Rapid Onset Thoracic Myelopathy Due to an Epidural Sarcoid-like Lesion in a Pediatric Patient

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Introduction

Isolated intraspinal neurosarcoidosis is a rare clinical entity, with most reports describing intramedullary involvement in adults. We detail a nine-year-old girl with rapid onset compressive myelopathy secondary to a thoracic epidural lesion. Although pathological diagnosis was challenging, a presumptive diagnosis of isolated extradural neurosarcoidosis was made in light of the patient's investigations and dramatic response to corticosteroids. Though less likely than neoplasia, rheumatologic processes such as inflammatory granulomatous disease warrant consideration in similar cases.

Methods

This nine-year-old otherwise healthy girl experienced two weeks of falls, progressively worsening lower extremity weakness, and bilateral thigh pain. She also developed episodic urinary incontinence, with urinary retention upon presentation. There were no antecedent infections or infectious symptoms. Pertinent neurological exam findings revealed evidence of thoracic myelopathy.

Results

Magnetic resonance imaging (MRI) of the brain and complete spine revealed an extensive circumferential enhancing epidural mass centered around T3-T6 causing severe spinal cord compression and cord signal change.An emergent open biopsy was performed. Steroids were initiated, and follow-up MRI showed extensive resolution of the lesion. Pathology results were inconclusive. Sections revealed scant fragments of fibroadipose tissue with focal infiltration by a population of medium-sized atypical histiocytic cells. No evidence of malignancy was found. The patient was eventually discharged on methotrexate. Her mother was non-compliant with the treatment, and the patient again returned with thoracic myelopathy and radiographic evidence of thoracic epidural lesions. A second open biopsy was again negative for a neoplastic process.

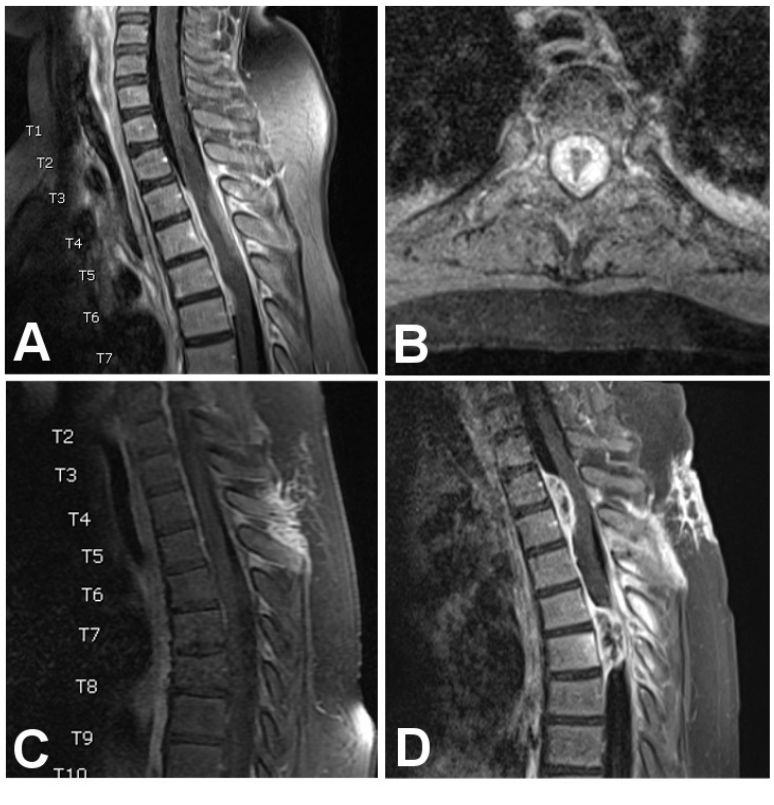
Conclusions

Extradural spinal lesions in the pediatric population presenting with clinical and radiographic features of compressive myelopathy are typically presumed neoplastic until proven otherwise. Infectious processes are quite rare in this region in otherwise healthy children. As demonstrated by our patient, rheumatologic processes such as inflammatory granulomatous disease should be kept in the differential diagnosis. Although our patient did not have systemic sarcoid-like features, her lesional pathology and dramatic response to corticosteroids point us in the direction of an isolated extradural case of neurosarcoidosis.

Learning Objectives

By the conclusion of this session, participants should be able to 1) Come up with a broad differential diagnosis for a spinal epidural lesion in a pediatric patient 2) Understand the extensive workup to rule out a neoplastic process in context of an epidural spinal mass

T1 MRI with contrast



(A and B) Sagital and Axial cuts revealing a circumferetial epidural lesion centered around T3-T6; (C) Sagital section revealing resolution of the epidural lesion after administration of steroids; (D) Sagital section displaying recurrence of the epidural lesion after non-compliance with medical management

Key References

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