Pallor, Abnormal Bones and Paravertebral Masses

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This 25-year-old man has had pallor, hepatosplenomegaly and stunted growth since childhood. There has been no bleeding. There is frontal bossing of the skull and the maxillae are prominent. The heart is enlarged and a systolic murmur is heard.

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**Diagnosis: Thalassemia Major**

The chest film reveals widening of the ribs with hazy, coarse architecture and dense, lobulated, bilateral paravertebral masses (Fig 1). Skull films showed increased diameter of the diploe with thinning of both tables. Other bones, including the vertebrae, showed increased diameters, decreased density and coarse disorganized cancellous structure (Fig 2).

The blood showed Hgb 3.5 gm percent, red blood cells 800,000/mm³, metamyelocytes 1 percent, reticulocyte count 2.2 percent, platelets 140,000/mm³. The peripheral smear revealed marked hypochromia and erythrocytosis, polychromia and occasional target cells. No inclusion bodies were found. The serum bilirubin was 2.2 mg percent. The urine showed increase in urobilinogen content.

Hemoglobin electrophoresis revealed 41 percent fetal hemoglobin. A sister had a fetal hemoglobin content of 44 percent. Thus the hematologic diagnosis of thalassemia major was confirmed.

The paravertebral masses are of particular interest in this patient. While neurogenic tumor is the most common cause of a posterior mediastinal mass, the multiple lesions seen here excluded that diagnosis. Knoblich described a similar patient in whom thoracotomy, performed for a possible neurogenic tumor, revealed osseous masses. Subsequent hematologic studies suggested the diagnosis of thalassemia.

Hemopoietic tissue in the thoracic cavity was first reported at autopsy in 1927 by Brannan, who found a mass in a patient suffering from hemolytic anemia. Dawson reported a patient with hemolytic anemia in whom four masses of bone, 4 cm each, were found in the left paravertebral region. Others have reported large intrathoracic masses that contained red marrow and blood in thalassemia. Sometimes the masses are bony enlargements; sometimes they are composed of soft tissue. These multiple, posterior mediastinal masses represent extramedullary hemopoiesis in the costovertebral areas.

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**REFERENCES**


**CULTURE AND BEHAVIOR**

Journalists and fiction writers speak of “unwritten law” by which they must mean the culture of a tribe. Culture comes ages before formal law. Nature people like the Eskimo have no formal law at all; there are no courts or statutes or jails, but the living law or culture may enjoin the death penalty just the same. Unless the formal law is in line with the living law, it cannot be enforced. The Prohibition law in the United States banning alcoholic beverages was an instructive example. One difficulty in setting up a formal international law today is that there is as yet no international living law, no planetary culture.


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