Not a Red Herring?:

A Potential Association of a Very Rare Spinal Tumor

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Disclosures

- None of the authors nor their immediate family members have a financial relationship with a commercial organization that may have direct or indirect interest in the content.

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Clinical Presentation

- 58-year-old female presents with history of stroke, thrombocytosis, and peripheral neuropathy
- Plasma cell dyscrasia identified on recent bone marrow biopsy
- Reported rapidly progressive numbness and weakness in lower extremities
- CT CAP performed for malignancy workup
- Epidural mass compressing spinal cord noted on thoracic MRI

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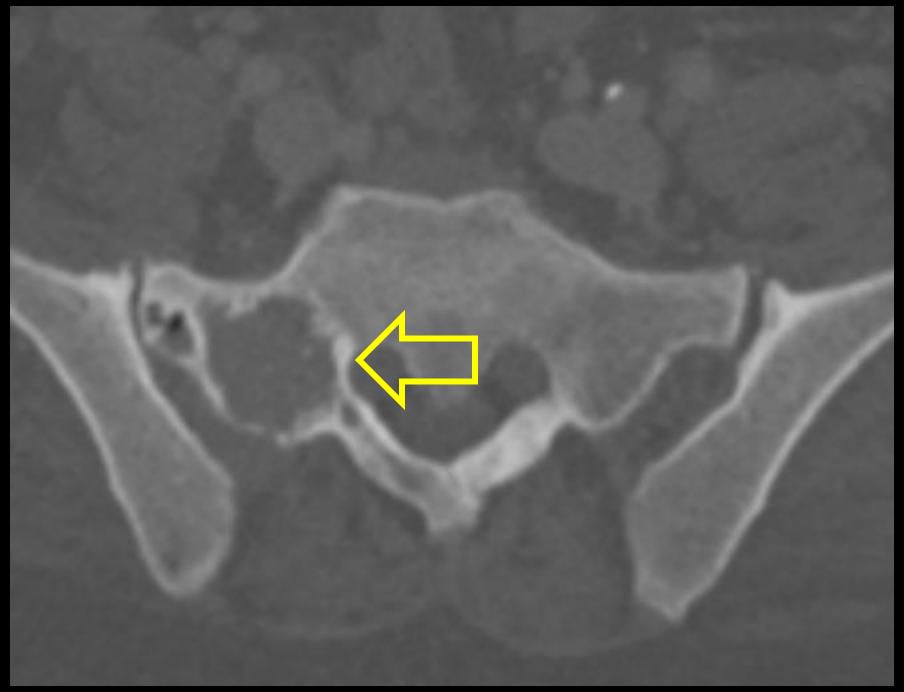
Imaging (CT CAP)



- Lytic lesion at the right sacral ala with a few small areas of cortical disruption, involving the right SI joint space, measuring 3.5 x 2.4 x 3.8 cm

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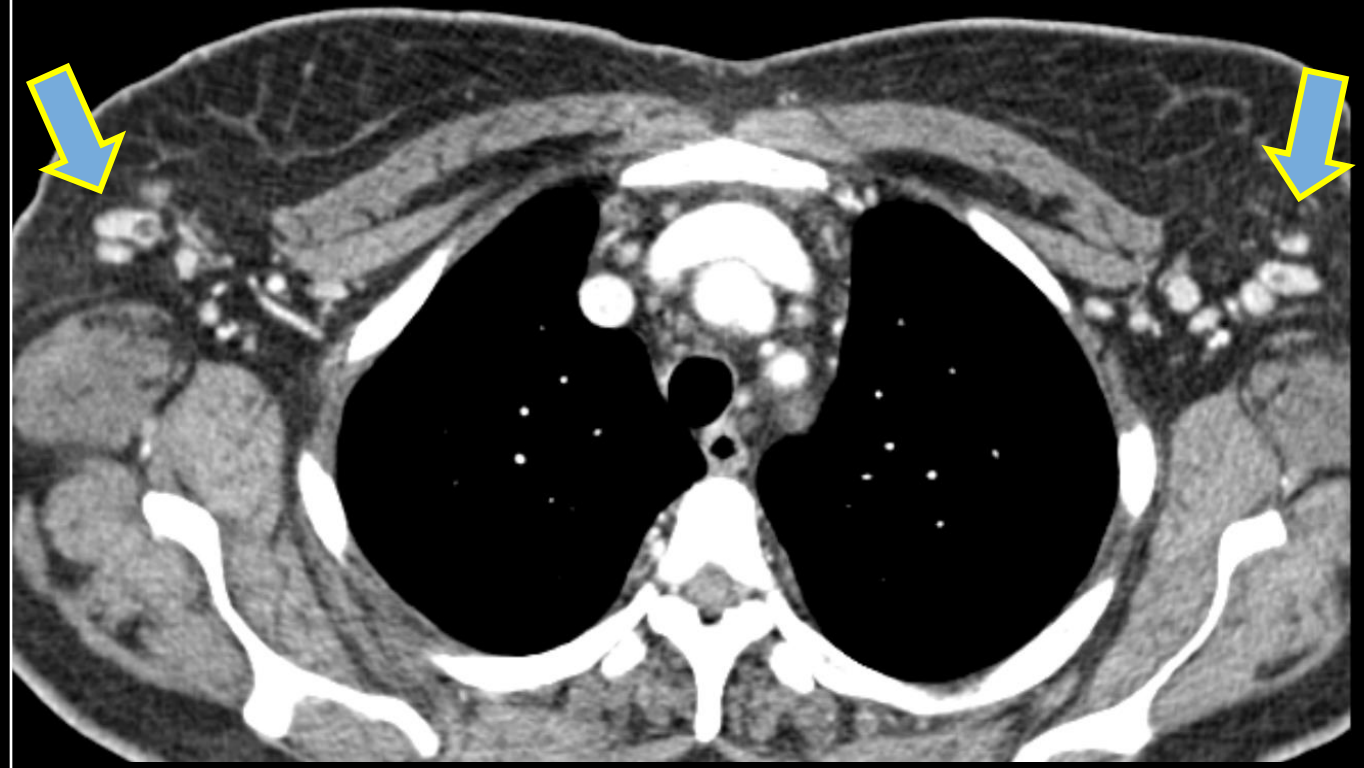
Imaging (CT CAP)



- Diffuse hypervascular lymphadenopathy most prominently in the axillary nodal stations

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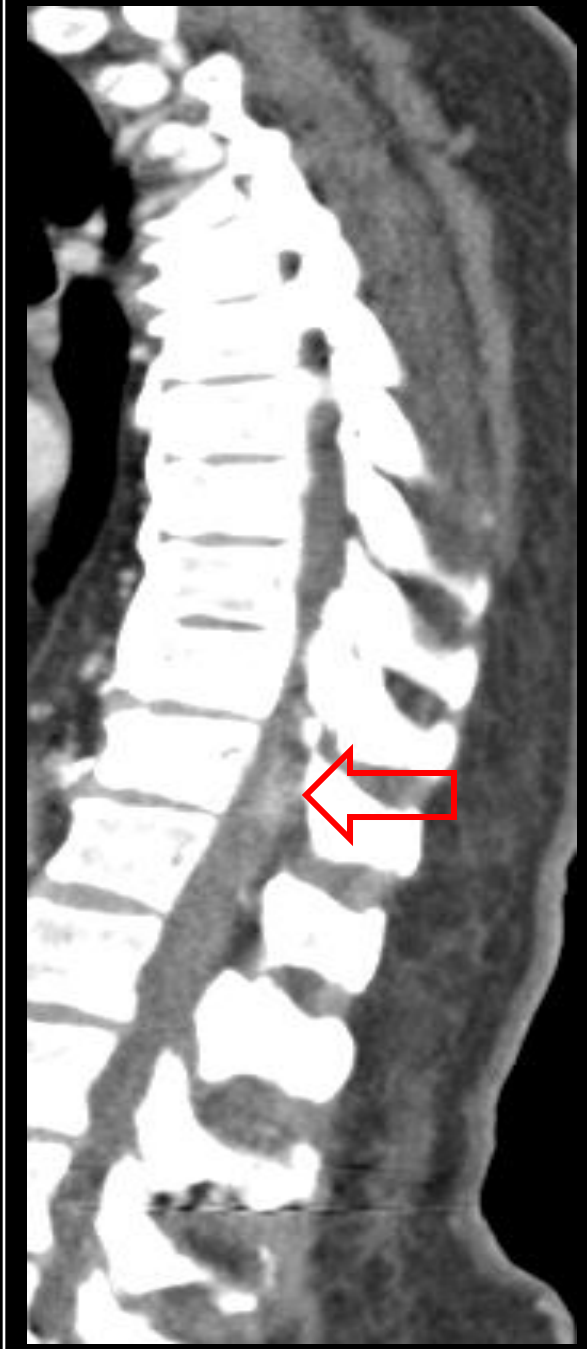

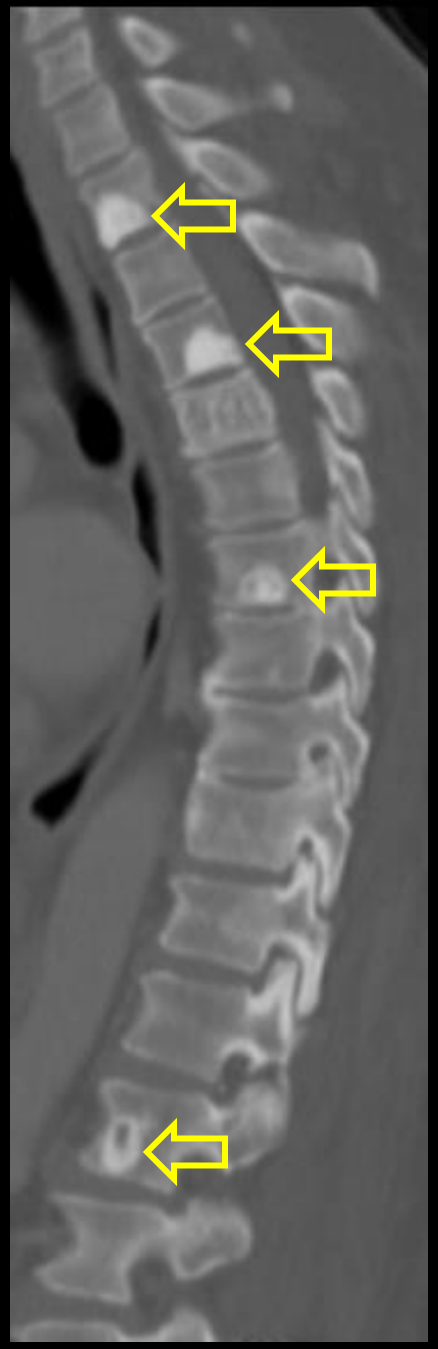

Imaging (CT CAP)

- The epidural mass is interspersed with fat
- Scattered sclerotic vertebral body lesions involving T2, T4, T7, T8 and L1
- Sclerotic lesion in the right pedicle of T11 is expansile and results in mild narrowing of the spinal canal

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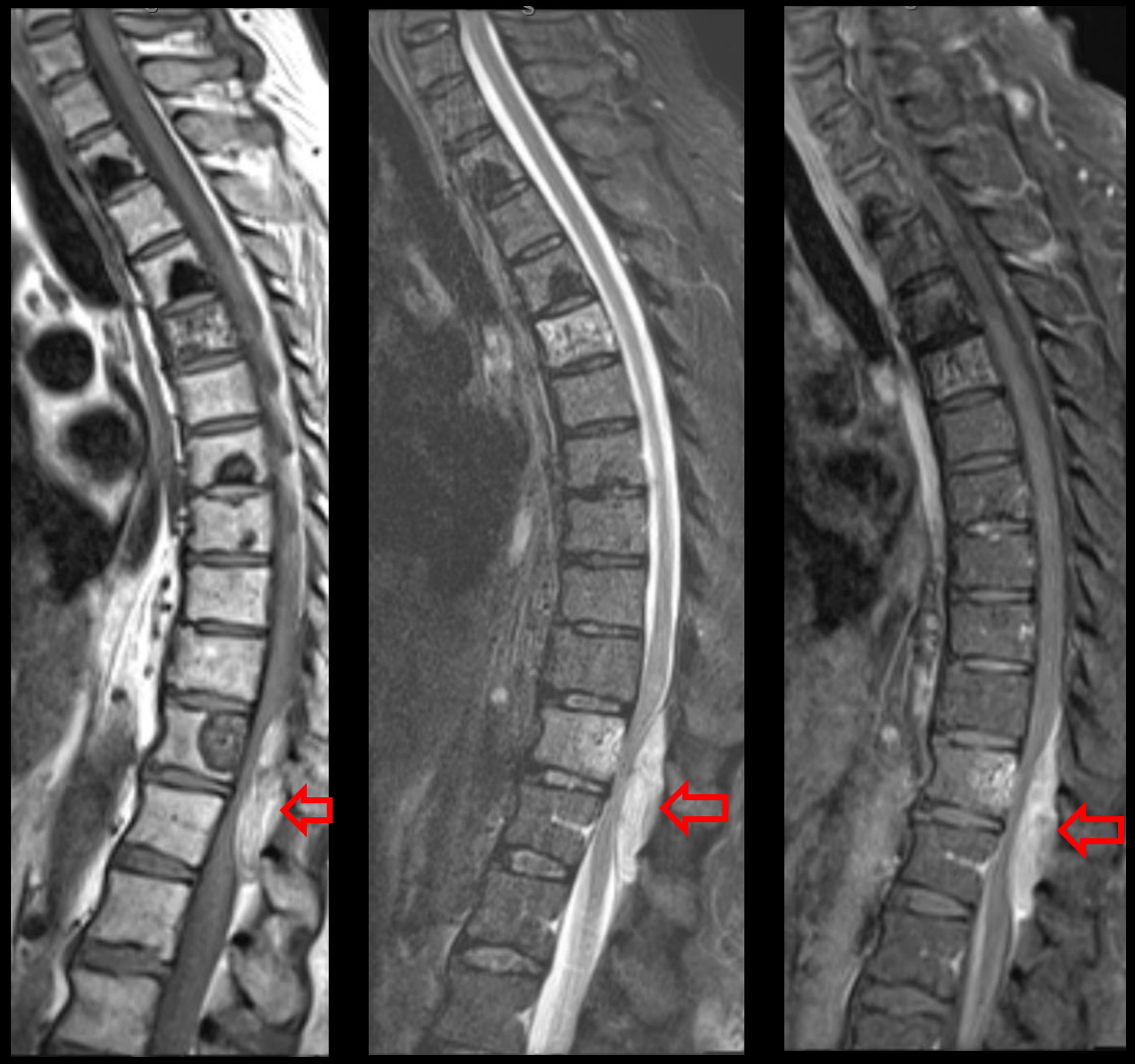
Follow-Up Imaging (MRI Thoracic Spine)



- Enhancing dorsal epidural mass compressing the spinal cord, with T1 hyperintense components that are mostly STIR hyperintense with a small sliver of interspersed macroscopic fat
- Adjacent T1 hyperintense, STIR hyperintense enhancing lesion in the posterior T11 vertebral body at the level of the epidural mass

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Management

- Patient underwent thoracic laminectomy for resection of epidural mass
- Biopsies of multiple sclerotic vertebral lesions were obtained during surgery
- She began acute rehabilitation with PT/OT and was discharged on day 10 to an inpatient rehabilitation facility
- Epidural mass pathology was consistent with benign angiolipoma

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Outcome

- On one month follow-up visit, she reported persistent numbness in upper and lower extremities but improvement in lower extremity movement
- She reported episodes of nausea, hypotension, and tachycardia
- During her three-year follow-up visit, she denied numbness in lower extremities but continued to show signs of polyneuropathy
- She was diagnosed with POEMS Syndrome based on clinical findings
- She receives care from hematology for thrombocytosis and IgG lambda gammopathy

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A Word About POEMS Syndrome

- POEMS syndrome consists of Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal gammopathy, and Skin changes
- It is a paraneoplastic disorder
- There are mandatory, major, and minor criteria
  - Mandatory: polyneuropathy, monoclonal plasma cell proliferative disorder
  - Major: sclerotic bone lesions, Castleman disease, elevated VEGF levels
  - Minor: organomegaly, endocrinopathy, skin changes, extravascular volume overload, optic disc swelling, thrombocytosis/polycythemia

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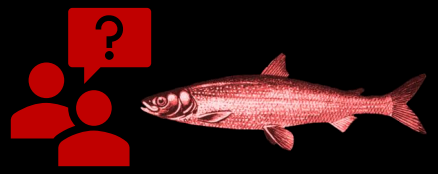

Spinal Angiolipomas

- Spinal angiolipomas are very rare benign lipomatous neoplasms
- Adults, 4th-5th decade, women > men
- Potential association between spinal angiolipomas and POEMS syndrome suggests a shared vascular pathogenesis
  - POEMS syndrome features include elevation of VEGF, hypervascular lymphadenopathy in the form of angiofollicular lymphoid hyperplasia, and glomeruloid hemangiomata in the skin.
  - Angiomyolipomas, rare vascular benign neoplasms that can occur in the epidural space, have been reported in at least one other case report of POEMS syndrome.
  - Given their vascular nature, we propose that this may be a previously unrecognized association between spinal angiolipomas and POEMS syndrome, providing insight into their pathogenesis.
  - At least one other case report exists with a spinal AML in a POEMS patient[1]

1. Al-Mayoof O, Al Sughaiyer H, Abuomar W, Khan M. POEMS syndrome: a rare cause of exudative ascites and chronic peripheral neuropathy. BMJ Case Rep. 2017 Jun 20;2017:bcr2016219022. doi: 10.1136/bcr-2016-219022. PMID: 28637843; PMCID: PMC5534653.

Take Home Points

- Spinal angiolipomas can mimic metastatic disease on imaging, potentially leading to diagnostic or treatment errors
- Key imaging clues include macroscopic fat and intrinsic T1 hyperintensity, which help distinguish angiolipomas from epidural malignancy
- Multiple spinal sclerotic lesions in a patient with hypervascular adenopathy or other features of POEMS syndrome should raise concern for this diagnosis

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