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Technical note

Primary spinal intradural extramedullary lymphoma: A novel management strategy

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ABSTRACT

Primary spinal intradural extramedullary lymphoma remains a very rare entity in spinal oncology. In this case report, we present the first treatment of a PSIEL diagnosed by cytopathologic analysis alone followed by urgent radio- and chemotherapy in the literature. At 18-month follow-up, our patient was ambulatory with near total imaging resolution of the lesion. In conclusion, surgical excision or biopsy may not be necessary when suspicion for PSIEL exists, and may delay prompt medical and radiation treatment due to necessity for wound healing. Further research into the management of extramedullary lymphoma treatment strategies is warranted.

1. Introduction

Metastatic spinal tumors remain the most common neoplastic lesion that involves the spine with majority of these being extradural in location [1]. There has been a tremendous advancement in the management of these tumors with an ever increasing role of radiation therapy particularly radiosurgery that can often obviate the role of surgery or allow lesser morbid operations [2]. On the other hand, while intradural tumors accounts for a far lesser number of spinal neoplasms; surgery remains the treatment of choice for intradural tumors that can be either intradural extramedullary (IDEM) or intradural intramedullary [3]. Spinal meningioma and nerve sheath tumors (schwannoma, neurofibroma) remains the most common pathological diagnosis for IDEM tumors [4]. We encountered an unusual case of primary spinal intradural extramedullary lymphoma (PSIEL) causing spinal cord compression that was treated with radiation therapy alone with an excellent outcome following a diagnosis obtained by lumbar puncture. There is no prior report of successful treatment of an IDEM lymphoma in the literature. Given the highly radiosensitive nature of lymphoma, the clinical significance of this cannot be overemphasized.

2. Case example

A 72-year-old immunocompromised female with past medical history of myelodysplastic syndrome with chronic leukopenia was admitted with a 2-month history of bilateral lower extremity weakness. Motor strength was 2/5 hip flexion bilaterally, 2/5 knee extension bilaterally, 4/5 right ankle strength and 3/5 left ankle strength.

Magnetic resonance imaging (MRI) with contrast showed a well-defined dural-based extramedullary lesion from T8-T12 surrounding the spinal cord. The lesion was iso- to hypo-intense on T1 and T2 weighted images with avid enhancement following administration of contrast. (Figs. 1 and 2). Considering the dural-based nature of the lesion and prior history of myelodysplasia, the possibility of intradural metastases or a hematogenous malignancy was entertained. Cerebrospinal fluid was obtained via lumbar puncture for cytopathological analysis (CSF) as an initial work up. Hematoxylin and eosin staining revealed atypical lymphocytes with prominent nuclei in large numbers (Fig. 3). Surface markers were consistent with a lymphocytic plasmocytic neoplasm: CD19+, CD20+, CD38+, and CD45+. Fluorescence *in situ* hybridization (FISH) for MLL, BCL1, BCL2, AML-m4, Burkitt's, AML1, BCR-ABL, MDS, CLL or MM were normal. Serum IgG, IgM, and IgA were within normal limits. Bone marrow aspirate revealed hypercellularity consistent with patient's known history of myelodysplasia and no plasma cell population was observed. Computed tomography (CT) and positron emission tomography (PET) studies for systemic work up were negative. These results were consistent with the diagnosis of primary spinal intradural

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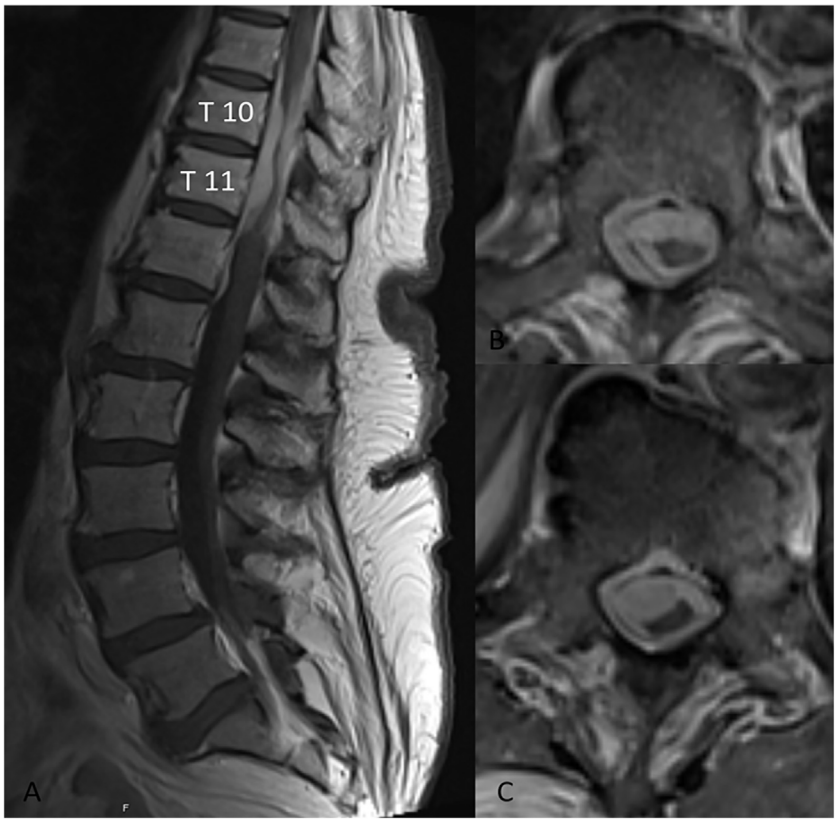

Fig. 1. MRI thoracic and lumbar spine. T2 weighted imaging showing severe cord compression from an intradural lesion with obliteration of all CSF high intensity signal on (A) sagittal and (B and C) axial imaging at T10 and T11.

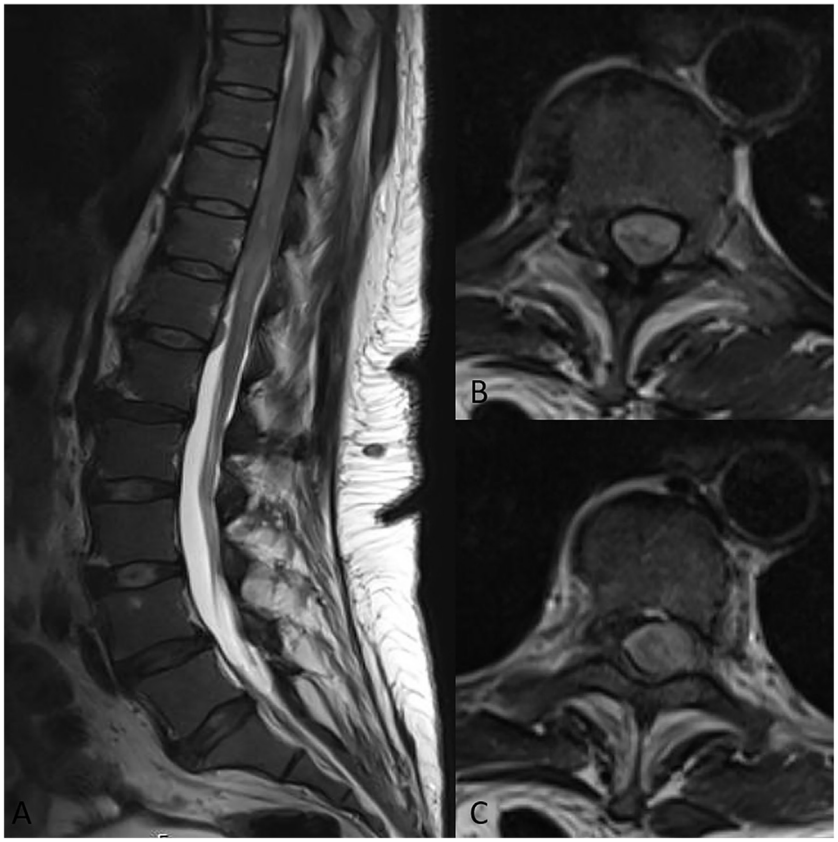

Fig. 2. T1 weighted MR imaging with gadolinium contrast. (A) Sagittal and (B and C) axial imaging reveal a homogenously enhancing intradural extramedullary lesion causing severe cord compression at T10 and T11.