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An Uncommon Cause of Abdominal Pain in a Patient with Thalassemia Intermedia

To the Editor:

Extramedullary hematopoiesis represents a compensatory mechanism to overcome ineffective erythropoiesis in patients with thalassemia as well as other hematological diseases.[1-7] Extramedullary hematopoiesis is more common in patients with thalassemia intermedia than in those with thalassemia major, and is frequently found within the spleen, liver, and occasionally, the lymph nodes.[2,3,5-8] Extramedullary hematopoiesis is typically asymptomatic and largely goes undiagnosed.[1,2,4-7] In particular, presacral extramedullary hematopoiesis is an extremely rare condition, with limited published case reports found in the literature.[1,2,5,8]

CASE SUMMARY

A 46-year-old woman presented to the Emergency Department with abdominal pain and exertional dyspnea. She had a history of beta-thalassemia intermedia and iron deficiency anemia secondary to menorrhagia from uterine fibroids. On physical examination, she appeared pale, with no evidence of petechia or lymphadenopathy. Abdominal palpation revealed a 20-week-sized uterus and splenomegaly. Laboratory studies showed hemoglobin of 5.3 mg/dL and hematocrit of 17%, with a mean corpuscular volume of 56 fL. Urine pregnancy test was negative and abdominal computed tomography scan was interpreted as presacral and retrocrural masses consistent with lymphadenopathy, with the largest mass measuring approximately 4 cm (**Figure**). A presumptive diagnosis of lymphoproliferative disorder was made and the patient underwent image-guided biopsy of one of the masses. Histopathology revealed cellular elements showing trilineage hematopoiesis along with marked erythroid hyperplasia, without any evidence of malignancy. These findings were consistent with extramedullary hematopoiesis. She was treated with red-cell transfusions, iron supplementation, and hormonal agents for menorrhagia.

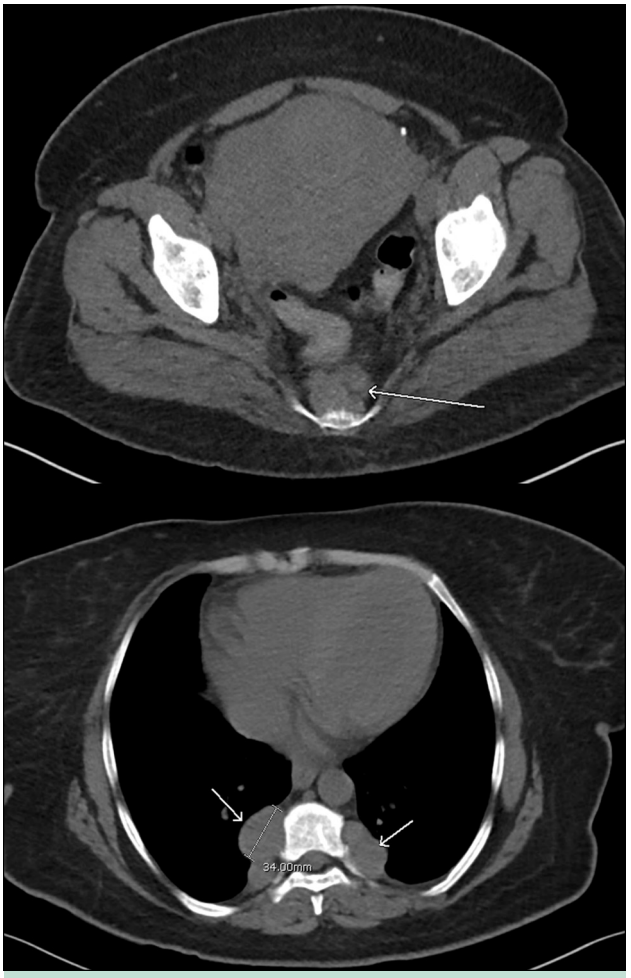

Figure **Top:** Computed tomography scan of the pelvis demonstrating a presacral mass (white arrow). **Bottom:** Computed tomography scan of the chest demonstrating retrocrural masses (white arrows).

DISCUSSION

In patients with thalassemias, frequent red-cell transfusion often ameliorates the physiologic stimulus for extramedullary hematopoiesis. In this case, iron deficiency anemia further augmented the drive for extramedullary hematopoiesis. Clinically, extramedullary hematopoiesis

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may masquerade as tumor formation in different parts of the body. Nearly 80% of extramedullary hematopoiesis cases are asymptomatic and incidentally diagnosed on imaging, unlike this patient, who presented with abdominal pain.[1,2,4-7] One of the most dreaded complications of extramedullary hematopoiesis is spinal cord compression secondary to a paraspinal mass.[2,3,7] Gadolinium-magnetic resonance imaging is the imaging modality of choice for diagnosing extramedullary hematopoiesis, and may help to avoid more invasive diagnostic tests.[1,4,6,7] Early diagnosis is essential to prevent catastrophic complications such as neurologic sequelae. The primary goal of management is to treat the underlying anemia and any associated conditions. Based on severity of presentation, additional management options may include radiotherapy, surgical decompression, hydroxyurea, or a combination of these modalities.[3,4,6,7]

CONCLUSION

The presence of iron deficiency anemia increases the risk of extramedullary hematopoiesis in patients with thalassemia. Extramedullary hematopoiesis should be considered as a differential diagnosis in any patient with thalassemia presenting with masses resembling lymphadenopathy or tumors, and appropriate treatment should be initiated in order to avoid potentially harmful complications.

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