Case 2465

Extramedullary haematopoiesis
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Section: Chest imaging
Case Type: Clinical Cases
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Patient: 51 years, female

Clinical History:
A 51-year-old female presented with hereditary spherocytosis and she had had multiple blood transfusions. She was found to be increasingly short of breath.

Imaging Findings:
Patient was known with hereditary spherocytosis and had received multiple blood transfusions over many years. She now presented with anemia and shortness of breath. Chest radiography was performed (Figure 1A and 1B), which showed large well-defined masses in the posterior mediastinum and paravertebral regions. Subsequently, unenhanced CT of the thorax and upper abdomen was performed, which confirmed the presence of soft-tissue masses in the paraspinal region with mixed high and low attenuation (Figure 2). However, the upper abdomen also showed high attenuation of the spleen and liver, mild hepatosplenomegaly, enlargement of the right adrenal gland and low attenuation of the aorta (Figure 3).

Discussion:
A differential diagnosis of lymphoma was contemplated, but the combination of the findings favoured the diagnosis of extramedullary haemopoiesis in response to underlying chronic anaemia (notice the low attenuation of the abdominal aorta) and haemochromatosis of the liver due to multiple blood transfusions. The commonest presentation of extramedullary haemopoiesis includes soft tissue paraspinal mass, rib expansion and hepatosplenomegaly. Some may present with spinal cord compression. These findings are not specific for extramedullary haemopoiesis and there is a wide radiological differential for each of these findings. More rare involvement has been described in kidneys, adrenal glands, the middle ear, skin and uterus. Finally, any site of lymph node chains can produce soft tissue masses. Low attenuation of aorta on a non-enhanced CT scan can be seen in severe chronic anaemias due to any cause. Thus, it has a wide differential of several primary haematological disorders causing anaemia but also may acutely be seen in severe blood loss following trauma or exsanguination following trauma. Furthermore the use of artificial blood, tissue expanders and major fluid overload (for instance, rapid infusion of a large amount of clear fluids) can cause decrease in the attenuation values in the aorta. A special mention must be made about repeated blood transfusions in patients with congenital haemolytic anaemias, such as hereditary spherocytosis as in this case. This leads to excessive iron loading and haemochromatosis, which increases the attenuation in liver. MRI is the investigation of choice for evaluation of posterior mediastinal and paraspinal masses, and is well suited to demonstrate the extent of disease. In cases of cord compression, use of steroid with external beam radiation has a favourable response, and MRI can assist in planning of radiation field and assess therapeutic response. The hallmark of MRI findings is a soft-tissue mass in any of the locations described above, which tends to be low to iso-intense on T1 and heterogenous to hyper-intense on T2, with only minimal Gadolinium enhancement. Bone marrow replacement is also dominant, as fatty bone marrow is replaced by red bone marrow (in later stages, this may reverse and indeed bone marrow fibrosis/sclerosis may develop). The liver, however, tends to show signal void in cases of excessive iron loading. Indeed, quantification of iron load using MR methods is feasible. Thus, it can be concluded that there is no single imaging
finding, which uniquely suggests extramedullary haemopoiesis, but a constellation of findings in the right clinical setting is very suggestive. Familiarity with cross-sectional imaging manifestations together with history and clinical features of the patient is vital in diagnosing extramedullary haemopoiesis. Some of the rare manifestations may need biopsy for definitive diagnosis. The importance of differentiating extramedullary haemopoiesis from a neoplastic process is crucial because of the favourable response to steroids and external beam radiation of the former and cross sectional imaging with CT (and MRI) is crucial in this regard.

**Differential Diagnosis List:** Extramedullary haematopoiesis with haemochromatosis.

**Final Diagnosis:** Extramedullary haematopoiesis with haemochromatosis.

**References:**

Georgiades CS, Neyman EG, Francis IR, Sneider MB, Fishman EK. Typical and atypical presentations of extramedullary hemopoiesis. AJR 2002 May ;179(5):1239-1243. (PMID: [12388506](pmid:12388506))


Description: A plain chest radiograph (PA view) at the time of admission demonstrating double contours with widening of the paraspinal lines indicating masses. The heart configuration appears enlarged, but the heart itself is actually within normal limits (in this case). Origin:
Description: A lateral chest radiograph demonstrating the paravertebral extent of the soft tissue masses. Origin:
Description: An axial CT image of the lower thorax demonstrating extensive soft tissue masses in a paravertebral position. Small pleural effusions are also present. Origin:
Description: An axial CT image of the upper abdomen demonstrating a mass in the right adrenal. The high attenuation of the liver and the spleen is striking, as is the low attenuation of the abdominal aorta.

Origin: