INTRATHORACIC EXTRAMEDULLARY HEMATOPOIESIS: A SCAN DIAGNOSIS

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Confirmation of the diagnosis of intrathoracic extramedullary hematopoiesis by scintillation scanning can avoid potentially difficult surgery. This unusual complication of chronic anemia has occasionally been reported in recent literature and colloidal ^{198}Au has been used to visualize these masses (*1–4*). Few cases, however, have been diagnosed prior to surgery or autopsy (*1,4*). We recently had the opportunity to evaluate posterior mediastinal masses in a young male with severe thalassemia intermedia by radioactive-tracer techniques.

CASE REPORT

C.E.H. #683691. This 25-year-old Negro salesman was known to be anemic since childhood, requiring three separate transfusions after intractable nosebleeds. For 2–3 months before admission he noticed increasing fatigue and dyspnea on exertion. These symptoms were not relieved by oral iron medication. There was no family history of anemia. On examination he was found to have scleral icterus, a systolic heart murmur and hepatosplenomegaly. Laboratory findings included hemoglobin concentration of 6.8 gm%, hematocrit of 24%, reticulocyte count of 4.6% and white blood count of 7895. White-blood-cell differential count was normal, and there were 26 nucleated RBC's/100 WBC. The red cells were markedly hypochromic and many target cells were present. The red cells varied greatly in size and shape. The bone marrow was hypercellular and most of the cells were erythroid precursors. A sickle-cell preparation was negative. The red cells showed increased resistance to osmotic lysis. Hemoglobin electrophoresis demonstrated no abnormal hemoglobins. Hemoglobin A_2 was found by column chromatography on DEAE-Sephadex A50 to be 4.2% of the hemoglobin present (normal 2.1–2.9%). Hemoglobin F was found by alkali denaturation to be 6.4% of the total (normal less than 2.5%). The serum uric acid level was 13 mg% (normal 3–6 mg%). Both parents and three of four siblings had abnormalities of red cell morphology, osmotic fragility, hemoglobin A concentration and/or hemoglobin F concentration consistent with thalassemia minor.

Roentgenographic studies revealed coarsened trabecular markings with medullary packing throughout most of the bones examined (Fig. 1). A multilobular posterior mediastinal mass was noted on chest roentgenograms (Figs. 2 A, B). Thick linear planigraphic sections showed the masses to be extrapleural and separate from the adjacent vertebral bodies and ribs (Figs. 3 A, B). A ^{51}Cr RBC survival test and liver-spleen sequestration study recorded a normal 7-day curve. Twenty-five μc of ^{59}Fe (ferrous citrate) was injected intravenously, and the $T_{1/2}$ time of plasma iron clearance was 30 min (normal $T_{1/2}$ = 60–120 min). Organ counts with a 2-in. thyroid-type scintillation probe showed rapid disappearance of the ^{59}Fe tracer from the precordium and accumulation by the sacrum, spleen and thoracic masses.

A posterior thorax scan was performed 24 hr after intravenous injection of 25 μc of ^{59}Fe. A commercial rectilinear scanner with a 5-in. scintillation crystal and a coarse 5-in.-focus collimator was used. The maximum counting rate was low (about 5,000 cpm) and much of this counting rate was due to collimator leakage. The standard collimation designed for energies up to 0.5 Mev was inadequate for the 1.1- and 1.29-Mev photons from ^{59}Fe. In spite of these limitations a patchy but diagnostic scan was obtained (Fig. 4 A).

The image was hardened by photocopying in the Blu-Ray Radiograph Duplicating Printer (*5*) (Fig. 4 B), achieving a cutoff of low image densities. This

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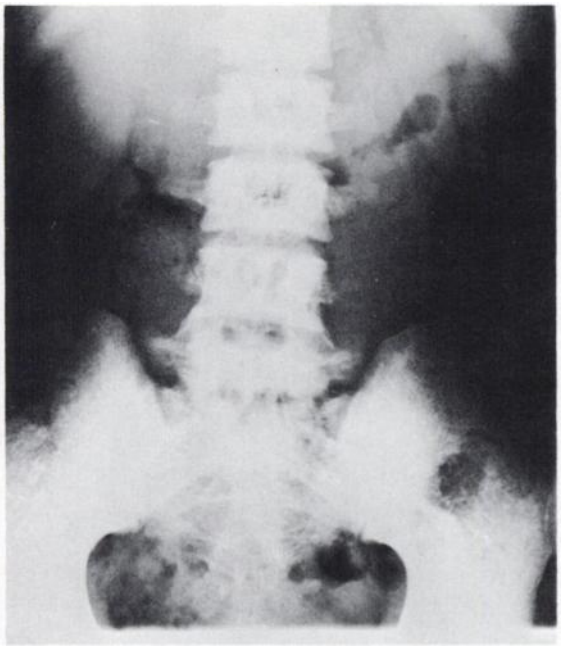

FIG. 1. Supine abdomen roentgenogram. Note prominent spleen and liver enlargement and coarsening of trabecular markings throughout the spine and pelvis.

copy was then superimposed on the PA chest roentgenogram to show a correspondence between the regions of increased uptake and the posterior mediastinal masses (Fig. 4 C).

A diagnosis of thalassemia intermedia with intrathoracic extramedullary hematopoiesis was made without biopsy. An 800-rad surface dose of external radiation was given to the posterior mediastinum to reduce the size of the mass and thereby reduce the possibility of cord compression. One return visit revealed no significant change in the size of the masses.

DISCUSSION

Extramedullary hematopoiesis is a compensatory phenomenon associated with long-standing severe hemolytic anemia. It has been found in the spleen, lymph nodes, liver, adrenal glands, cartilage, broad ligaments and even within thrombi and adipose tissue (*6*). Intrathoracic extramedullary hematopoiesis gives a rather typical roentgenographic appearance of multiple, lobular paravertebral masses usually between T6 and T12 vertebral segments. These are well circumscribed without bony erosion or spiculation (*1,4*). It is unknown whether these masses represent embryonal rests of osteogenic tissue or marrow emboli (*1,3*). Planigraphic sections clearly separated the masses from adjacent vertebral bodies and ribs (Figs. 2 A, B). Therefore the thoracic masses are not direct extension of hematopoietic tissue from the marrow.

Spinal-cord compression did not occur in this patient but has been reported to occur by extension

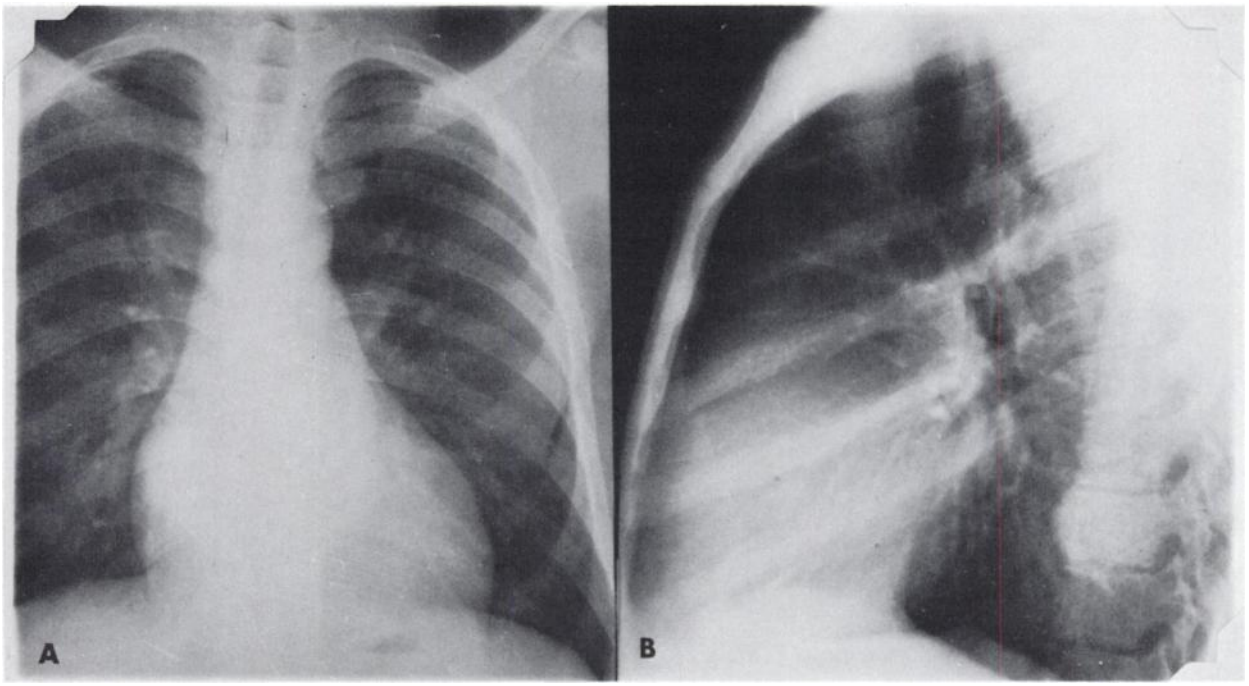

FIG. 2. PA (A) and lateral (B) chest roentgenograms record multilobular mediastinal masses visible through the heart shadow and overlying the spine on lateral view.

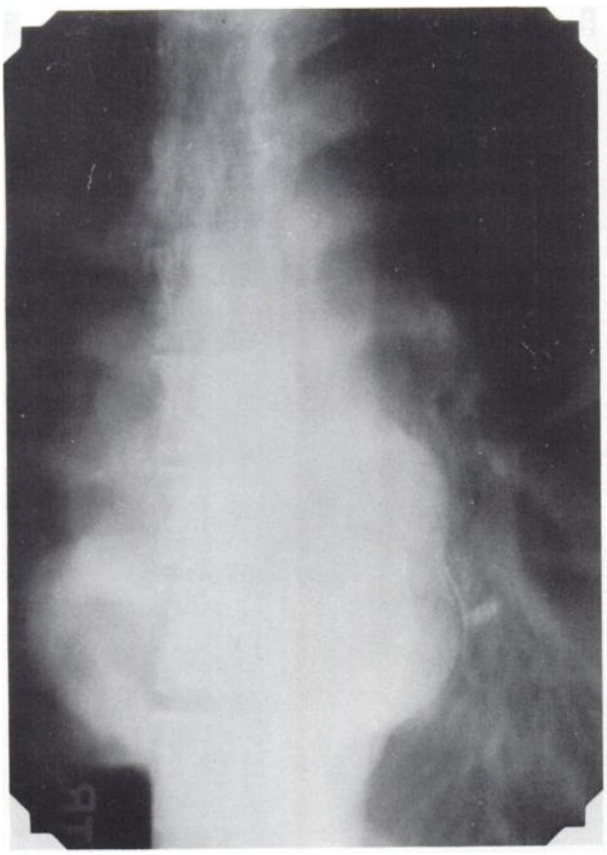

FIG. 3. Planigraphic sections through mid-point of masses, revealing intact vertebral body segments and extrapleural nature of masses.

of the hematopoietic mass into the spinal canal (*1,3,4*). Laminectomy is difficult when this occurs because of the highly vascular nature of the masses. This complication of intrathoracic extramedullary hematopoiesis can be avoided by early radiation therapy (*3*).

The definitive diagnosis of any new growth within the thorax is essential. Percutaneous biopsy in cases of extramedullary hematopoiesis is hazardous because of the danger of hemothorax. It is therefore advantageous to make a diagnosis without surgery. Scanning with ^{59}Fe, usually requiring large doses, can be combined with a plasma ^{59}Fe-clearance study. The quality of the scan is less than optimal but the scan may be helpful in establishing the location and presence of extramedullary red-cell production (*8*). The scan is best performed after organ counting reveals disappearance of the tracer from the heart pool with concentration in areas of active hematopoiesis. This is usually 24 hr after injection in patients with hemolytic anemia and active erythropoiesis.

SUMMARY

A diagnosis of intrathoracic extramedullary hematopoiesis was confirmed in a young Negro man with thalassemia intermedia by scanning following a plasma ^{59}Fe-clearance study. Surgical biopsy was not thought necessary because of the clear evidence of ^{59}Fe incorporation into the masses. Increased tracer concentration was further clarified by using an elevated intensity factor in a film-copying process.

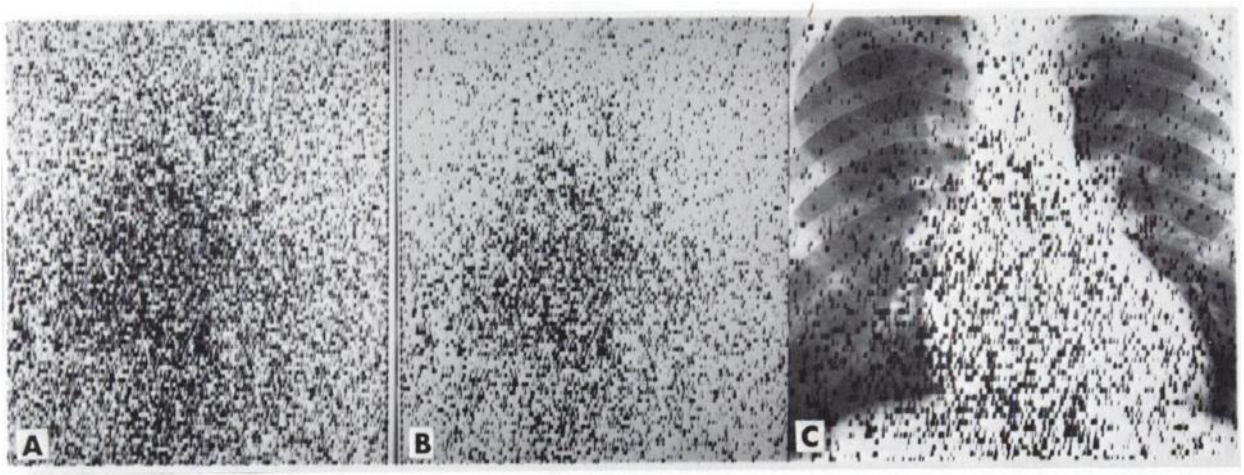

FIG. 4. A: Initial posterior thoracic photoscan 24 hr following intravenous administration of 25 μc of ^{59}Fe. B: Copy of initial photoscan at full-scale intensity setting of photocopier. C: Photoscan copy oriented over PA chest roentgenogram showing correlation of ^{59}Fe concentration with masses. Scan superimposed on the PA chest roentgenogram in C is same as B. Composite is enlarged by factor of 1.6 compared with A and B.

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