A RARE CAUSE OF CHEST PAIN IN CHILDREN

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HISTORY

Syed, a 6-year-old male child, presented with a history of trauma to the right side of the chest 3 months back. He consulted a pediatrician on the 5th day for persistent pain and swelling at the same site. Given NSAIDS for 5 days, he got some relief only to experience the pain and swelling reappear upon stoppage of treatment.
ON FOLLOW UP

- No previous history of fever or chills
- No history of cough, cold, fever, breathing difficulty, hemoptysis, or bony pains elsewhere
- History of Koch's contact + (on AKT) No lymphadenopathy
- Immunized as per NIS
- Vitals stable
- X-ray and USG chest was advised
USG CHEST (13/3/13)

IMPRESSION:
A c/o trauma now showing possibly loculated hematoma in extra pleural soft tissue of right anterior chest wall. Patient treated with NSAID and antibiotics for 7 days.
On reappearance of symptoms -- Advised repeat X ray and HRCT chest and referred to our hospital.

H/o not gaining weight since last 8 to 10 months

Vitals stable

Anthro:
- Wt - 14.5 kg (exp 21 kg)
- Ht - 104 cm (exp 116 cm)

Both less than 3rd centiles.

RS: AE decreased right IAA, IMA, ISA. Dull note on same area.

Other systemic examination – Not contributory.
Homogenous opacity in RLZ
Thinned out and partly eroded anterior aspect of right 7th rib
Possibly loculated hematoma in RLZ with associated osteomyelitic changes
DIFFERENTIAL DIAGNOSIS

Ø EWING'S FAMILY OF TUMOUR
Ø OSTEOCYTOMA ? TUBERCULOUS ETIOLOGY
Ø OSTEOSARCOMA, BONE CYST
Ø EOSINOPHILLIC GRANULOMA
Ø ENCHONDROMA, HEMANGIOMA, METASTATIC NEUROBLASTOMA
Ø FIBROUS CORTICAL DEFECT
Ø METASTASIS
HRCT FINDINGS

- Right 7th rib erosive destruction noted in MA, AA regions with secondary periosteal reaction
- Saucerisation seen
HRCT FINDINGS

SUN-BURST APPEARANCE
SPICULATED PERIOSTEAL REACTION
HRCT FINDINGS

• RLL anterior basal superior segment and lateral basal segment air space opacification
• RML plate atelectasis
• Minimal pleural effusion

IMPRESSION:
-- ? Neoplastic (Ewings sarcoma, PNET) ?
-- ? Tuberculous etiology
7th RIB BIOPSY: IMPRESSION

- Metastatic carcinoma: Round to polygonal tumour cells. Mitosis ++
- Differentials:
  - Small cell carcinoma lung
  - Lymphoma
  - Neuroendocrine carcinoma
- Suggested: Immunohistochemistry

Suggested
**IMMUNOHISTOCHEMISTRY**

<table>
<thead>
<tr>
<th>MARKERS</th>
<th>REACTIVITY</th>
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</thead>
<tbody>
<tr>
<td>CD99 (MIC2)</td>
<td>Strong membrane +ve reaction</td>
</tr>
<tr>
<td>KERATIN</td>
<td>+ve reaction</td>
</tr>
<tr>
<td>SYNSPTOPHYSIN / NSE</td>
<td>+ve</td>
</tr>
<tr>
<td>CHROMOGRANIN</td>
<td>+ve</td>
</tr>
<tr>
<td>CD68</td>
<td>faint background positivity</td>
</tr>
<tr>
<td>LCA / VIMENTIN / CD57</td>
<td>negative reaction</td>
</tr>
<tr>
<td>EMA</td>
<td>faint +ve reaction</td>
</tr>
<tr>
<td>Ki 67 - 60% tumour cells</td>
<td>show +ve reaction</td>
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</tbody>
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**IMPRESSION**

ASKIN'S TUMOUR (ESFT)
BONE SCAN

Increased uptake is seen in the 6th and 7th ribs.

BONE MARROW ASPIRATION

Negative study.
CBC --
Hb 10.8
PCV 34%
TLC 11050
N52
L40
E4
MONO 3
BASO 1
platelets 3.5 lac
RBC 4.49/cumm

ESR --
100

MONTUOX -- NEGATIVE

LDH --
464 U/L (200 to 400 U/L)
TREATMENT

- Started on Chemotherapy - VAC/IE protocol. Alternate cycles.
  - V - VINCRISTINE
  - A - ADRIAMYCIN (DOXORUBICIN)
  - C - CYCLOPHOSPHAMIDE
  - I - IFOSFAMIDE
  - E - ETOPOSIDE
• PLAN: TO GIVE CHEMO CYCLES WITH PROPER FOLLOW UP
EXCISION OF AFFECTED RIB / RADIOTHERAPY AFTER CHEMOTHERAPY
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EXCISION OF AFFECTED RIB / RADIOTHERAPY AFTER CHEMOTHERAPY
POST 1ST VAC CYCLE X RAY CHEST

- The soft tissue mass appears to be resolving clinically and in the X-ray.
DISCUSSION ON ESFT

James Ewing (1866-1943) first described the tumour.

Malignant small, round, blue cell tumour.

2nd MC malignant bone tumour in children.

Incidence – approx. 3 cases / million / year

MC in teenagers and young adults

M:F – 1.6 : 1

MC sites – lower extremity (41%), pelvis (26%), chest wall (16%), upper extremity (9%), spine (6%), skull (2%)
Ewing's sarcoma, peripheral PNET of bone and soft tissue, pPNET of chest wall (ASKIN'S TUMOUR), ES with neural differentiation are grouped together as Ewing's sarcoma family of tumours.

Origin – hotly debated

- Neuroectodermal
- Mesenchymal cells of hematopoetic and non-hematopoetic origin

Human MSCs currently appear to be strong candidates as the cell of origin.
Clinical findings – localized pain and swelling
Additional – fever, weight loss, anemia, increased ESR, leucocytosis
Causes – 85% t(11;22) Others t(21;22), t(7;22)
Histology – Classical and Atypical/Variant
Diagnosis – Multimodality
**IMAGING FINDINGS**

**Moth eaten appearance**

**Periosteal reaction**

**saucerization**

**MRI** – assessment of soft tissue involvement / evaluates response to chemotherapy
TREATMENT

- Multidisciplinary
  - Chemotherapy – VAC/IE protocol
  - Radiotherapy
  - Surgery

PROGNOSIS

- 5 year survival rate
  - Localised disease – 60 to 70%
  - Metastatic disease – 20%
PROGNOSTIC MARKERS

1. Site of tumour
2. Tumour size
3. Age of patients
4. Sex
5. LDH
6. Metastasis
7. Ki-67 Expression
LITERATURE REVIEW

• Askin’s tumour: A case report by medical oncology dept, Hassan university hospital, Morocco

Source: World Journal of Surgical Oncology, January 2013

Presented two similar cases of Askin’s tumour.
CONCLUSIONS:
Rib tumors are rare entities in the pediatric population. However, a significant number of rib lesions are malignant. Therefore, proper diagnosis and expeditious treatment are critical.
Pediatric Ewing sarcoma of the rib: role of neoadjuvant chemotherapy in tumoral shrinking and sterilization. A case report.


Source—hôpital militaire d'instruction Mohamed V, Rabat, Morocco.

Abstract

The management and prognosis of these tumors have markedly improved with the use of multimodal therapy including adjuvant chemotherapy, surgery and/or irradiation.
• Prognostic factors in primary non metastatic Ewing sarcoma of the rib in children and young adults.

• Qureshi SS, Kembhavi S, Vora T, Ramadwar M, Laskar S, Talole S, Kurkure P.

• Source--

• Relapses occurred more often at distant sites and had a poor outcome. In this study, poor histologic response to chemotherapy \((P = .04)\) and the infiltration of adjacent lung parenchyma \((P = .01)\) are adverse prognostic factors.
Chest wall Ewing sarcoma family of tumors: long-term outcomes.


Source
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CONCLUSIONS:

This is the largest single-institution series describing the treatment of chest wall Ewing tumors. Despite improvements in survival, obtaining local control is challenging and often accompanied by morbidity. Effort should be focused on identifying tumors amenable to combined-modality local therapy and to improving RT techniques.
Rib tumors are rare entities in the pediatric population. However, a significant number of rib lesions are malignant. Therefore, proper diagnosis and expeditious treatment are critical.
THANK YOU !