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Ewing-like Sarcoma – Hiding in PA view

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Learning Objectives

- Identify the clinical and radiological findings of Ewing-like sarcoma
- Review the importance of obtaining a thorough history and physical exam, and recognize their value in the process of medical decision making
- Recognize that non-resolving pneumonia may be the first manifestation of malignancy involving the lung

Case Description

- A 14-year-old boy presented to the emergency department after 3 days of fever, pleurisy, and worsening left mid-back pain. Chest x-ray (CXR) showed left lower lobe (LLL) airspace opacity consistent with pneumonia without evidence of effusion. He was admitted for IV antibiotics and discharged 6 hours later with oral antibiotics.
- Two weeks later he presented to his primary care physician with continued pain. Repeat CXR showed LLL pneumonia with effusion. **Figure 1**. Patient was then directly admitted after failed outpatient treatment for pneumonia with effusion.
- Further history and physical exam unveiled an unintentional 20 lb weight loss over the course of 6 months, as well as a mass protruding between his posterior ribs.
- Lack of response to antibiotics prompted testing with computer tomography (CT) of the chest, which showed a large heterogeneous mass at the posterior left upper quadrant (LUQ) of the abdomen protruding through the posterior aspect of the left hemidiaphragm causing atelectasis. **Figure 2**.

Figure 1

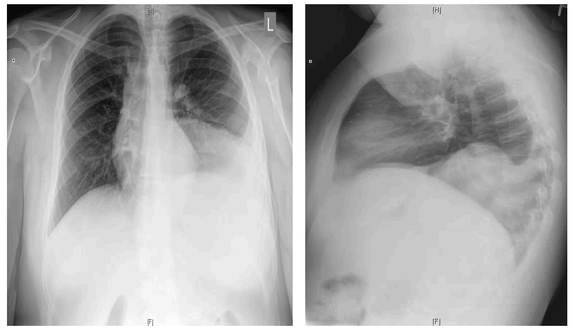

Figure 1: PA/Lateral CXR. LLL pneumonia with left pleural effusion.

- Magnetic resonance imaging (MRI) demonstrated a 17.9 x 10.6 cm heterogeneous hyperenhancing hypervascular posterior LUQ mass which invades the posterior wall of the thorax and abuts if not invades the left hemidiaphragm. **Figure 3**.
- A biopsy was performed, which revealed this mass to be an Ewing-like sarcoma with a BCOR-CCNB3 rearrangement.
- Patient was treated with chemotherapy prior to surgical resection and radiation.

Figure 2

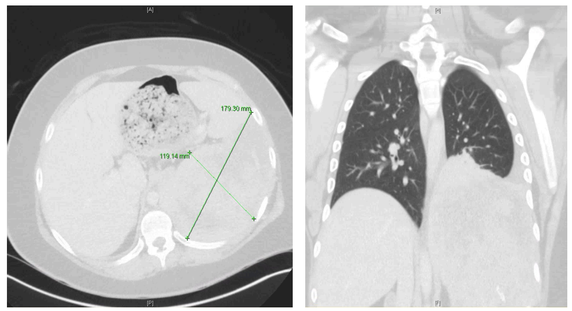

Figure 2: CT chest with contrast. Large LUQ mass causing atelectasis.

Figure 3

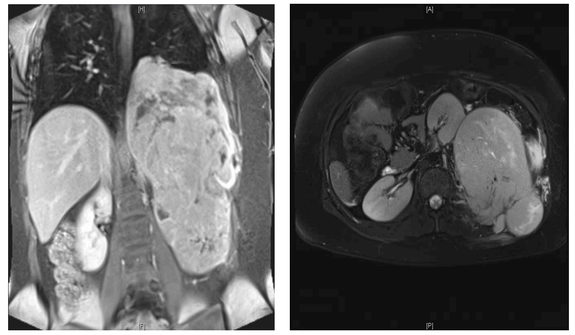

Figure 3: MRI of the chest. Mass invades the posterior aspect of the chest wall and displaces the left kidney and spleen anteriorly.

Discussion

- Broad differential diagnoses apply to non-resolving pneumonia. These include infectious, inflammatory or neoplastic processes.
- Neoplasms may be associated with non-resolving pneumonia either by compromise of the airway lumen and secondary postobstructive pneumonia or by mimicking an infiltrative process.
- Ewing-like sarcomas (ELS) are a rare heterogenous group of round cell sarcomas that typically occur in the bone and soft tissue of pediatric and young adult patients.
- ELS share various degrees of morphological, molecular, and clinical similarity with Ewing sarcomas. However, these tumors lack the pathognomonic molecular hallmark of Ewing sarcoma, which is defined as translocation between a gene of the RNA-binding TET family with a gene of the ETS-transcription family.
- Accurate classification and distinction from classical Ewing sarcomas is important for patient management.
- The diagnosis is suggested based on a combination of morphologic and immunophenotypic features, but requires molecular testing for confirmation.
- A subset of ELS harboring the BCOR-CCNB3 rearrangement have been described recently.
- BCOR-rearranged sarcomas most commonly arise in bones of patients in their second decade (with a median age of 15 years).
- There is a striking male predominance, with nearly 85% of BCOR-rearranged sarcoma cases occurring in males.
- The 5-year overall survival for BCOR-rearranged sarcomas is approximately 75%.
- BCOR-rearranged sarcomas located in the axial skeleton and soft tissues show a significantly shorter survival.

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