Ewing-like Sarcoma – Hiding in PA view

Andrew Donahue
Abigail Cruz

Department of Pediatric Hematology/Oncology, College of Medicine, East Tennessee State University

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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 revealed a 20 lb weight loss over the course of 6 months, as well as a mass protruding between his posterior ribs. Figure 2. A biopsy was performed, which revealed this mass to be an Ewing-like sarcoma with BCOR-CCNB3 rearrangement. Patient was treated with chemotherapy prior to surgical resection and radiation.

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 after failed outpatient treatment for pneumonia with effusion.

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.

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

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

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

References


