Extramedullary Hematopoiesis Presenting as Spinal Cord Compression in Thalassemia

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SENRS 2026 Interesting Case Presentation

VANDERBILT HEALTH

Clinical Presentation

- 34 y/o M with hx of thalassemia minor s/p childhood splenectomy
- Presented to ED with 4 months of lower back pain and ataxia, 10 days of bilateral leg stiffness and neurogenic claudication
- Neuro exam: spastic gait, generalized hypertonia, diffuse pathologic reflexes, 3-4 beats of clonus in bilateral ankles → upper motor neuron process
- Labs: leukocytosis, thrombocytosis, anemia, mildly elevated LFTs
- First step: non-contrast CT head

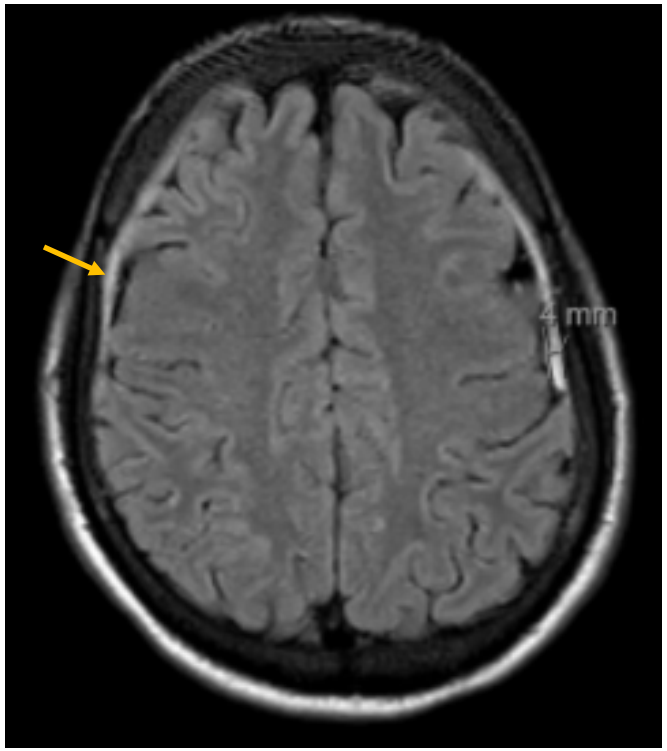
Imaging Discussion: CT head w/o contrast

- Diffuse calvarial marrow signal abnormalities, heterogeneity
- Lytic and ground-glass changes
- Thin lucencies in diploic space → **hair on end** appearance
  - Differential: anemia, sickle cell disease, G6PD deficiency, hyperparathyroidism, hereditary bone disorders, unknown malignancy
- Next step: neuroaxis imaging





Imaging Discussion: MRI brain and spine w contrast

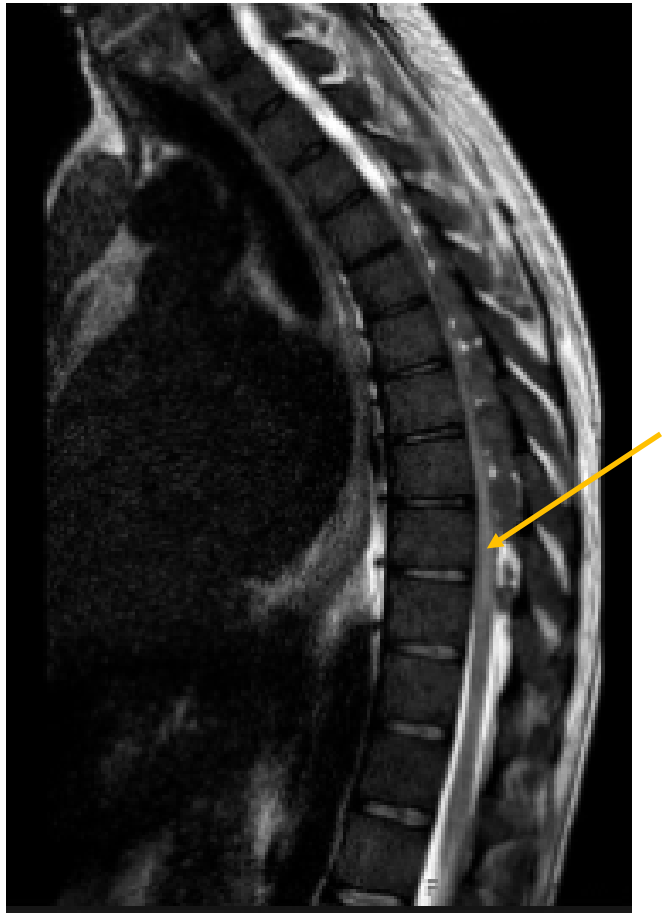


T2 FLAIR: bilateral frontotemporal pachymeningeal thickening and enhancement

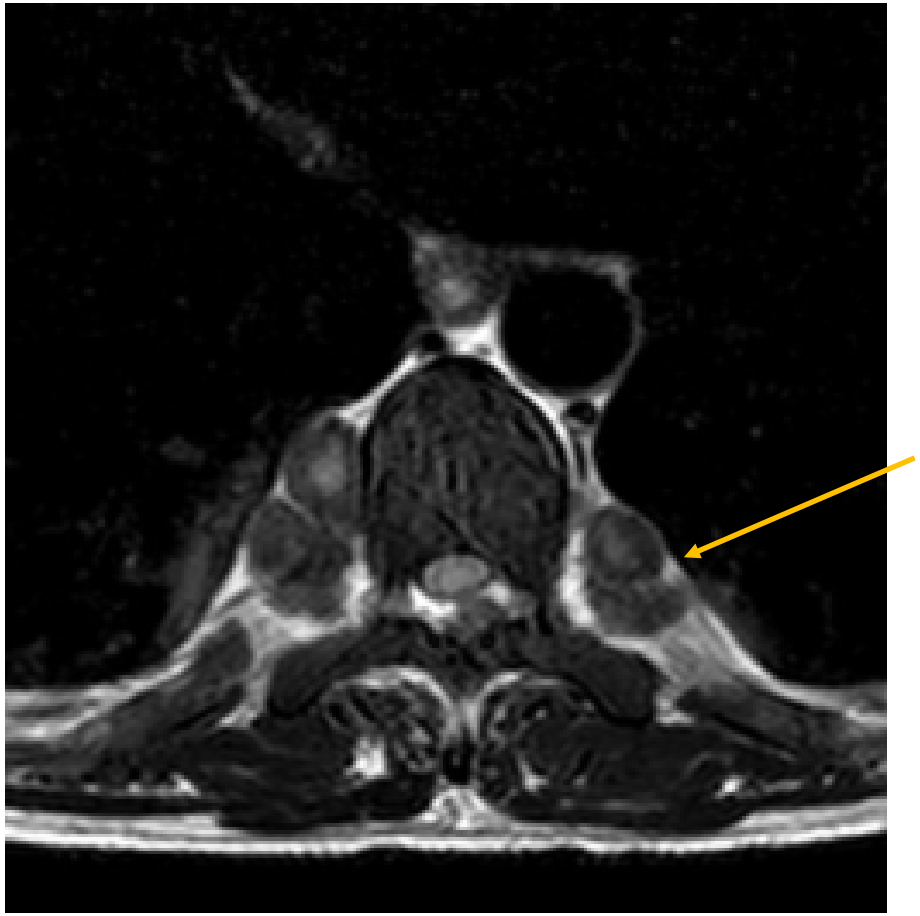


T1: thoracic spine (T4-T9) dorsal epidural space with hypo/isointense lobulated structures causing >50% spinal cord stenosis

Imaging Discussion: MRI brain and spine w contrast



T2: cord compression and edema

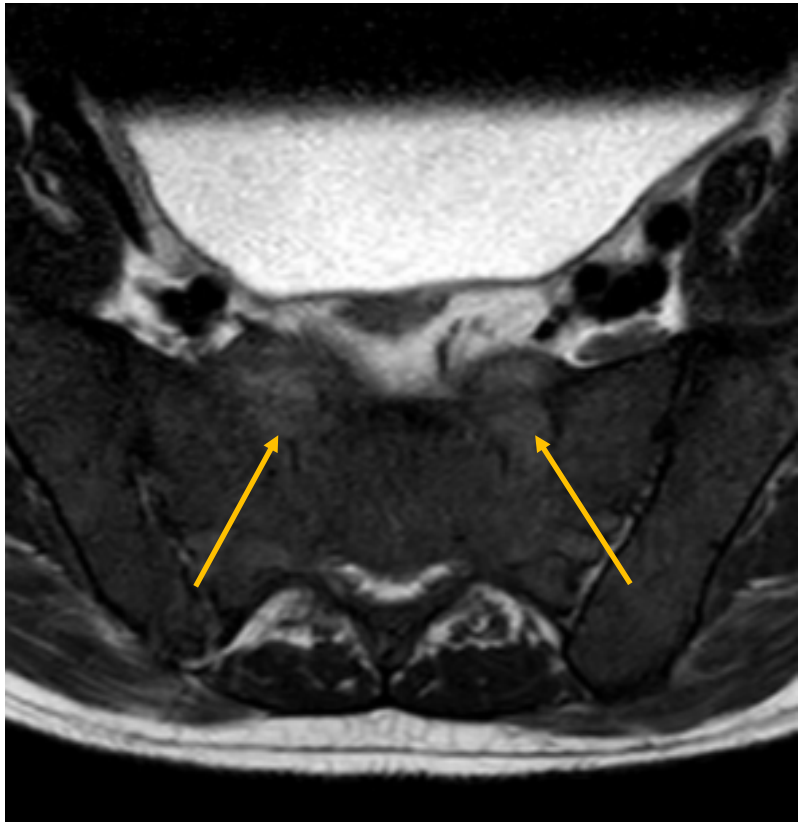


T2: soft tissue nodules at costovertebral angles

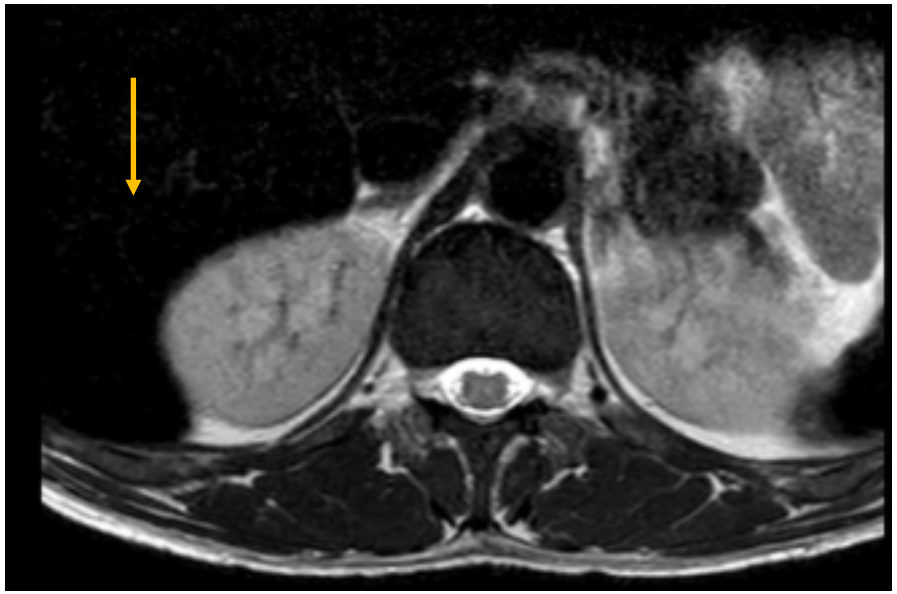
Imaging Discussion: MRI brain and spine w contrast



T1: hypointense lobules in L5-S1 fat around ventral cord, contributing to severe spinal stenosis

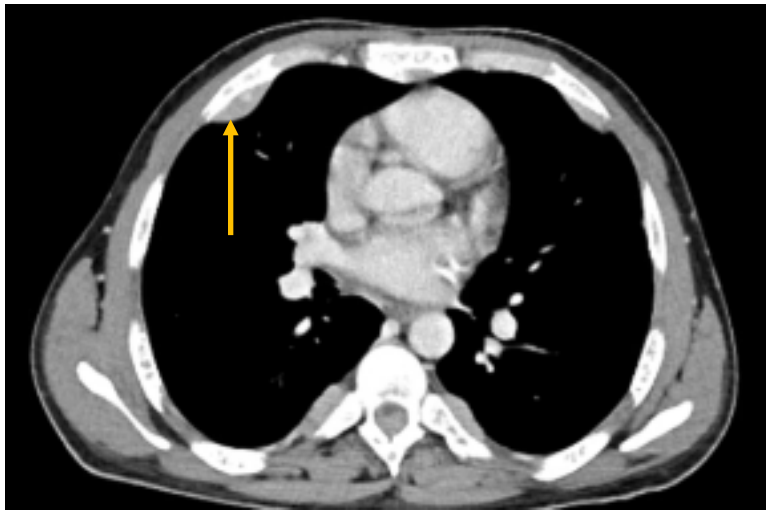


T2: lobulated, bulky soft tissue nodules anterior to sacrum and adjacent to vertebral bodies

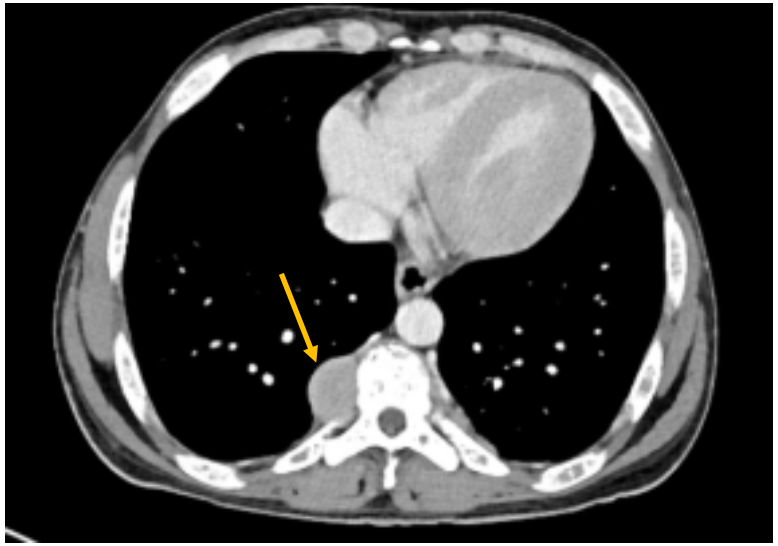


T2: hypointense liver signal due to chronic anemia

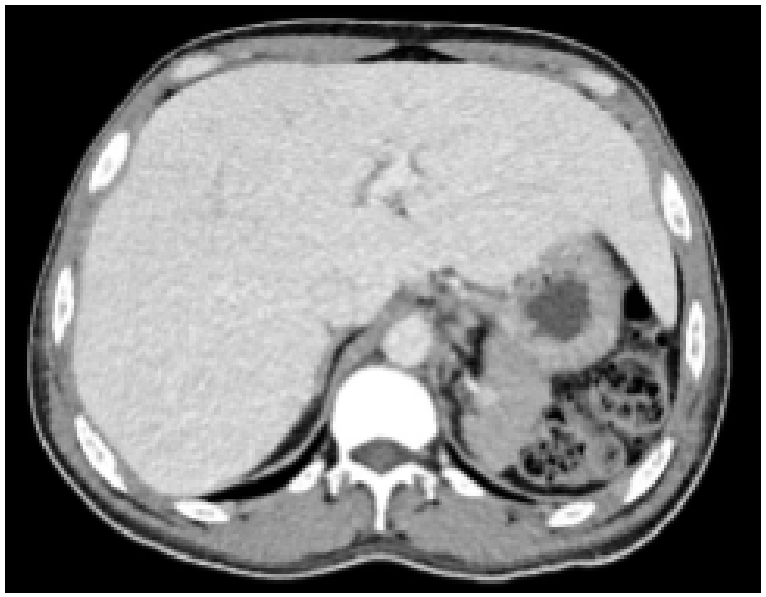
Imaging Discussion: CT chest, abdomen, pelvis w contrast



Pericostal soft tissue mass



Costovertebral junction mass



Hepatomegaly, surgical asplenia



Diffuse lytic/ permeative bone changes

Management

- Biopsy deferred due to compatible clinical and radiologic diagnosis, and high bleeding risk for biopsy of highly vascular epidural and paraspinal tissues
- Flow cytometry results: mixed B and T cell subsets without evidence of lymphoma, supporting extramedullary hematopoiesis over malignancy
- Extramedullary hematopoiesis management:
    - Blood transfusions to address the underlying cause
    - Steroids to reduce inflammation
- **RED FLAG SIGNS: spinal cord compression, neurologic compromise → risk of paralysis**
    - Emergency treatment: surgery (resection, laminectomy)

Outcome

- Final unifying diagnosis: extramedullary hematopoiesis in the setting of thalassemia minor
    - Classic radiologic distribution of soft tissue masses + underlying thalassemia + hepatomegaly
- Common sites for distribution of masses in extramedullary hematopoiesis:
    - Speen and liver (hepatosplenomegaly)
    - Bone marrow
    - Epidural space
    - Paraspinal soft tissues
    - Thorax
- Although this patient should have received emergent surgical decompression, he ultimately left against medical advice and was lost to follow up due to lack of insurance.

Take-Home Points

- Extramedullary hematopoiesis should be considered in anyone with a hemoglobinopathy presenting with epidural or paraspinal masses causing cord compression.
- Classic imaging clues include hair-on-end calvarial changes and bilateral fat-containing paraspinal masses with a peripheral fat rim.
- Extramedullary hematopoiesis may mimic lymphoma radiographically, as it tends to distribute along the liver and spleen (hepatosplenomegaly), bones, paraspinal muscles, and epidural space.
- Biopsy of epidural/spinal cord lesions should be reserved for scenarios without a unifying diagnosis due to bleeding risk.
- Cord compression from extramedullary hematopoiesis is a neurologic emergency, managed with transfusion, steroids, and/or surgical decompression.

References

Huang, A.H., Sun, H.H., Skolnick, G.B. et al. Thickness of calvarium and diploic space in children ages 0 to 17 as assessed by computed tomography. Eur J Plast Surg 38, 193–198 (2015). https://doi.org/10.1007/s00238-014-1060-3

Lam NG, Ahmadpour A, Gimelfarb A, Nanda R, Ares WJ. Epidural Spinal Cord Compression due to Extramedullary Hematopoiesis: Systematic Review of the Literature with an Illustrative Case. World Neurosurg. 2025 Jan;193:119-130. doi: 10.1016/j.wneu.2024.10.034. Epub 2024 Nov 9. PMID: 39427876.

Radswiki T, Weerakkody Y, Silverstone L, et al. Extramedullary hematopoiesis. Reference article, Radiopaedia.org (Accessed on 28 Apr 2026) https://doi.org/10.53347/rID-12220

Roberts AS, Shetty AS, Mellnick VM, Pickhardt PJ, Bhalla S, Menias CO. Extramedullary haematopoiesis: radiological imaging features. Clinical radiology. 2016;71(9):807-814. doi:10.1016/j.crad.2016.05.014

Weerakkody Y, Niknejad M, Hair on end sign (mnemonic). Reference article, Radiopaedia.org (Accessed on 28 Apr 2026) https://doi.org/10.53347/rID-9384

Thank you!

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