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

Orak Hücre Hastalığı Olan Bir Kadında Dev İntratorasik Kitlenin Nadir Bir Nedeni: Ekstramedüller Hematopoiezis

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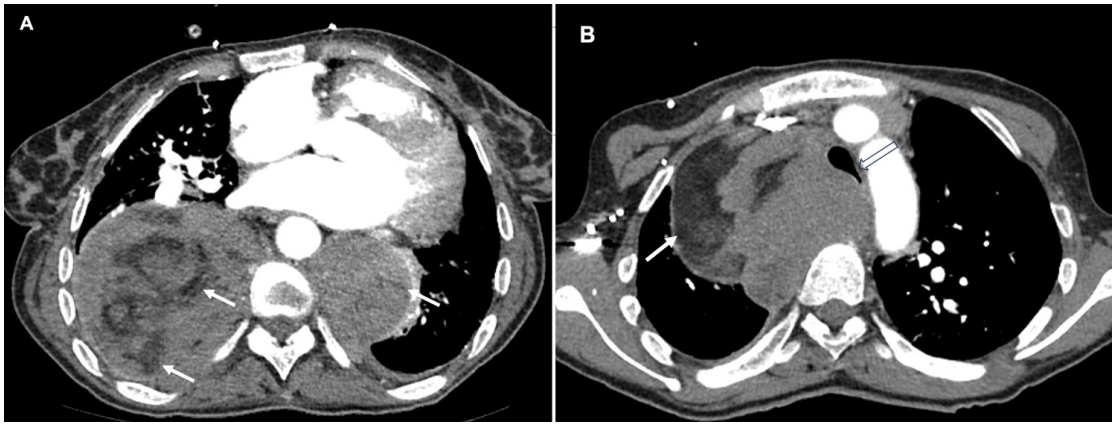

Figure 1. (A, B) Axial contrast-enhanced chest computed tomography images show well-defined giant masses in the mediastinum with areas of macroscopic fat densities (filled arrows). Note the tracheal compression and narrowing due to the right paravertebral mass (hollow arrow).

Figure 2. 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.

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A 41-year-old woman presented with worsening dyspnea and chest discomfort persisting 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 diminished 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 assessments revealed prerenal kidney failure. A right jugular vein catheter was placed and hemodialysis was planned. Chest radiography showed giant intrathoracic masses and chest computed tomography (CT) was performed for further evaluation. CT revealed well-defined, fat-containing, giant masses in the mediastinum, suspicious for extramedullary hematopoiesis (EMH) (Figures 1 and 2). A fine-needle aspiration biopsy confirmed the diagnosis and multiple blood transfusions were begun.

In cases of EMH, 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 a mass or masses in the posterior mediastinum with smooth margins and visible fat [2], as in the present case. Although treatment of the underlying disease is often sufficient for EMH, effects of masses due to EMH occur rarely, and radiotherapy, surgery, or medical treatment may be required in such cases [1,2].

Keywords: Sickle cell disease, Computed tomography, Extramedullary hematopoiesis, Chest

Anahtar Sözcükler: Orak hücre anemisi, Bilgisayarlı tomografi, Ekstramedüller hematopoiezis, Göğüs

Ethics

Informed Consent: Obtained from the patient.

Authorship Contributions

Concept: İ.O.; Design: F.U.; Analysis or Interpretation: L.C., L.L.; Literature Search: F.U., L.C.; Writing: F.U., İ.O., L.L.

Conflict of Interest: No conflict of interest was declared by the authors.

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