyema thoracis with cold abscess or tuberculosis, were ruled out based on imaging and cytological findings.

The EES is a rare tumour within the Ewing Sarcoma Family of Tumours (ESFT), including Ewing Sarcoma of bone, peripheral primitive neuroectodermal tumour (pPNET), Askin tumour, and atypical Ewing Sarcoma.[3,4] The diagnosis of EES requires a combination of radiological, histopathological, and molecular findings. In this case, the characteristic small round blue cells with neuroendocrine features and radiological evidence of a massive thoracic lesion supported the diagnosis of EES. Confirmation through IHC markers (CD99, FLI1) and FISH testing is critical to differentiate EES from other small round cell tumours.[5-7]

The primary EES involving the lung or pleura is exceedingly rare, with fewer than 20 cases reported in the literature, often classified as Askin tumours.[8] The rarity of EES with pleural involvement highlights the need for high clinical suspicion and a multidisciplinary approach for diagnosis and management.

7. Conclusion

We present the diagnostic challenges of EES, particularly in rare presentations involving the pleura. Comprehensive diagnostic evaluation that includes imaging and histopathological analysis is essential for accurate diagnosis and appropriate management. Early diagnosis of such rare presentations provides an opportunity to improve outcomes for affected patients.

8. Source of Funding

None.

9. Conflict of Interest

None.

10. Acknowledgements

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