A Diagnostic dilemma!

C. Rekha
ICH&HC
12 year old female child, admitted with h/o

- Breathlessness of 3 month duration
- Loss of weight and appetite
- 1 episode of hemoptysis
- Evaluated and diagnosed to have hemorrhagic pleural effusion for which she was treated with repeated aspirations, iv antibiotics and ATT
- Referred for persistent symptoms (repeated accumulation of effusion)
- No cough
- No fever
- No palpitation/ oliguria/ pedal edema
- No jaundice / hematemesis/ melena
- No vomiting / chronic diarrhea
- No skin rashes/bony pains/bleeding manifestations.
Past history...

- H/O trauma in the form of fall from tree one month before onset of symptoms.
- Contact h/o – father known pulmonary tuberculosis completed ATT 6 months earlier.
O/E

- Alert
- Wasted [WHO charts]
  - weight for age $<-2$ z score,
  - weight for height $<-2$ z score
- Afebrile
- Pallor +
- Tachypneic
- Dyspneic
- No lymphadenopathy
- No clubbing
- No cyanosis
• **Vitals**
  - Respiratory rate: 48/min, abdominal type of respiration
  - Pulse rate: 104/min
  - BP: 90/60
  - Temperature: N.

• **Respiratory system**
  - Confirmed features of right sided pleural fluid

• **Other systems with in normal limits**
Provisional diagnosis.

- Tuberculous effusion.
- Traumatic hemothorax
- Malignancy.
Investigations..

- CBC- N
- ESR- N
- Peripheral smear study- N
- CXR- massive right side effusion with mediastinal shift.
- USG chest- massive R sided turbid fluid
- Mantoux - neg
- Sputum for AFB- neg
- Sputum gene Xpert studies - neg.
- BT/CT/PT/aPTT - N
- LFT, RFT- N
- Serum for iron studies- N
- Pleural fluid analysis-
- Protein - 6.8 g/dl, glucose - 87,
  - Field full of RBC, 3-4 degenerated cells, AFB- Neg,
    gram stain- neg, C/S- NG, ADA- neg.
- Gene Xpert from pleural fluid - neg.
- Pleural fluid cytology- discrete aggregate of lymphocytes and neutrophils, histiocytes and few mesothelial cells.
- Pleural fluid LDH : serum LDH - 15
- Vascular malformations, sequestrations, traumatic rupture
  CT angiogram of chest.

- Bone marrow studies
  - R/O leukemia, lymphoma, myelofibrosis - neg.
  - Serum uric acid - N
Provisional diagnosis..

- Tuberculous effusion. NO EVIDENCE
- Traumatic hemothorax NO
- Malignancy. NO
Other Rare causes...

- Pancreatitis
- Rheumatoid arthritis
- Other rare connective tissue disorders
- Serum amylase - N
- Serum lipase - N
- RF - N
- ANA, Anti-dsDNA, pANCA - neg
Pleural biopsy done - showed features suggestive of extramedullary hematopoiesis.
DEPARTMENT OF HISTOPATHOLOGY

Patient Name: Ms. ABIRAMI E
UHID: AC01-0002070369
W/BNo/RefNo: GP
Received on: 29/06/2014 01:22:54 PM
Printed on: 05/06/2014 11:55:41 AM
Ref Doctor: GOVT CHILDREN'S, HOSPITAL EGMORE

HISTOPATHOLOGY TEST (SMALL)

Specimen:
1. Pleural biopsy
2. Lung biopsy

Macroscopic Description:
- Bottle marked pleura: Multiple irregular reddish brown tissue fragments ranging from 1.0 to 3.0 cms A1 - 4 bits A2 - 4 bits
- Bottle marked lung: Single reddish to pale white tissue measuring 0.8 x 0.6 x 0.2 cms B - 2 all
  (Dr SNS)

Microscopic Description:
- A1 & A2: Section shows fragments of fibrinous material with poorly preserved lymphocytes, plasma cells, eosinophils and neutrophils. Also there is a fragment of fibrous and loose areolar tissue with congested blood vessels and crushed lymphoid infiltrates. No granulomas are seen.
- B: Section shows lung and pleura with pleural aspect showing lymphocytes, plasma cells, eosinophils, few neutrophils, nucleated erythrocytes and megakaryocytes are also seen. Crushed lymphoid infiltrates are also seen. No granulomas are seen.

IMPRESSION:
- BIOPSY FROM LUNG AND PLEURA SHOWING SUBACUTE INFLAMMATION WITH EOSINOPHILS AND WITH FEATURES SUGGESTIVE OF EXTRAMEDULLARY HEMATOPOIESIS.

Comments:
- Please correlate with clinical findings.

* END OF REPORT *
Causes of hemorrhagic effusion...

- The causes of hemorrhagic pleural effusion are divided into the following eight groups:
  - Pleuro pulmonary infections
  - Pleuropulmonary malignancy
  - Connective tissue diseases,
  - Abdominal causes
  - Cardiovascular (aneurysm rupture, pulmonary infarction).
  - Bleeding disorder
  - Miscellaneous causes (superior vena caval syndrome, Kawasaki disease)
Rarest of the rare - pleural extramedullary hematopoiesis.
Extramedullary hematopoiesis

- Extramedullary hematopoiesis is a physiological compensatory phenomenon occurring because of insufficient bone marrow function that becomes unable to meet circulatory demands.

- EMH is seen in many hematological diseases, such as myelofibrosis, polycythemia vera, leukemia, lymphoma or after bone marrow irradiation.
Sites....

- Most common areas
  - paraspinal regions of the thorax,
  - liver, and
  - spleen,
- Rarer sites
  - adrenal gland,
  - bowel,
  - dura mater
  - breast
  - perirenal and pelvicaliceal
  - pleura
  - pericardium
Theory behind EMH...

Hematogenous spread of multipotential stem cells occurs with eventual infiltration of various tissues and organs.
Pleural extramedullary hematopoiesis...

- Pleural EMH is a rarest phenomenon and is the most rarest cause of pleural effusion.

- Even if effusion occurs it becomes very rarely symptomatic.

- In children pleural extramedullary hematopoiesis has not been noted so far.

- Most commonly diagnosed only on postmortem examination in adults.

- Most common association is with MMM (myelofibrosis and myeloid metaplasia)
Investigations...

- Contrast enhanced CT.
- Technitium labelled bone marrow scan/sulphur colloid scan (best non-invasive method for confirming diagnosis)
- Pleural fluid analysis
- Pleural biopsy (gold standard).
Treatment...

- External beam radiation - total radiation of 150 centigray (71% response rate).
Back to our case....
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<thead>
<tr>
<th><strong>Favouring</strong></th>
<th><strong>Against</strong></th>
<th><strong>Favouring</strong></th>
<th><strong>Against</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Pleural biopsy</td>
<td>No hepatosplenomegaly</td>
<td>Most common in our country</td>
<td>Investigations neg.</td>
</tr>
<tr>
<td>findings</td>
<td>No evidence of hemolysis</td>
<td>Wasted and malnourished child</td>
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<tr>
<td></td>
<td>Peripheral smear – N</td>
<td>Father a known case</td>
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<td>Bone marrow – N</td>
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Is this really EMH

Why not tuberculosis
## Mixed connective tissue disorders?

<table>
<thead>
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<tbody>
<tr>
<td>Age</td>
<td>No external markers</td>
</tr>
<tr>
<td>Sex</td>
<td>No other serosal involvement other than pleura</td>
</tr>
<tr>
<td>Serositis (pleural effusion)</td>
<td>ESR- N</td>
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<tr>
<td></td>
<td>No thrombocytosis</td>
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<td></td>
<td>RF, ANA, dsDNA, ANCA-N</td>
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Further course...

- Treated with intercostal drainage.

- Started on ATT with steroids after discussion with hemato-oncologist / pulmonologist.

- Child became symptomatically better after 2 weeks of therapy with persistence of mild tachypnea.

- 4-5 months lapsed and child had been expected to come for follow up.
Dr. G. Durai Arasan (lead consultant)
Department of pulmonology, ICH&HC
Department of hematology, ICH&HC
Dr. Gowri Shankar, and Dr. Revathy Raj
Thankyou.
• Contact no of lead consultant

• Dr. G.Durai Arasan - 9381014773

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