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A Rare Case of Extraskeletal Ewing's Sarcoma Presenting as Hemorrhagic Pleural Effusion

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Abstract:

- ***Background:***
Extraskeletal Ewing's sarcoma (EES) is a rare and aggressive small round cell tumor, primarily affecting soft tissues outside the skeletal system. Pulmonary involvement is uncommon, and hemorrhagic pleural effusion as an initial presentation is exceedingly rare. Due to its rarity, EES is often misdiagnosed, particularly in endemic regions where tuberculosis is prevalent.

- ***Case Presentation:***
We report a case of a 33-year-old female with no known comorbidities who presented with progressive breathlessness, chest pain, and chronic dry cough for three months. She also reported intermittent fever and unintentional weight loss. Initial imaging suggested a right-sided pleural effusion, which was hemorrhagic on thoracentesis. Pleural fluid analysis revealed an exudative effusion with a lymphocyte-predominant cytology but was negative for malignancy. A contrast-enhanced CT (CECT) thorax demonstrated a large heterogeneously enhancing mass involving the parietal pleura, with adjacent rib erosion. Pleural biopsy and immunohistochemistry confirmed the diagnosis of EES. The patient was initiated on combination chemotherapy, resulting in a significant reduction in tumor size on follow-up imaging.

- ***Conclusion:***
EES should be considered in young adults presenting with unexplained hemorrhagic pleural effusions. Early recognition and histopathological confirmation are critical for timely intervention. Multimodal therapy, including chemotherapy, surgical excision, and radiotherapy, remains the mainstay of treatment.

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I. INTRODUCTION

Extraskeletal Ewing's sarcoma (EES) is a rare entity of the Ewing sarcoma family of tumors (ESFT), typically arising in the soft tissues without bony involvement [1]. Unlike skeletal Ewing's sarcoma, which primarily affects the long bones of adolescents, EES occurs in a wide age range and often involves the trunk, retroperitoneum, or extremities [2]. Pulmonary involvement is rare, and its presentation as a hemorrhagic pleural effusion is even more unusual [3].

Given the overlap in clinical and radiological features with pulmonary tuberculosis, malignant effusions, and mesothelioma, diagnosing EES can be challenging [4]. Immunohistochemistry plays a crucial role in differentiation, with CD99 positivity being a hallmark of Ewing sarcoma [5].

We report a rare case of EES presenting as a massive hemorrhagic pleural effusion in a young female, initially misdiagnosed as tuberculosis.

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II. CASE PRESENTATION

A 33-year-old female homemaker with no known comorbidities presented with progressive breathlessness, dry cough, and right-sided chest pain for three months. She also reported intermittent fever for one month and a 10 kg unintentional weight loss. There was no history of hemoptysis, night sweats, or tuberculosis exposure.

A. Clinical Examination

- Moderately built and nourished
- Vital signs: PR - 97 bpm, BP - 120/80 mmHg, SpO2 - 89% on room air, RR - 26 breaths/min
- Respiratory system: Reduced chest movements on the right side, diminished breath sounds, and dullness on percussion

B. Investigations

➢ *Chest X-ray:*

- Right-sided massive pleural effusion

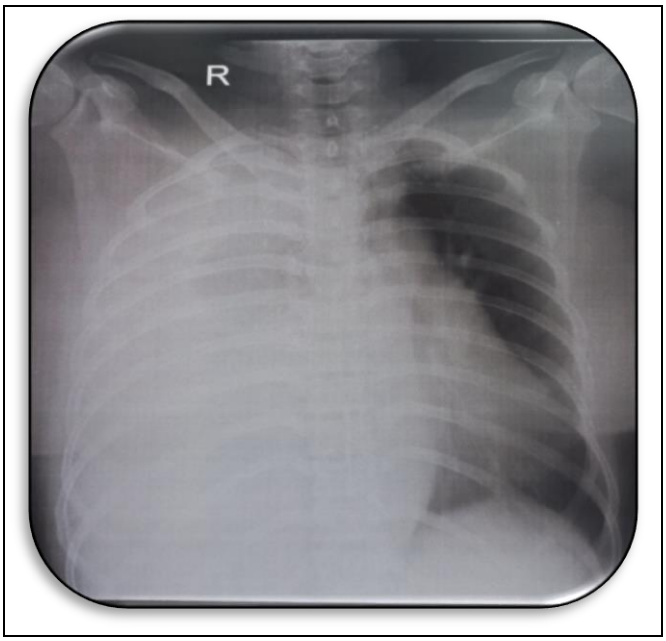

Fig 1(a): CXR at Presentation s/o Right Sided Gross Pleural Effusion

➢ *Pleural Fluid Analysis:*

- Hemorrhagic fluid (~2.5 L drained)
- ADA: 21.5 U/L (borderline)
- LDH: 209.6 U/L
- Protein: 5.6 g/dL
- Cytology: Predominantly lymphocytic, negative for malignant cells

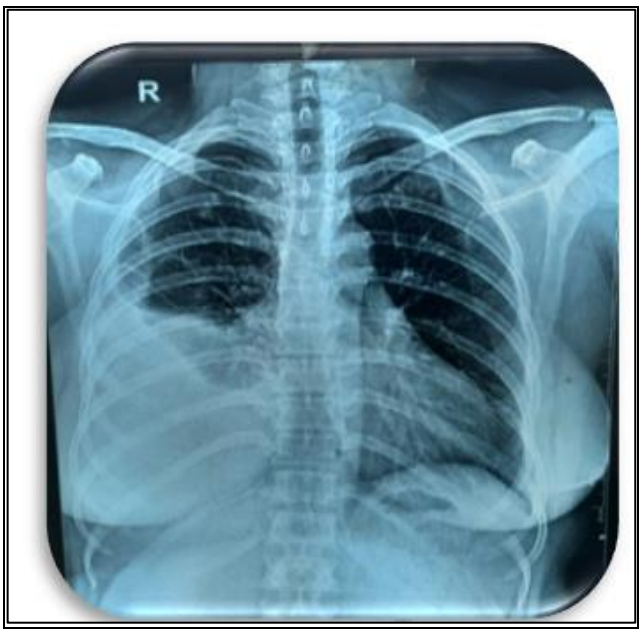

Fig 1(b): CXR Post Therapeutic Tapping

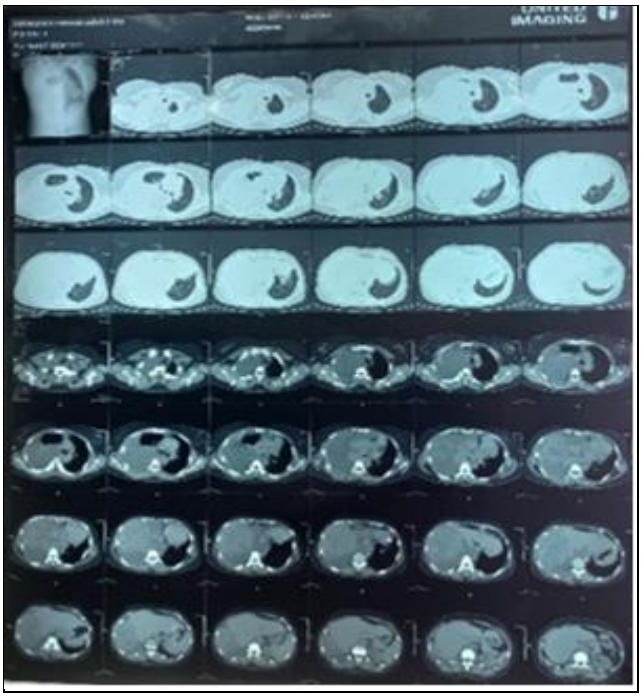
Fig 2: CECT Showing Heterogeneously Enhancing Mass Lesion Involving Parietal Pleura Right Side with Effusion

C. CECT Thorax:

Large heterogeneously enhancing multilobulated mass involving the parietal pleura (12 x 11 x 15 cm) with rib erosion and right-sided pleural effusion (Figure 2)

➢ *Pleural Biopsy and Immunohistochemistry:*

- CD99 and Vimentin: Positive
- Ki-67 proliferation index: 40%
- CD45, CK, Synaptophysin, Chromogranin: Negative

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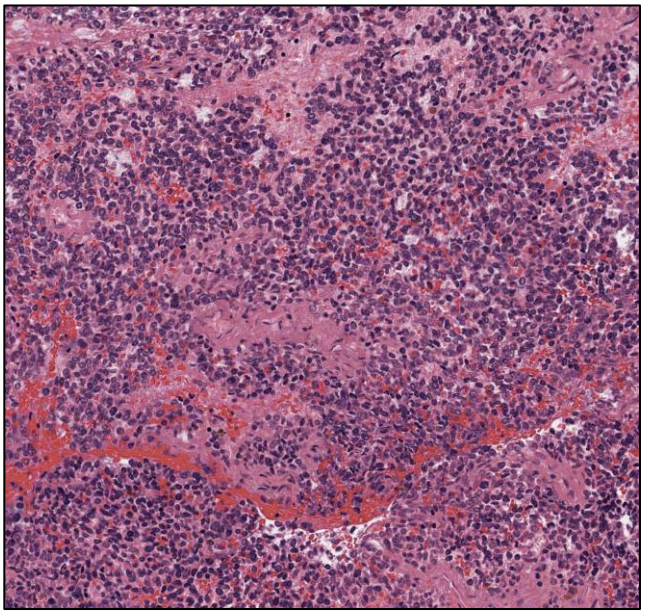

Fig 3: Small Round Blue Cell Tumor

D. Diagnosis

Extraskeletal Ewing's sarcoma involving the pleura with hemorrhagic pleural effusion.

III. TREATMENT AND OUTCOME

The patient was initiated on multi-agent chemotherapy with Vincristine, Adriamycin, Cyclophosphamide (VAC) alternating with Ifosfamide and Etoposide (IE) every three weeks. After four cycles of chemotherapy, a repeat CECT thorax showed a significant reduction in tumor size (7.7 x 5.1 x 7.2 cm). She was planned for wide local excision with segmental resection of the affected rib followed by adjuvant radiotherapy.

IV. DISCUSSION

Extraskeletal Ewing's sarcoma is an aggressive malignancy that poses a diagnostic challenge due to its nonspecific clinical presentation. While pleural effusions have been reported in cases of metastatic Ewing's sarcoma, primary pleural involvement with hemorrhagic effusion is exceptionally rare [6].

Histopathological examination with immunohistochemistry is crucial for diagnosis. CD99 positivity is a hallmark of Ewing's sarcoma, and genetic confirmation via EWSR1 translocation testing is recommended [7].

V. CONCLUSION

Extraskeletal Ewing's sarcoma should be considered in cases of unexplained hemorrhagic pleural effusion. Early histopathological evaluation and immunohistochemical markers are essential for timely diagnosis. A multidisciplinary approach, including chemotherapy, surgery, and radiotherapy, provides the best outcomes.

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