Case of Mediastinal Ewing’s Sarcoma/Primitive Neuroectodermal Tumor Presenting as Pleural Effusion

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I. Introduction

Ewing sarcoma is a small round blue cell tumour with regular sized primitive appearing cells. It is closely related to the soft tissue tumours pPNET, Askin tumour and neuroepithelioma, which collectively are referred to as Ewing sarcoma family of tumours (ESFT). They share not only microscopic appearances but also demonstrate a non-random (11;22)(q24;q12) chromosome rearrangement.

II. Material

A young male aged 22yr presented with complaints of fever, breathlessness, dry cough, left sided chest pain since 1 month to JMJ Medical College. Patient was subjected to Chest x ray and CECT (16 slice TOSHIBA machine) and blood investigations.

III. Examination

On physical examination vitals were stable, signs of left pleural effusion with trachea shifted to right side. Blood Investigations shows Complete Blood Count, Renal Function Test, Liver Function Test- Within Normal Limit, sputum for A F B-negative. CXR- Signs of moderate pleural effusion, mediastinum shifted to the right side; Pleural fluid analysis: straw-colored, [TC– 170/cumm, L- 80%, N-15%, ADA-122 IU, protein-3.7g/dl, sugar- 86 mg/dl] malignant cells – negative.

CECT THORAX

CT machine used was 16 slice TOSHIBA machine
CT findings- Well-defined heterogeneous soft tissue density mass lesion noted predominantly within the anterior mediastinum measuring 15x13x10 cm making obtuse angles with anterolateral the chest wall causing medial displacement of the pleural and erosion of the left 5th rib with minimal pleural effusion. Lesion shows significant enhancement on contrast study. F/S/O Ewing’s Sarcoma of chest wall/ primitive neuroectodermal tumor (PNET). Other differentials include chondrosarcoma (malignant tumour of the rib seen in elderly age group with calcification) and osteosarcoma; less likely pleural based tumour as pleura is displaced medially.
Axial CECT images showing: Well-defined heterogeneoussoft tissue density mass noted measuring 15x13x8cm making obtuse angles with anterior lateral chest wall causing medial displacement of the pleural and erosion of 5th rib with minimal pleural effusion.


IV. Management

Patient underwent surgery tumor was excised completely from anterior chest wall and subjected to Histopathology examination of mass lesion shows: Small round cells with round nuclei containing fine chromatin and scant eosinophilic cytoplasm with indistinct cell borders. The cells are arranged in lobules separated by thin fibrovascular septa. Occasional rosette formation made out. Tumor shows capsular invasion, skeletal muscle and bony infiltration F/S Op Ewing’s sarcoma /primitive neuroectodermal tumor.
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Post operative specimen

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**SURGICAL PATHOLOGY REPORT**

**Patient Name:** Mr. Marti

**Address:**

**Age/Sec:** 22 yrs/Male

**Physician/Surgeon:** Dr. H.L. Subha Ray

**Unit:**

1685/1685/2015

**MICROSCOPIC IMPRESSION:** Features are suggestive of "Ewing’s sarcoma / primitive neuroectodermal tumor – Anterior mediastinum".

- Please note: The portion of the rib sent is destroyed with the presence of tumour cells, suggesting the possibility of Ewing’s Sarcoma arising from the rib and occupying the anterior mediastinum.

**Advised:** Immunohistochemistry: CD-99 marker for confirmation of Ewing’s Sarcoma.

1685/2015

**MICROSCOPIC DESCRIPTION:** Sections studied from mass (A to J) show an infiltrating tumor composed of uniform small round cells with round nuclei containing floe chromatin and scant eosinophilic cytoplasm with indistinct cell borders. The cells are arranged in lobules separated by thin fibro vascular septa. Occasional rosette formation made out. Peri-tumour areas of necrosis +. Tumor cells have perivascular distribution. Stromal hyalinization +. Mitoses 0-1/local. Tumor shows vascular invasion, skeletal muscle and bony infiltration.

**MACROSCOPY:** Received nodular tissue attached to bony fragment ms 15x13x8cm. E/n lobulated & glistening. Other surface attached to elongated piece of bone shows few areas of hemorrhage. C/s grey white, variegated appearance. Also received in same bottle, multiple irregular tissue bits all together ms 16.3x12x2cm, dark brown to pale brown. C/s pale white.

1685/2015

**MICROSCOPIC DESCRIPTION:** Sections studied from lung nodules (A,B,C) show presence of above described tumor.

**MACROSCOPY:** Received multiple nodular tissue bits altogether ms 4.5x4x1cm. C/s pale white.

**Clinical Diagnosis:** Anterior mediastinum tumor.

**Tissue sent:** Mediastinal tumor + lung nodule.

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Corrected by

[Signature]

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V. Observation

A young male patient presented with fever, cough with a left pleural effusion. CECT thorax revealed mass lesion, patient underwent USG guided FNAC report S/O PNET. Patient underwent surgery, anterior mediastinal mass arising from chest wall resected with rib fragments subjected to HPE reported as “Ewing’s sarcoma/primitive neuroectodermal tumor-Anterior mediastinum” The differential diagnosis for chest wall tumor are

<table>
<thead>
<tr>
<th>Benign Soft tissue</th>
<th>Skeletal (rib cage)</th>
<th>Malignant Soft tissue</th>
<th>Skeletal (rib cage)</th>
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<tbody>
<tr>
<td>Haemangioma</td>
<td>Fibrous dysplasia (MC)</td>
<td>Rhabdomyosarcoma (MC)</td>
<td>Chest wall metastases (MC)</td>
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<td>Lymphangioma</td>
<td>Aneurysmal bone cyst</td>
<td>Ewing’s sarcoma; including Askin tumor/pPNET</td>
<td>Myeloma</td>
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<tr>
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<td>Giant cell tumor</td>
<td>Ganglioneuroblastoma</td>
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<td>Osteochondroma</td>
<td>Angiosarcoma</td>
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<td>Paraganglioma</td>
<td>Mesenchymal hamartoma of chest wall</td>
<td>Malignant fibrous histiocytoma</td>
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VI. Conclusion

Ewing’s sarcoma is highly malignant primary tumor. The tumor is derived from red bone marrow, most frequently, it is observed in children and adolescents aged 4-15 years and rarely develops in adults older than 30 years. Ewing sarcoma is the second most malignant tumor in young patients and it is the most lethal bone tumor. Males are affected than females.

Most frequently, the tumor is diagnosed as a monostotic lesion in the metaphysis or diaphysis of the long bones of the extremities. The tumor also may occur, although less frequently, in the pelvic area, ribs, and scapulae.

References:

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