Answer

Extramedullary Hemopoiesis

Extramedullary hemopoiesis is the result of a compensatory mechanism against many chronic hematological diseases that are characterized by bone marrow depression. These conditions include thalassemia major or intermedia, sickle cell disease, polycythemia vera, chronic myelogenous leukemia, and hereditary spherocytosis. The most common sites of occurrence are the liver, spleen, and lymph nodes,[1] but many tissues have the potential to exhibit extramedullary hemopoiesis, such as the kidney, adrenal glands, breasts, spinal cord, pleura, endometrium, intracranial cavity, and intrathoracic cavity. Intrathoracic extramedullary hemopoiesis is rarely seen and is usually asymptomatic, requiring no therapy.[2] However, it may sometimes present with spinal cord compression, pleural effusion, massive hemothorax, or dyspnea.[3] It is important to distinguish a mass caused by extramedullary hemopoiesis from other lesions involving the posterior mediastinum. Diagnosis is based on clinical and radiological findings, with magnetic resonance imaging as the diagnostic modality of choice.[4] The differential diagnosis includes neurogenic tumors, lymphomas, primary and secondary malignant lesions, extrapleural cysts, and paravertebral abscesses. Biopsy may sometimes be necessary to confirm the diagnosis,[2] and surgery and radiotherapy may be needed to treat symptomatic patients with spinal cord compression.

Although extramedullary hemopoiesis is a rare cause of an intrathoracic mass, the diagnosis must be considered in any patient with congenital hemolytic disorder. In this regard, a potentially invasive procedure such as surgery should be avoided.

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