Disseminated Oligodendroglioma-Like Leptomeningeal Neoplasm
A Case Report

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Overview

- Introduction
- Radiology
- Histology
- Case presentation

Introduction

- Rarely encountered neoplasm predominantly seen in children
- Diffuse Leptomeningeal thickening
- Incidence difficult to assess
- Primarily Leptomeningeal?

Introduction

- Presenting symptoms include: focal neurologic deficits, elevated intracranial pressure, seizures and altered mental status
- Imaging: diffuse leptomeningeal thickening and enhancement involving the brain and spinal cord
- Mimics infectious or inflammatory etiology or high grade metastatic neoplasm
- Hydrocephalus is common and often requires a shunt

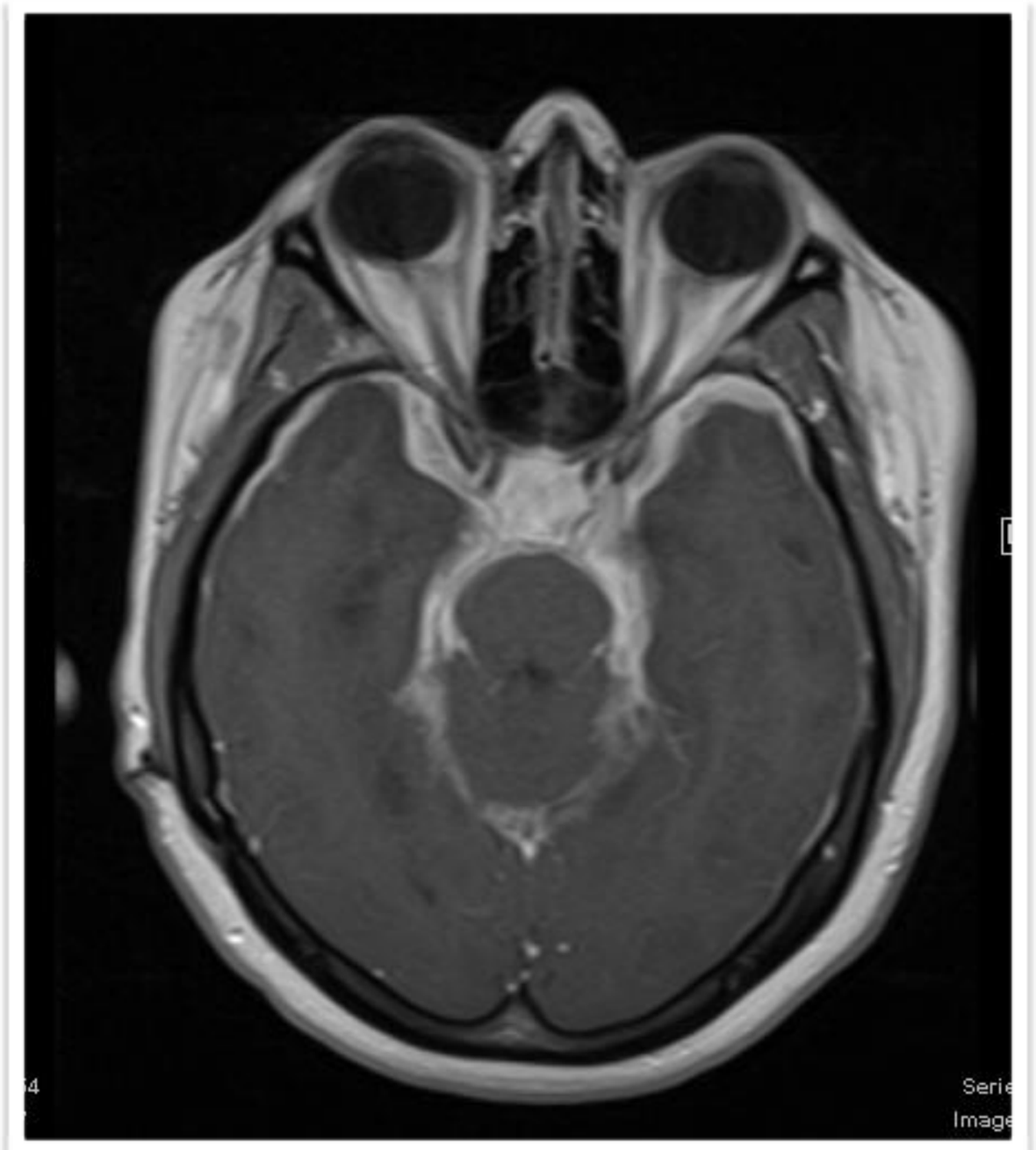
Introduction

- Intraparenchymal disease is variable and may not be seen at presentation
- Development of lesions in the spinal cord have been shown in follow up imaging
- Extent of disease is often confined to the leptomeninges, brain and spinal cord.

Introduction

- CSF demonstrates elevated protein with no neoplastic cells
- Biopsy may yield unclear results due to small sample size and/or uneven spread of leptomeningeal thickening and fibrosis.

Radiology



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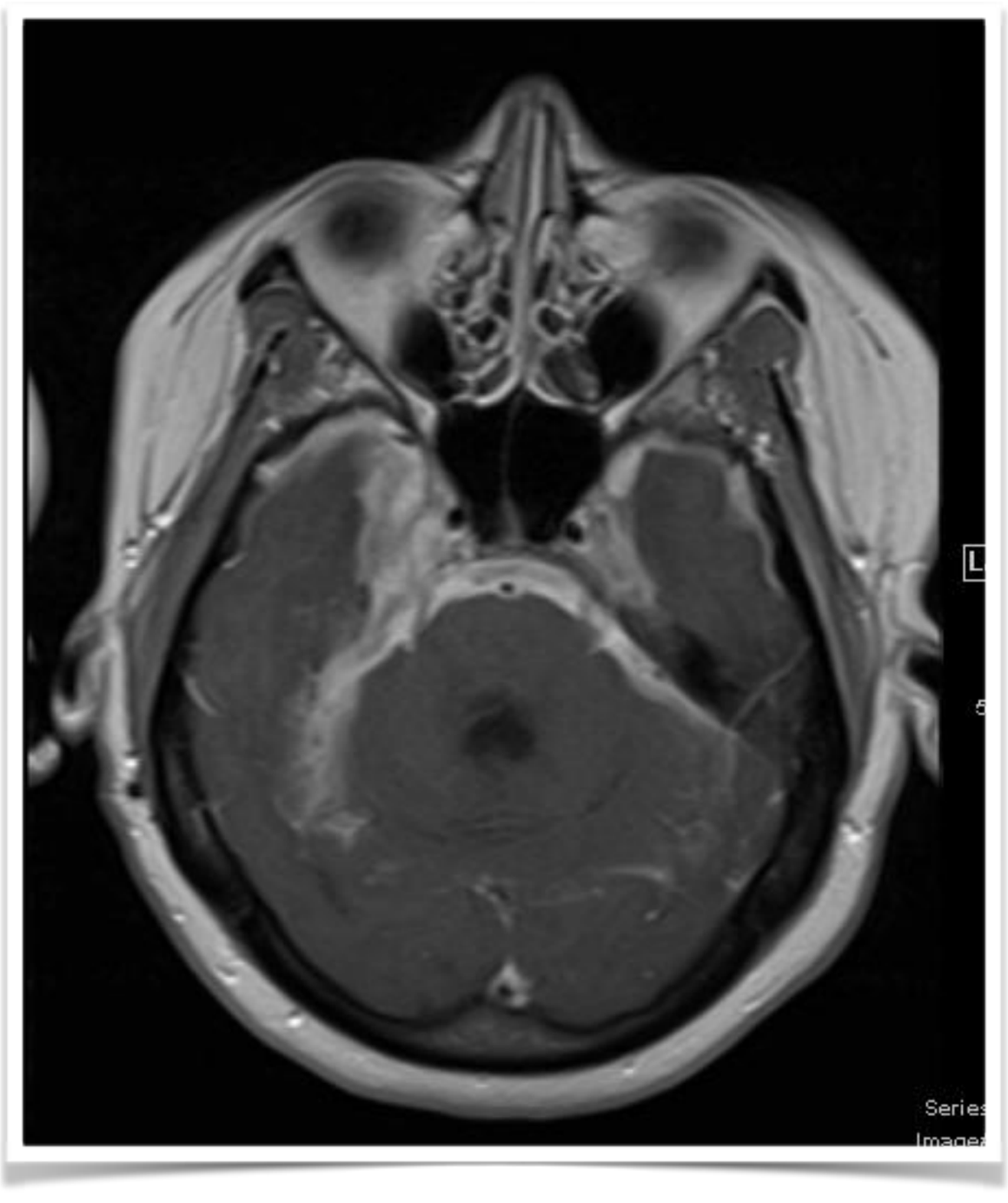

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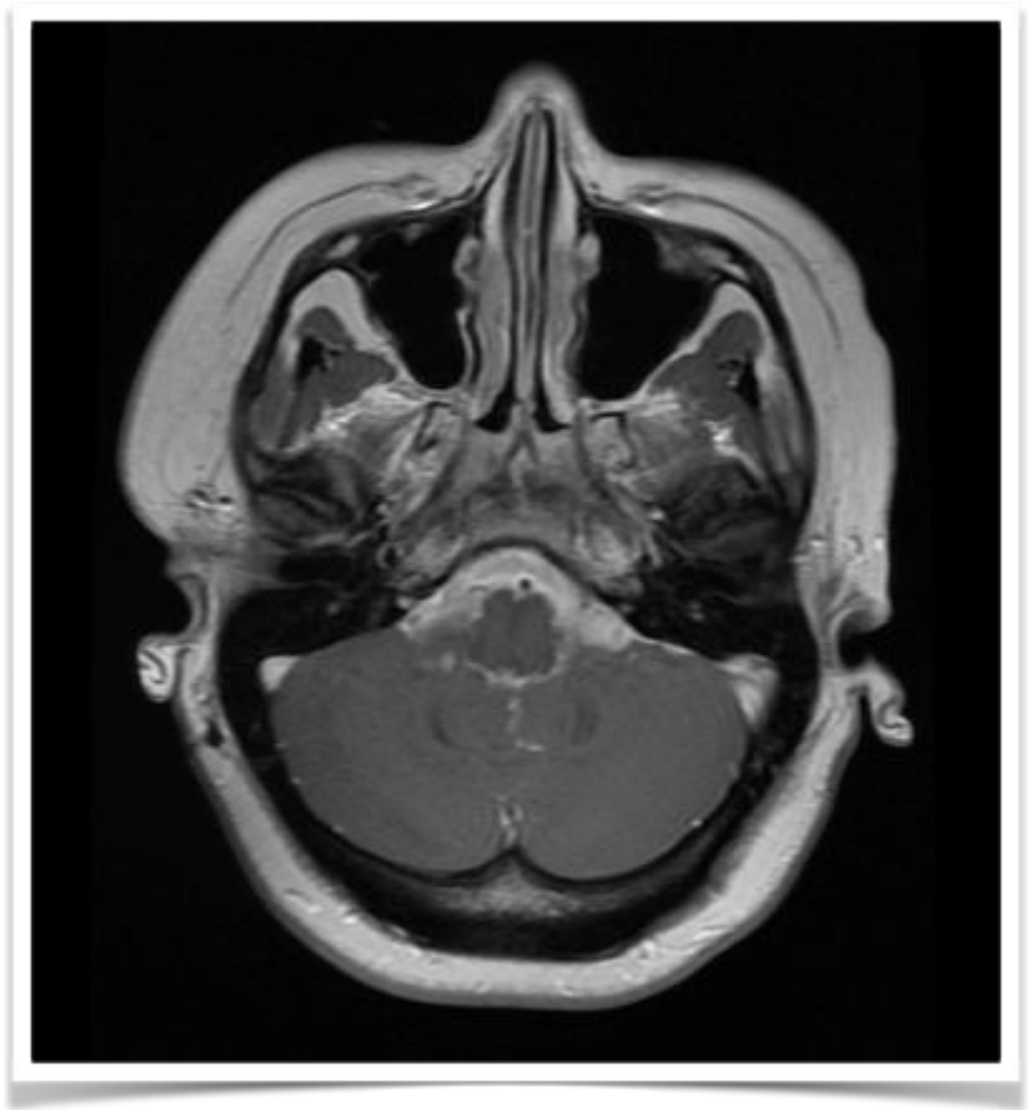
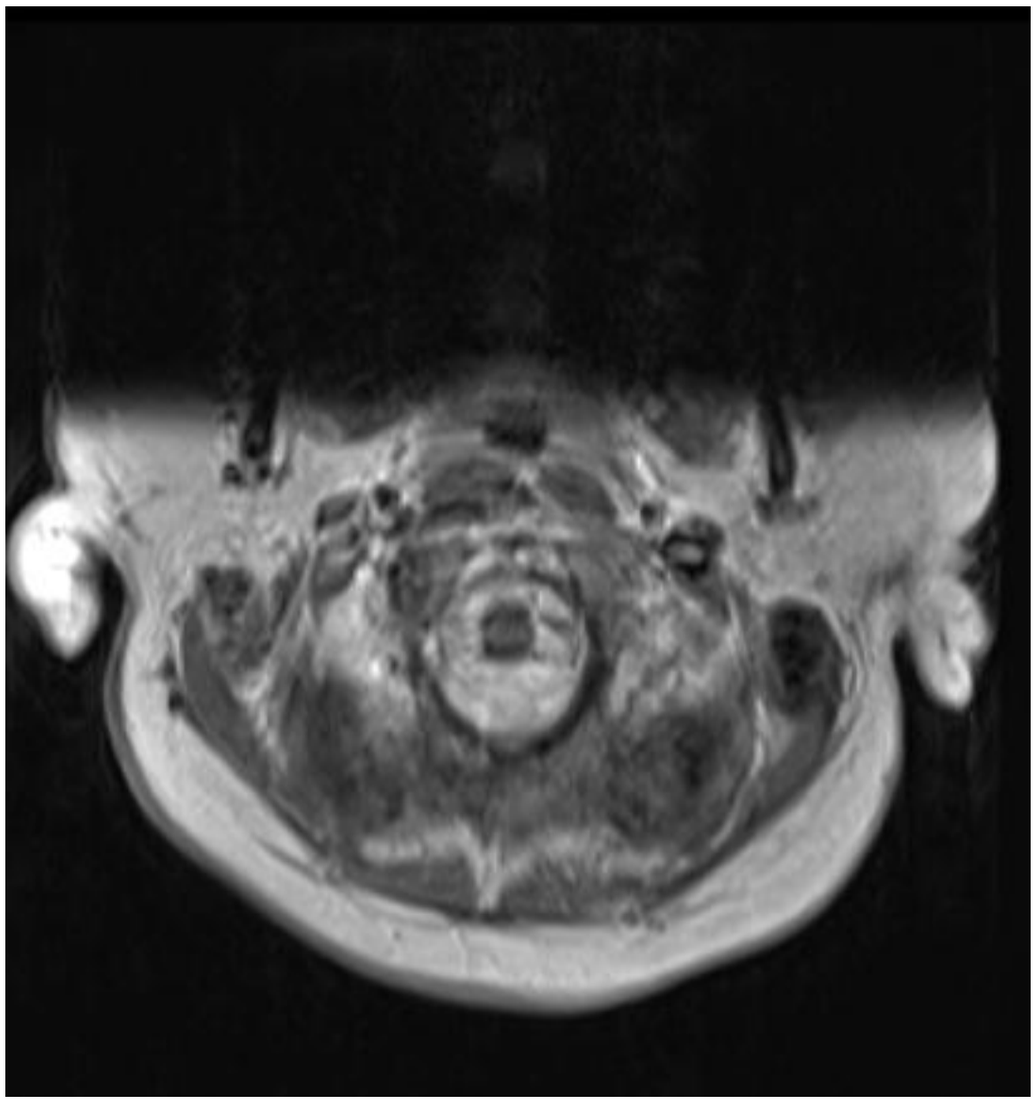


Radiology



Histology

- Grossly thickened and fibrotic leptomeninges, pial involvement
- Uniformly OLIG-2 positive
- Inconsistent immunoreactivity for GFAP, synaptophysin, and NeuN and have been described as glial, glioneural and neurocytic
- 1p19q and IDH1 or 2 mutations

Disease progression/treatment

- Indolent with survival but with substantial morbidity
- Optimal treatment is unknown
- Chemotherapy followed by craniospinal irradiation

Case Presentation

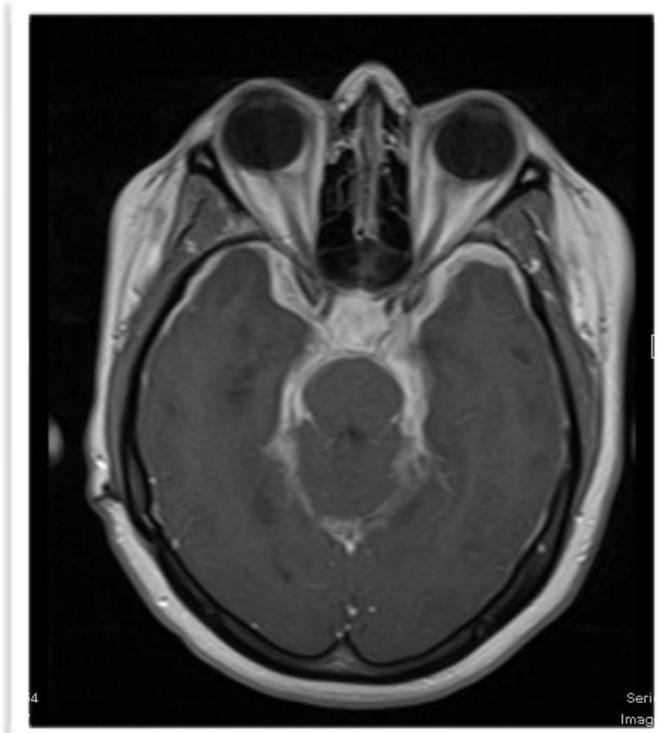
- 31yo F presents with 6-8 week history of slowly worsening headache, memory loss, gait imbalance and nausea. She had an acute episode of aphasia with altered mental status (a blank stare) for 6 hours. Her symptoms progressed to include bilateral lower extremity weakness

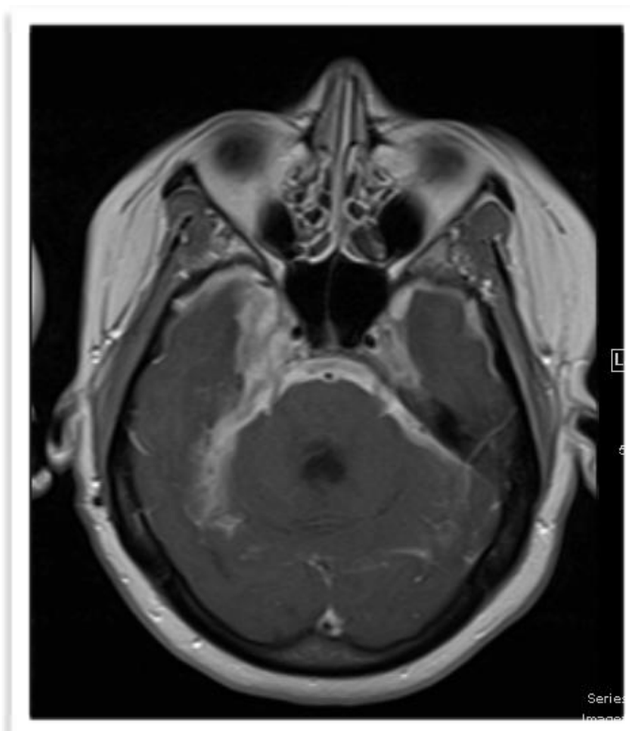
- PMH: VPS at age 3 due to hydrocephalus of unknown etiology. Revised once a year prior to presentation due to fractured tubing.

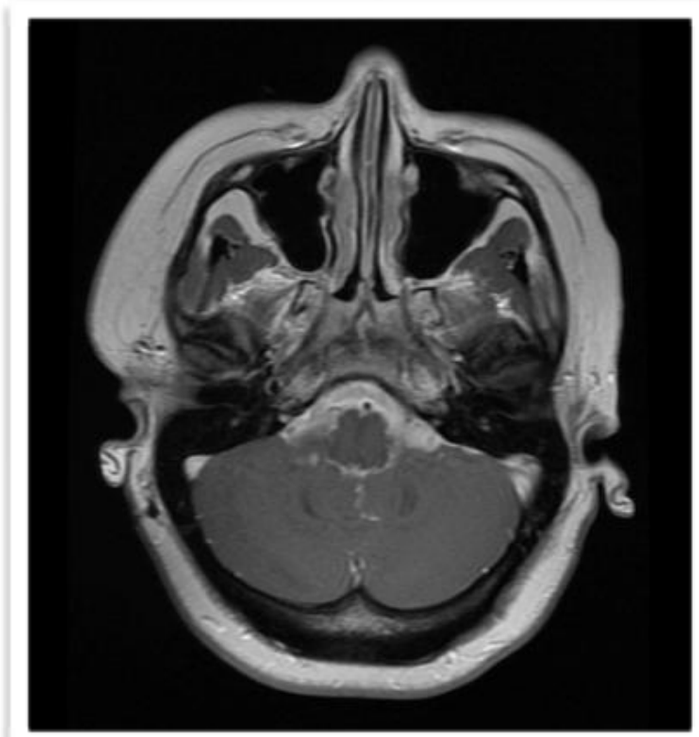
Case Presentation

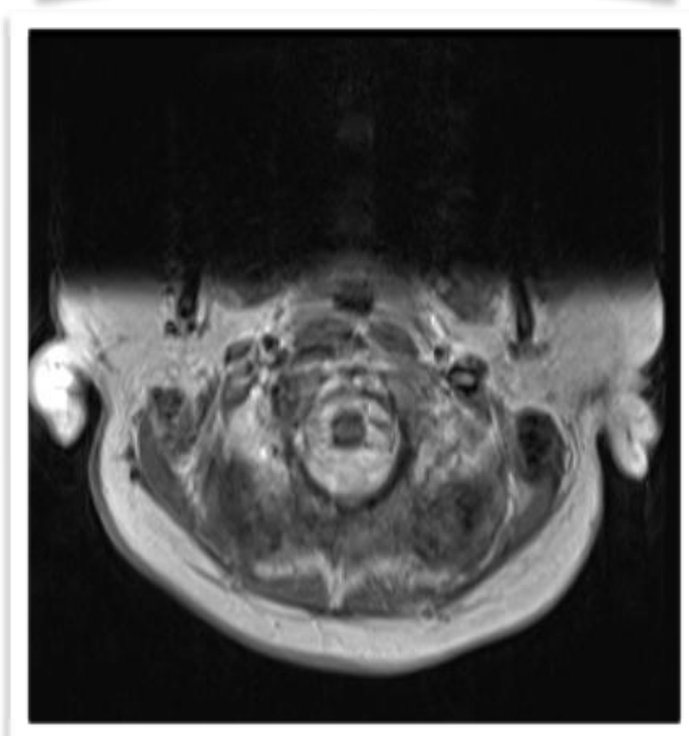
- Imaging showed extensive Leptomeningeal enhancement
- WBC 16K, Afebrile
- Admitted and treated for meningitis with shunt externalization. Cultures returned negative

Case Presentation Imaging

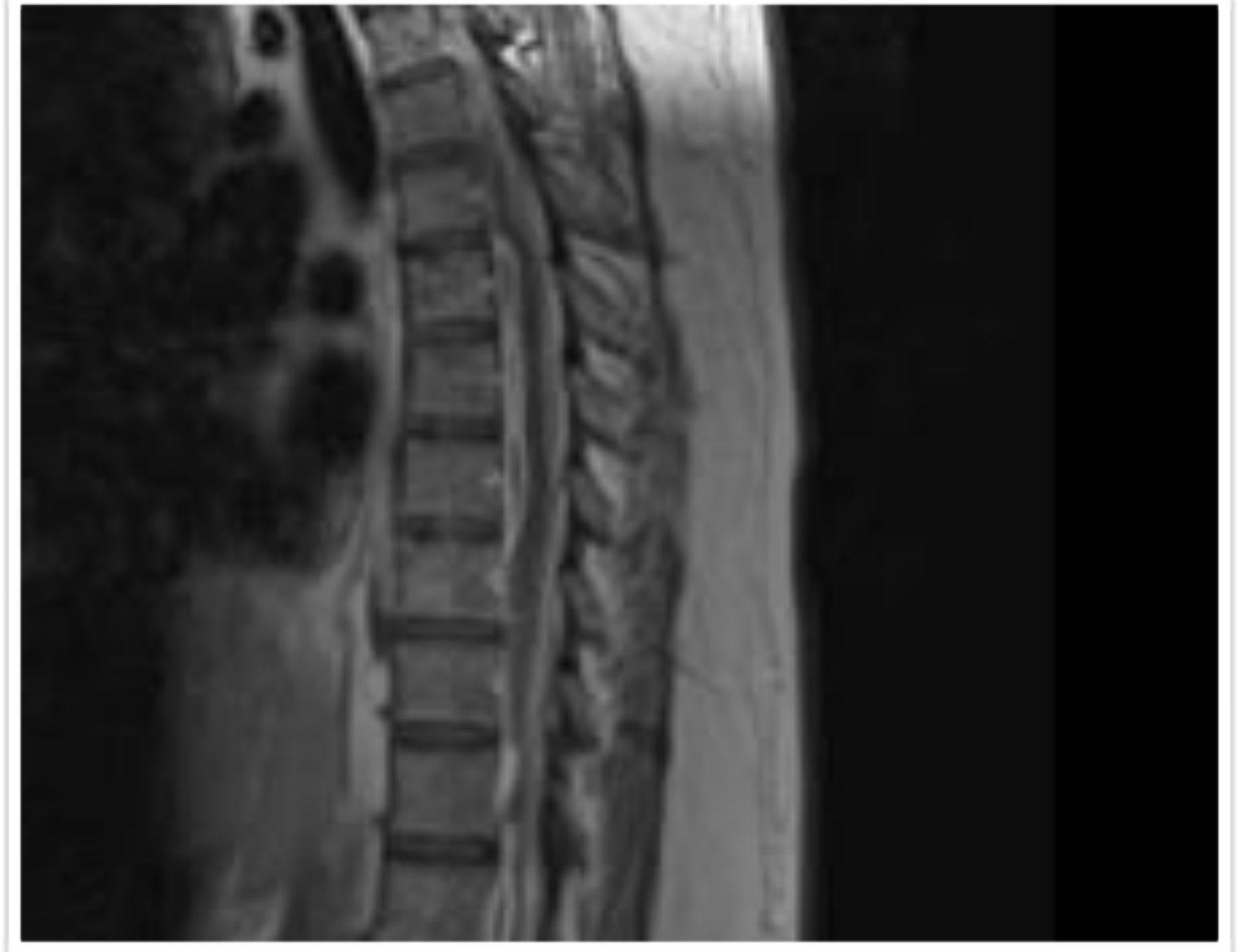








Case Presentation-Imaging



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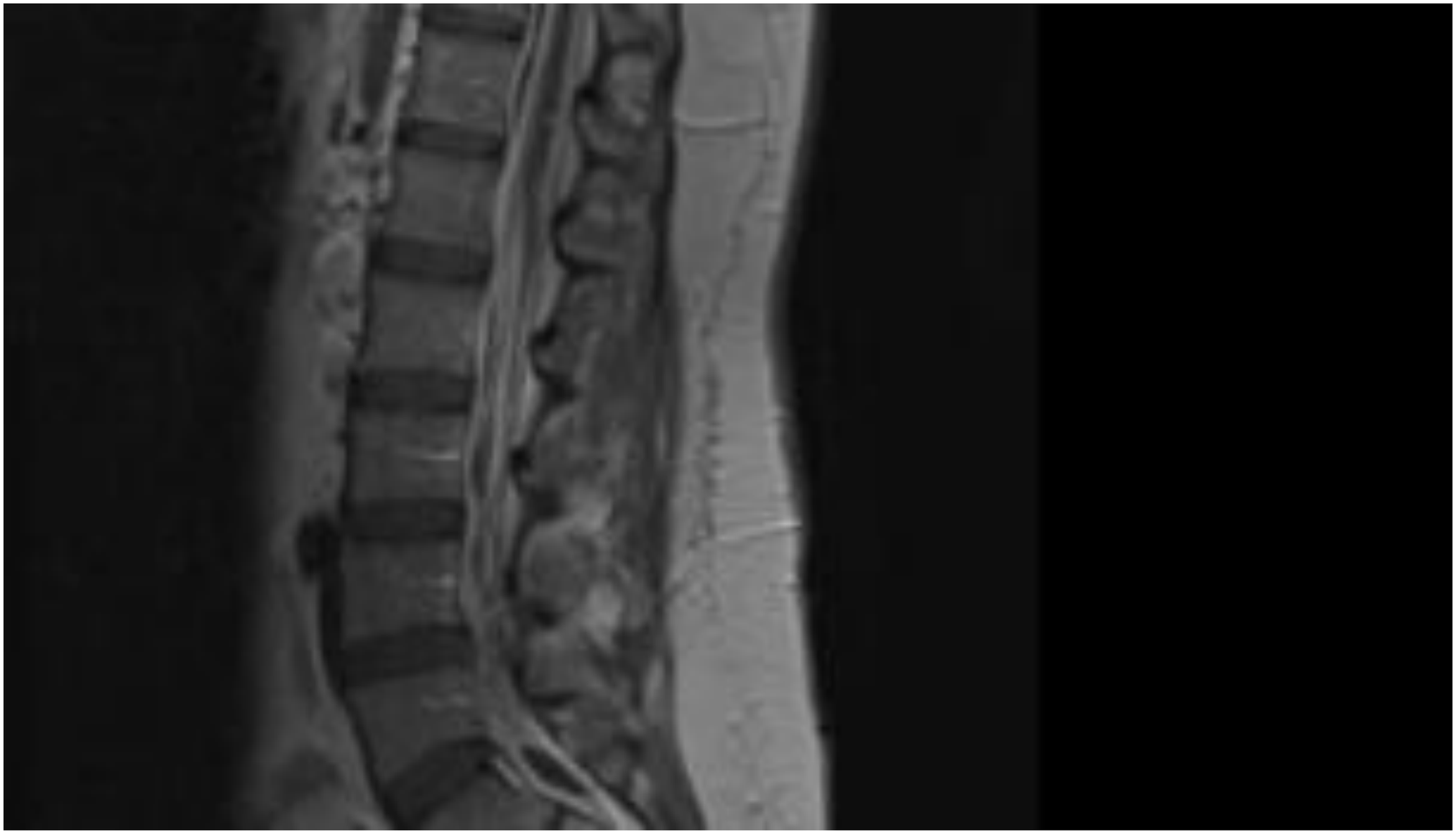

Case Presentation

- No improvement after treatment for meningitis. Biopsy performed, right temporal crani, dural excision.

Case Presentation-Pathology

- Severe fibrous thickening, crushed cells, and focal nests of cells with rounded nuclei and clear cytoplasm
- Scattered atypical mitotic figures
- Strongly positive for S100 and synaptophysin
- Focally positive for GFAP and OLIG-2
- Negative for IDH 1 or 2 mutation
- Loss of 1p, no 19q loss

Case Presentation

- Variable prognosis

- Discharged to hospice

Conclusion

- Rare diagnosis
- More common in children
- Mimics infection
- No clear treatment
- Poor prognosis? Less than 1yr survival