



How do I diagnose *Extramedullary Hematopoiesis*

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Differential diagnoses: Extramedullary myeloid proliferations, Myeloid sarcoma.

Abstract

Extramedullary hematopoiesis occurs in bone marrow disorders and is most commonly seen in thalassemia and myelofibrosis. We present a case of a 58 year-old male, with a history of thalassemia, which presented with a para-vertebral mass composed of hematopoietic tissue. When considering the differential diagnosis, a primary concern was an extramedullary proliferation of neoplastic hematopoietic elements, like extramedullary myeloid sarcoma. Neoplastic extramedullary proliferations can consist of trilineage marrow elements and closely mimic the appearance of benign extramedullary hematopoiesis. The morphologic appearance may be a diagnostic clue, but cannot be completely relied upon.

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Anamnesis / History

A 58 years-old male, smoker, with a personal history of thalassemia minor, elevated blood pressure and cardiac insufficiency, presented with left thoracic pain and perspiration for the last four months. The blood count revealed microcytic hypochromic anemia: hemoglobin (Hb), 110 g/l; mean corpuscular volume, 65.5 fl; mean corpuscular Hb concentration, 299 g/l. TC and MRI imaging revealed a left para-vertebral mass, not compromising the vertebral space and suggestive of a neurogenic tumor.

Gross - microscopic findings

We received a CT-guided transthoracic needle aspiration biopsy of the mass: two filiform fragments of 15mm each.

Microscopy

Microscopy revealed hematopoietic tissue with trilineage proliferation of bone marrow elements (positive immunostaining for CD61, MPO, Glycophorin and CD68), with no evidence of immature myeloid cells (negative immunostaining for CD34, CD117 and TdT) .

Expression of markers

CD61(pos), Glycophorin (pos), CD68 (pos), MPO(pos), CD34(neg), CD117(neg), TdT (neg).



Discussion

Extramedullary hematopoiesis occurs in bone marrow disorders and is most commonly seen in thalassemia and myelofibrosis. Common locations include the spleen, liver, lymph nodes and para-vertebral regions. There are reports of extramedullary hematopoiesis presenting as a para-vertebral mass in middle aged men with a history of thalassemia. A common complaint is chest discomfort. Fever and sweating have also been reported.

When considering the differential diagnosis, a primary concern is an extramedullary proliferation of neoplastic hematopoietic elements. Extramedullary myeloid sarcoma is a rare extra medullary tumor mass consisting of myeloid blasts. It occurs in about 5% of patients with acute myeloid leukemia or rarely presenting as an isolated mass. The tumor consists of populations of blasts, but mixtures of cell types are seen in the more differentiated subtypes that can consist of trilineage marrow elements and closely mimic the appearance of benign extramedullary hematopoiesis.

Both neoplastic and benign proliferations may present as unilineage or multilineage. A neoplastic proliferation typically presents as a monomorphous population of intermediate size, noncohesive hematopoietic cells, with immature eosinophils as an important morphologic clue. Clusters of immature myeloid cells (eg blasts) may be seen. Multilineage benign proliferations often display all developmental stages of hematopoietic cells, but predominantly mature forms.

Thus morphologic appearance may be a diagnostic clue, but cannot be completely relied upon. Special studies are necessary to rule out neoplastic proliferations, the most useful being CD34, CD117 and TdT immunostains. CD34 expression is associated with an immature myeloid phenotype; CD117 staining suggests a myeloid origin and the presence of immature myeloid cells; TdT staining strongly supports acute lymphoblastic leukemia/lymphoma or myeloid sarcoma.

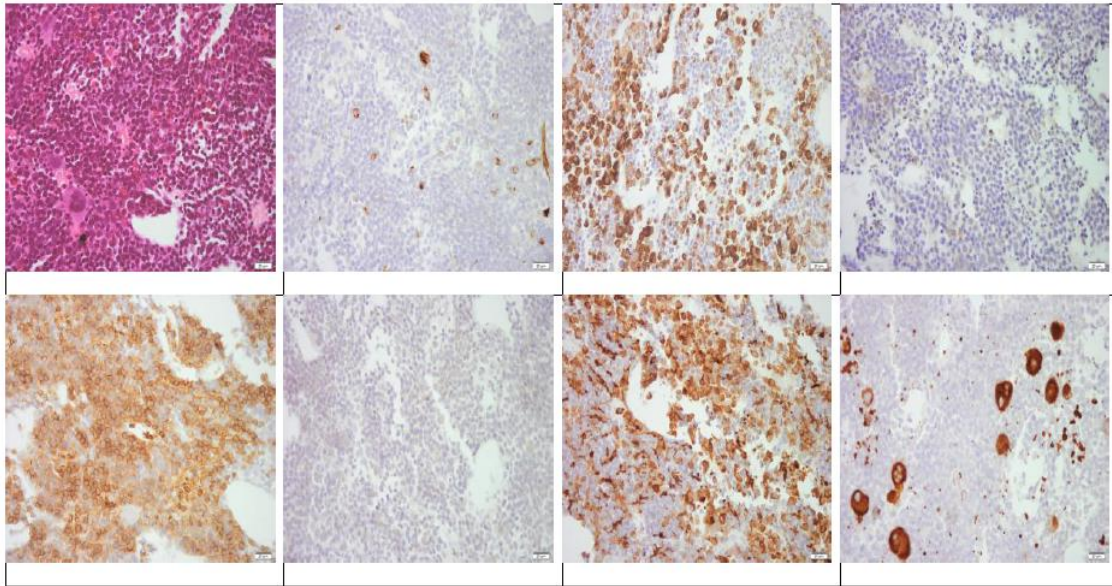
In this case, CD34, CD117 and TdT were negative, excluding a neoplastic proliferation. Glycophorin, MPO, CD61 and CD68 were used to confirm hematopoietic multilineage.

Extramedullary hematopoiesis should be considered in the differential diagnosis of an intrathoracic mass, particularly if a history of thalassemia or chronic anemia is known. Awareness of the differential diagnosis is important to avoid overdiagnosing this benign condition.

Hallmarks of Diagnosis

Personal history of thalassemia or chronic anemia; no evidence of blasts [rare or no expression of CD34, CD117, TdT].

Images (for full size images see supplements)



Keyword - Diagnosis: [Extramedullary hematopoiesis](#)

Keyword - differential diagnosis: [Extramedullary myeloid sarcoma](#)

Keyword - side findings:

Keyword - organ: [Posterior mediastinum, para-vertebral mass](#)

Keyword - methods: [CD34, CD117, TdT](#)

Online References (PubMed)

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