A rare tumour of the chest wall
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A 66-year-old male with a 6 months h/o masses beneath both his arms pits presented with symptoms of gradually worsening shortness of breath for a week. He had significant orthopnoea and slight pleuritic chest pain but did not report any other symptoms and denied any work-up being done for these masses.

On examination, he was noted to be in moderate respiratory distress and his chest wall showed two hard masses on either side adjacent to the axillae. He had dullness to percussion over his lung fields and breath sounds were absent on both sides posteriorly. X-rays of his chest and abdomen confirmed pleural effusion and also demonstrated the calcified masses over the lower part of the chest wall (Figure 1 and Figure 2).

Figure 1. Posterior anterior chest X-ray revealed bilateral effusions and calcified masses over the chest wall
Figure 2. Abdominal radiograph demonstrated the calcified masses on either sides of the lower chest wall

A CT scan of his chest showed bilateral masses arising from the ribs measuring 10 × 15 cm in the coronal plane and 5–6 cm in the antero-posterior dimensions and extending into the mediastinum (Figure 3). The tumor on the left side even extended intra-abdominally, displacing the spleen.
Figure 3. Thoracic computerised tomography demonstrated 2 masses arising from the ribs bilaterally

There was no evidence on bone scan imaging to suggest continuity between these 2 masses and they were distinctive to each other in origin.

Thoracentesis was done and haemorrhagic fluid was aspirated which was otherwise unremarkable on analysis.

Are these tumors related and what is the diagnosis?—A core-biopsy from the mass on the right side revealed atypical spindle cells that were strongly positive for CD31, CD34, Ulex and Factor VIII (the latter only focally) on immunohistochemistry. The tumor was noted to surround the trabeculae of mature bone and because of this fact and it's location in the chest wall, it was suggested to be a primary Epithelioid Hemangioendothelioma (EH) of the bone with soft tissue extension. Later, a similar biopsy obtained from the mass on the left side was also diagnostic of EH. Patient declined any surgical or chemotherapeutic options for his management. However, he was repeatedly admitted with recurring hemothoraces that required therapeutic thoracentesis and blood transfusions until his death 2 months later.

Discussion—This is a case of multicentric EH of the bone originating synchronously from the ribs on either sides of the chest wall and invading into the mediastinum as well as causing hemothoraces. EH is a rare low-grade tumor of vascular origin that can arise from any site in the body and has a tendency to be multifocal. It is unclear whether the separate lesions represent metastases or multicentric disease. Diagnosis requires immunohistochemistry and though it is a low-grade tumor, prognosis can be poor if left untreated.

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