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A Rare Cause of Giant Intrathoracic Mass in a Woman with Sickle Cell Disease: Extramedullary Hematopoiesis

Ufuk F. et al.: Extramedullary Hematopoiesis

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A 41-year-old woman presented with worsening dyspnea and chest discomfort for a few months. She had a history of transfusion-dependent sickle cell disease since the age of 4, cholecystectomy, and splenectomy. Physical examination revealed conjunctival pallor, pretibial edema, and decreased breath sounds in both lungs. Laboratory analysis showed decreased hemoglobin (6.3 g/dL) and increased serum creatinine level (2.9 mg/dL). Clinical and laboratory assessment revealed prerenal kidney failure. A right jugular vein catheter was placed, and hemodialysis was planned. Chest radiography showed giant intrathoracic masses, and a chest CT was performed for further evaluation. CT revealed well-defined, fat-containing, giant masses in the mediastinum, suspicious for extramedullary hematopoiesis (EMH) (Figs. 1, 2). A fine needle aspiration biopsy confirmed the diagnosis and multiple blood transfusions started.

EMH is the formation where blood cell precursors are produced outside the bone marrow (1). Sickle cell disease is a relatively unusual cause of EMH. Radiologically, thoracic EMH is characterized by mass/masses in the posterior mediastinum with smooth margins and visible fat, as in the present case (2). Although treatment of the underlying disease is often sufficient for EMH, mass effects due to EMH may rarely occur, and radiotherapy, surgery, or medical treatment may be required (1, 2).

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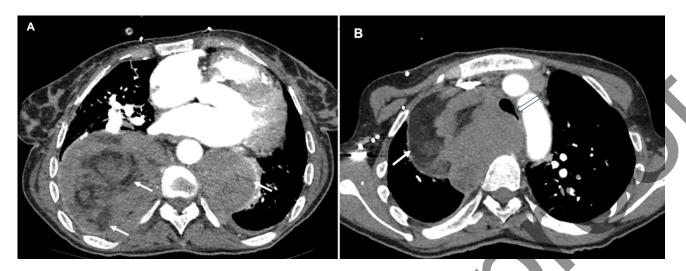


Figure 1: A, B) Axial contrast-enhanced chest CT images show well-defined giant masses in the mediastinum with areas of macroscopic fat densities (arrows). Note the tracheal compression and narrowing due to right paravertebral mass (open arrow).



Figure 2: A three-dimensional volume rendering image of the chest demonstrates huge paravertebral masses (arrows). Note the H-shaped vertebral bodies (central endplate depression) due to sickle cell disease.