

Primary Spinal Epidural Lymphoma As a Cause of Spontaneous Spinal Anterior Syndrome: A Case Report and Literature Review

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Abstract

Background Primary spinal epidural lymphoma (PSEL) is one of the rarest categories of tumors. Spinal cord compression is an uncommon primary manifestation and requires to be treated with surgery for the purpose of diagnosis and decompression.

Case Presentation A 45-year-old man presented with a new onset thoracic pain and progress to an anterior spinal syndrome with hypoesthesia and loss of thermalgesia. Magnetic resonance image showed a paravertebral mass that produces medullary compression at T3. The patient was taken up to surgery, where the pathology examination showed a diffuse large B-cell lymphoma.

Conclusions PSEL is a pathological entity, which must be considered on a middle-aged man who began with radicular compression, and the treatment of choice is decompression and biopsy. The specific management has not been established yet, but the literature suggests chemotherapy and radiotherapy; however, the outcome is unclear.

Keywords

- ▶ spinal epidural lymphoma
- ▶ epidural
- ▶ spinal compression

Introduction

Primary spinal cord tumors are one of the rarest categories of tumors, representing approximately 4 to 16% of all tumors arising from the central nervous system (CNS).¹ An epidural location for lymphoma is observed in 0.1 to 6.5% of the cases.

We describe the diagnostic and management of a 45-year-old man who presented an anterior spinal syndrome secondary to an epidural lesion, which had a histological confirmation of a diffuse large B-cell lymphoma (DLBCL).

Case Presentation

A 45-year-old man presented with a new onset of high-intensity thoracic pain limiting his movements; a month later he was accompanied by decrease in the strength of

the left pelvic limb. After 2 months, he started with weakness of both lower limbs and impaired urinary sphincter control.

The neurologic examination demonstrated a minor motor impairment of lower limbs and reflexes were hyperactive. He presented a sensory level of hypoesthesia and loss of thermalgesia at T5. Anal sphincter tone was without damage, negative Hoffman, Trömner and Babinski's sign.

Magnetic resonance images showed a paravertebral mass hypointense on T1 and T2 with homogeneous enhancement, which produced medullary compression at T3 (▶ **Figs. 1 and 2**).

The patient was taken into surgery, where we performed a neural decompression by posterior way and biopsy of the extradural spinal lesion. Intraoperatively, we found a fibrotic, gray-colored, vascularized lesion on the right side above the T3–T4 level (▶ **Fig. 3**).

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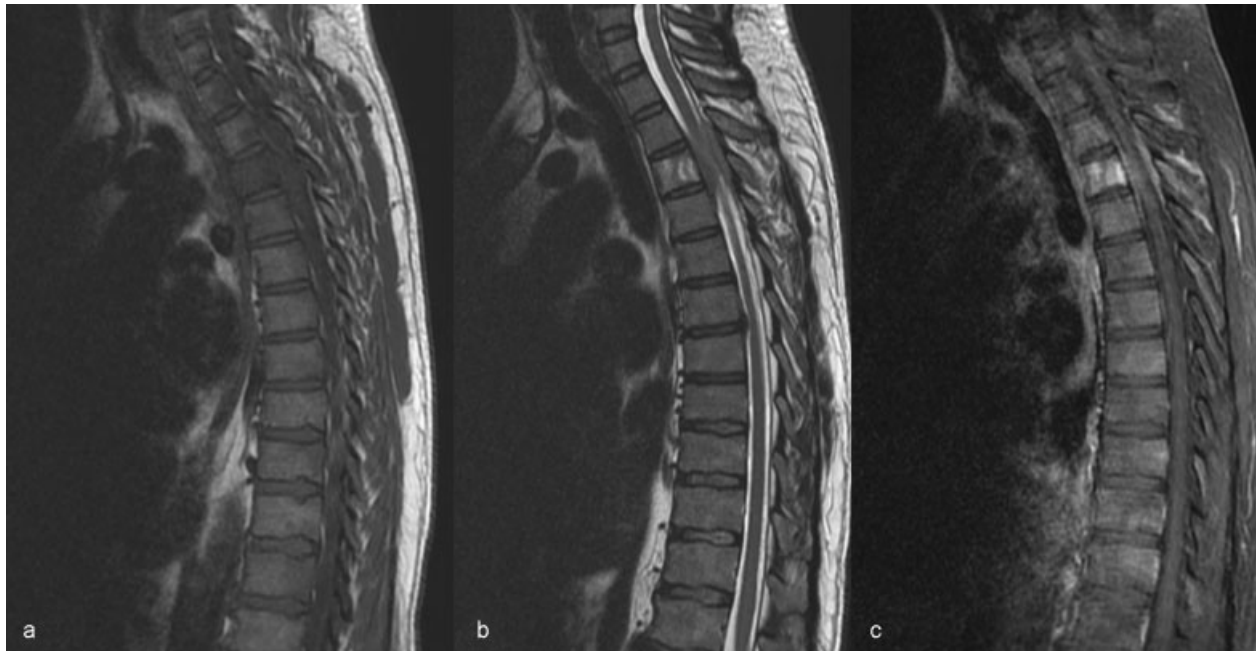


Fig. 1 (a) Sagittal thoracic spine T1-weighted image. (b) Sagittal thoracic spine T2-weighted image. (c) Sagittal thoracic spine contrast material-enhanced T1-weighted image. Vertebral body osteophytes at anterior marginal. Vertebral bodies show multiple irregular areas, hypointense on T1 and hypertense on T2, with heterogeneous enhancement in postcontrast sequence mainly in the vertebral body of T3, where we also observe a paravertebral mass hypointense on T1 and T2, with homogeneous enhancement, which produces medullar compression. Schmorl nodes at the T9–T11 level. Protrusion of the disk at T6–T7 level.

The final pathology examination showed a diffuse large B-cell lymphoma (► **Fig. 4**). Immunohistochemistry reported CD20⁺, BCL-2⁺, CD3⁺ CD5⁺, CD10⁺, CD30⁻, and Ki67 positive in 20% of neoplastic cells (► **Fig. 5**).

During the postoperative course, the patient denied pain, walked again without weakness, and also regained control of the urinary sphincter. The complementary studies were negative to another primary site of lymphoma; the final diagnosis is a primary spine epidural lymphoma (PSEL).

Discussion

Extranodal non-Hodgkin's lymphoma (NHL) accounts for 24 to 48% of all NHL.² It is reported that lymphoma limited to the spinal epidural space participates in up to 3.3% of all lymphomas, 9% of epidural spinal tumors,³ and 0.9% of all extranodal

NHL. Male preponderance of 69% and a male/female ratio of 2:7 were observed.⁴ Patients clinically present most commonly in the fifth to seventh decade of life with more than 80% being older than 40 years.⁵ The midthoracic spine (69%) is the most common site of involvement, followed by the lumbar spine (27%) and the cervical spine (4%).³ According to this review of the literature, our patient fits in all this statistics.

The pathogenesis of PSEL is still unclear, and many hypothesis have been formulated, including the role of chronic inflammatory process, chronic infection, autoimmune disease, and the meningoepithelial component.⁶

DLBCL is the most common category, representing approximately 30% of all NHL. Immunohistochemically, the large lymphoid cells were CD20⁺ and CD3⁻. The DLBCL involving the spinal epidural space could be categorized into germinal center B cell (GCB) type (CD10⁺ o CD10⁻/BCL-6⁺/MUM1⁻) and

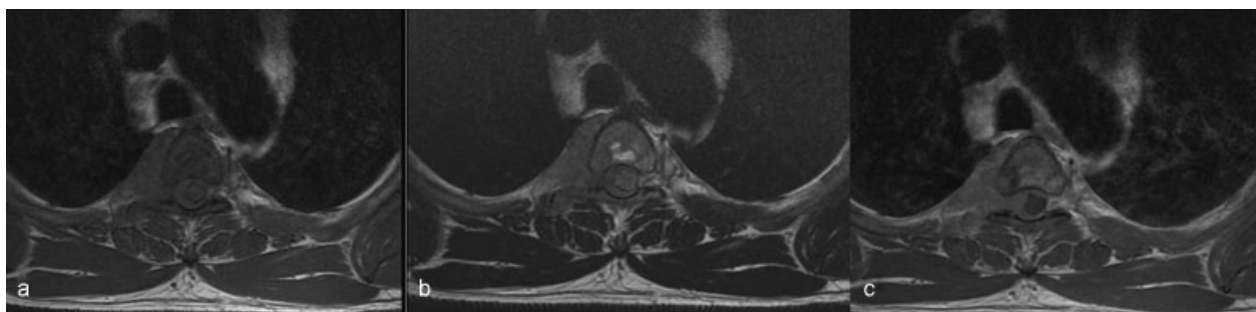


Fig. 2 (a) Axial thoracic spine T1-weighted image. (b) Axial thoracic spine T2-weighted image. (c) Axial thoracic spine contrast material-enhanced T1-weighted image. The vertebral body at T3 level, with hypointense areas in T1 and hypertense in T2, with heterogeneous enhancement in postcontrast sequence. Right epidural mass with homogeneous enhancement; it involved the vertebral body and transverse apophysis that condition radicular compression at the right neural foramen and spinal cord compression

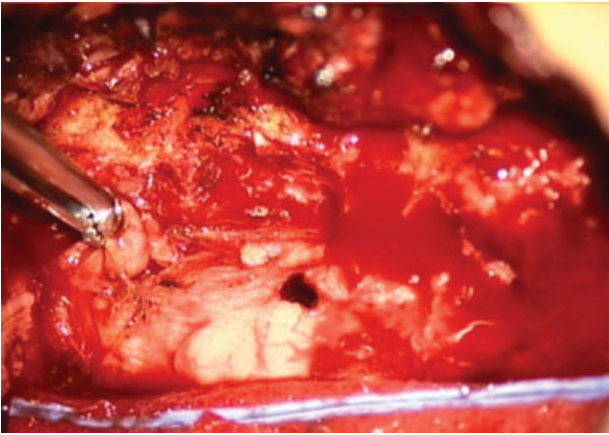


Fig. 3 Surgical view. Fibrotic, gray colored, vascularized lesion; approximately 3 × 1.5 × 1 cm in diameter.

non-GCB (CD10⁻/BCL-6⁻ o CD10⁻/BCL-6⁺/MUM1⁻) type. The ratio of the GCB to the non-GCB subgroup was significantly higher in the spine DLBCL (17:9) than in CNS DLBCL (5:16).⁷

Spinal cord compression is an uncommon primary manifestation of NHL, occurring in less than 5% of newly diagnosed cases,⁸ and may be responsible for mayor morbidity in patients with NHL. Back pain or radicular pain can be the first symptomatic manifestation, followed by neurologic deficits, such as paresis, ataxia, and sensory disturbance. Bladder and bowel disturbance appear only later in the course.⁹ Two phases of clinical presentation have been described: in the first prodromal phase, they suffer from local pain in the back, sometimes accompanied by radicular pain to legs and abdomen, and this pain persisted for several months to 1 year; then the phase of spinal cord compression followed and, within 2 to 8 weeks, the rapidly developing signs of

compression.¹⁰ Based on these clinical manifestations, our case was in the second phase, which can lead us to think that the beginning of the pathology was at least 1 year ago.

The diagnosis requires exclusion of a systemic primary lymphoma and does not apply to patients with immunodeficiency conditions.¹¹ Myelography and computed tomography, as well as magnetic resonance imaging (MRI) are useful for detecting epidural compression. MRI appearance was isointense on T1-weighted images and iso- to hyperintense on T2-weighted images, with marked contrast enhancement; occasionally, MRI demonstrates an extraforaminal component.⁵

The proper management of patients with primary epidural NHL is not yet certain,² and several studies suggest that patients with features of spinal cord compression require to be treated with surgery for the purpose of both tissue diagnosis and decompression.³ Generally, acute paresis and/or loss of bladder/bowel control require emergency decompression of the spinal cord. The surgical approach depends mainly on the location of the tumor, the degree of spinal cord compression, spinal instability, and the patient's general condition.⁹

The researchers suggest that the combination therapy of surgical and radiotherapy and/or chemotherapy should be done.³ Radiation therapy doses of at least 25 Gy and chemotherapeutic agents such as cyclophosphamide, vincristine, and prednisone have been recommended.⁵

The prognosis for functional recovery in patients with spinal cord compression due to epidural NHL is relatively better than that of patients with metastatic carcinoma.² It is reported in many series that nonambulant patients with spinal cord compression have only a 10 to 30% chance of becoming ambulant after therapy; however, earlier diagnosis and treatment are associated with improved functional outcome.⁸

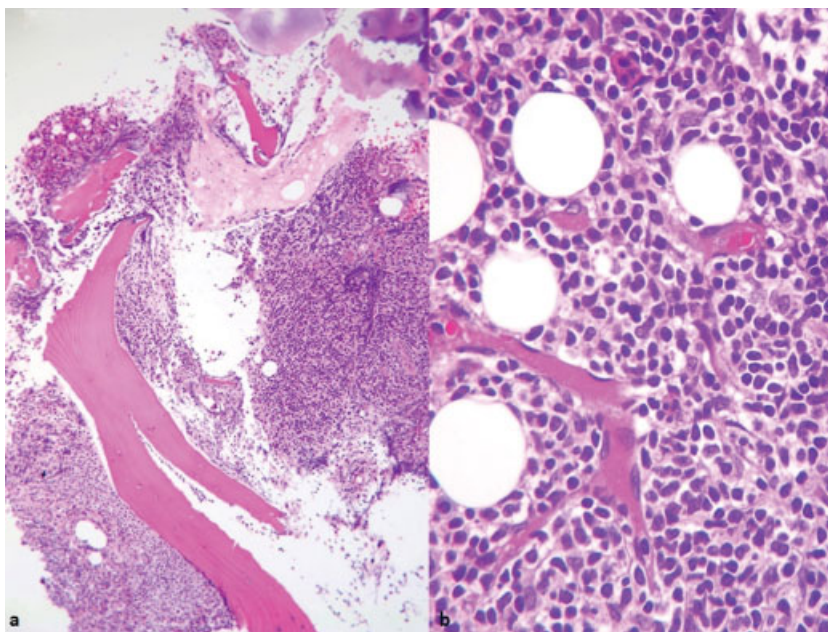


Fig. 4 Pathology examination. (a) Histologically between bone trabeculae. (b) Proliferation of atypical lymphoid cells with hyperchromatic nuclei and scant amount of cytoplasm is observed.

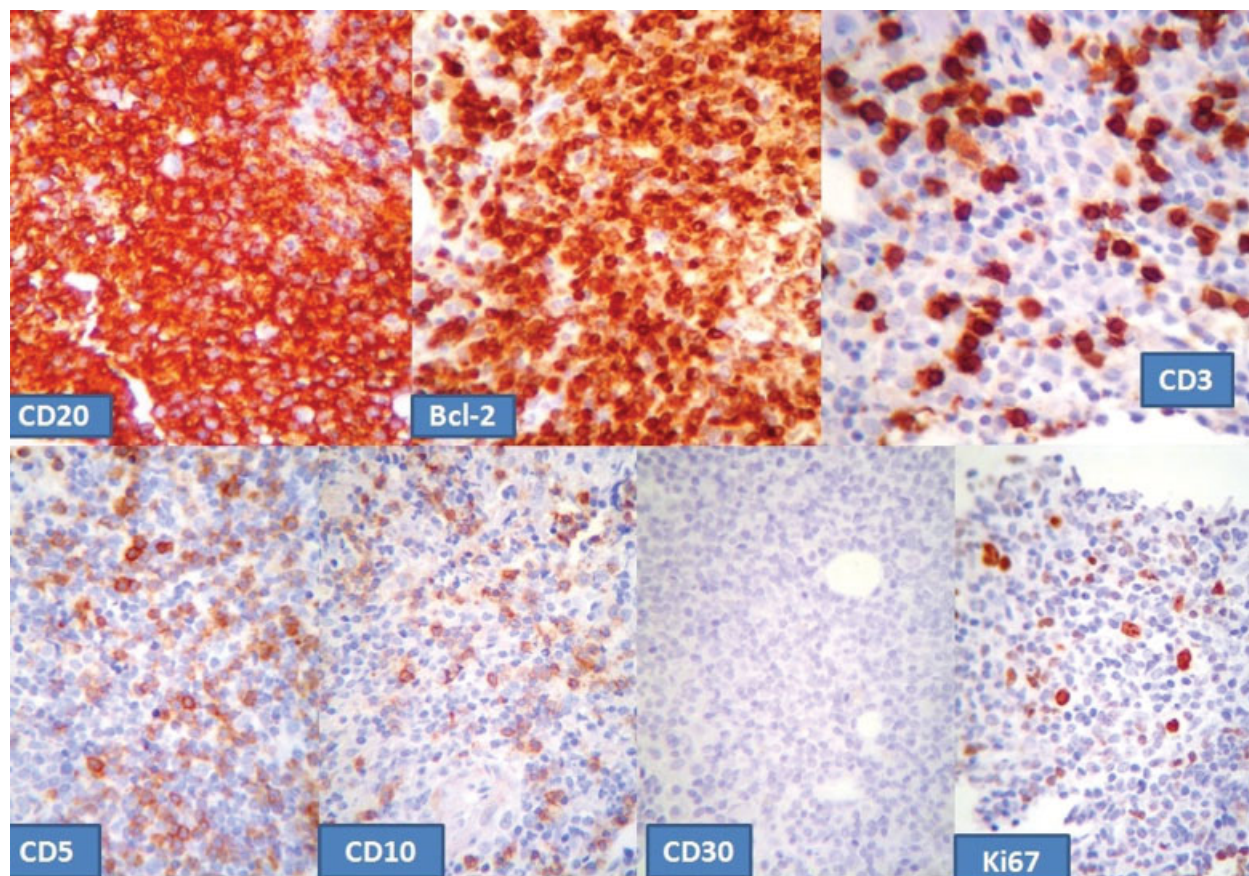


Fig. 5 Neoplastic cells were strongly positive for CD20 and BCL-2, focally positive for CD5 and CD10, and negative for CD30; CD3 was positive in small lymphocytes reactive appearance, and the cell proliferation marker Ki67 positivity was found in 20% of neoplastic cells.

Our patient presents an excellent evolution, and he has just begin with his chemotherapy and radiotherapy. According to the literature, the prognosis should be to recover motor and sensitive functionality, as our patient presented, but it still reserved about life.

Conclusion

Primary spinal epidural lymphoma is a rare pathological entity, which must be considered on a middle-aged man who began with radicular compression.

The use of spinal decompression with biopsy is what promotes better outcome in patients with spinal compression secondary to PSEL; however, subsequent handling is still debated, as being a disease with such low prevalence, the chemotherapy or radiation schemes have not yet been fully established.

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