Leptomeningeal Myelomatosis: An enhanced look into a rare cause of weakness

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OHSU

INTRODUCTION

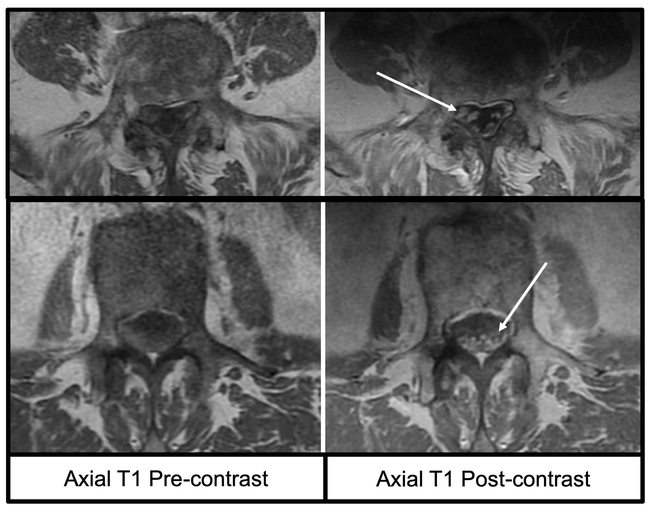
- Multiple myeloma (MM) is a clonal plasma cell neoplasm that can manifest with classic symptoms of hypercalcemia, renal disease, anemia, and bone lesions ("CRAB" criteria).
- Treatment involves multiagent chemotherapy, with consideration of autologous stem-cell transplant.

CASE

Patient
- 65-year-old male with refractory MM was started on ixazomib (proteasome inhibitor) as 3rd line therapy.
- Developed acute-onset generalized weakness and an episode of bowel and bladder incontinence.

Exam
- Vitals unremarkable. 4/5 strength in his bilateral lower extremities, mildly decreased rectal tone

Studies
- Worsening pancytopenia
- MRI total spine showed lumbar canal stenosis and diffuse smooth enhancement of cauda equina nerve roots



- Neurosurgery had no concern for cord compression or cauda equina syndrome given lack of correlation of imaging and exam.
- Differential included deconditioning given co-morbidities; nerve root enhancement was possibly post-surgical changes from remote L-spine decompression. Could not exclude leptomeningeal myeloma.

IMAGES

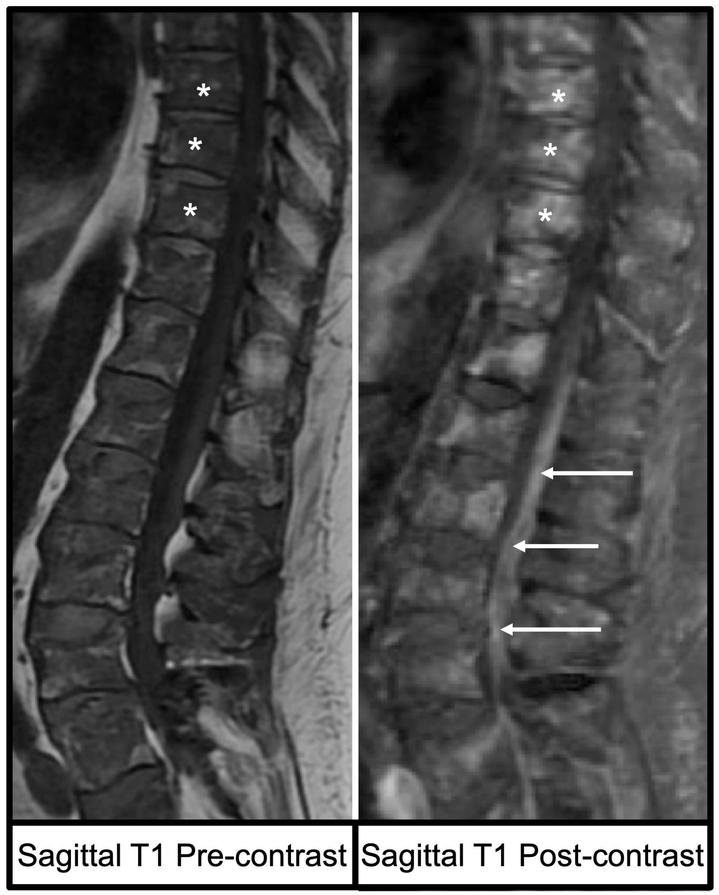


Fig 1 (Above)
Sagittal T1 images of the lumbar spine show enhancement of the cauda equina nerve roots (arrows) on post-contrast sequence. Heterogeneous T1 hypointensity in the vertebral bodies on the pre-contrast sequence with corresponding heterogeneous enhancement on the post-contrast sequence (*) is consistent with multifocal multiple myeloma lesions.

Fig 2 (Left)
Axial T1 images of the lumbar spine demonstrate smooth enhancement of the cauda equina nerve roots (arrow), involving all of the visualized nerve roots, a nonspecific finding.

CASE CONCLUSION

- CSF from LP with lymphocyte predominance and M-spike; flow cytometry revealed monoclonal plasma cells with CD20, CD38, CD138, kappa+; FISH showed t(11;14).
- Leptomeningeal myelomatosis diagnosed. Transitioned to comfort care for worsening cytopenias and infection, died soon after.

DISCUSSION

- CNS myeloma is rare, <1% of MM patients.
- Often hematogenous or contiguous spread from nasopharyngeal or intraparenchymal plasmacytoma, extension from skull, or leptomeningeal disease.
- Cerebral symptoms include visual changes, headache and seizure. Spinal cord symptoms include radiculopathy, sensory changes and motor loss.
- **Imaging modality of choice:** MRI with contrast
- **Gold standard:** CSF cytology
- No defined optimal treatment, chemotherapy does not significantly improve prognosis. Poor prognosis, literature suggests 1-4 months survival from time of diagnosis.

Take away

- CNS myeloma can be a challenging diagnosis due to mild, heterogenous, and nonspecific symptoms in a complex picture (treatment side effects, deconditioning, cytopenia).
- In myeloma patients with nonspecific neurologic symptoms, MRI with contrast of CNS is best.
- Lower threshold for LP to examine CSF for unexplained CNS findings.

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