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Case Report

Primary Spinal Epidural Lymphoma: A Rare but Treatable Emergency

Karima Sif Nasr*, Abir El Hamzi, Mohammed Abdellaoui, Meriem Edderai, Rachida Saouab, Jamal El Fenni and Hassan En Nouali

Radiology Department, Mohamed V Military Hospital, Rabat, Morocco

***Corresponding Author:** Karima Sif Nasr, Radiology Department, Mohamed V Military Hospital, Rabat, Morocco.

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Abstract

Primary spinal epidural lymphoma is a very rare condition. In cases where spinal cord compression occurs, surgical decompression is required, followed by chemotherapy.

A 46-year-old man presented with upper back pain accompanied by progressive sensory disturbances and weakness in the upper limb, particularly affecting the hand.

Magnetic resonance imaging revealed an extradural, extramedullary lesion is identified at the level of T1, occupying the posterior epidural space with left foraminal extension. The patient underwent surgical decompression with tumor resection. Histopathological analysis identified the lesion as diffuse large B-cell lymphoma of the germinal center type.

Chemotherapy was subsequently initiated by the hematology team, and the patient was referred to physical therapy due to mild paraparesis. Postoperative positron emission tomography showed no evidence of residual disease.

Clinicians should consider primary spinal epidural lymphoma in patients presenting with back and leg pain. Early diagnosis and appropriate treatment are essential, as the prognosis is generally favorable when managed promptly.

Keywords: *Thoracic Spine; Primary Spinal Epidural Lymphoma; Foraminal Extension; Chemotherapy*

Introduction

Primary spinal epidural lymphomas are uncommon, accounting for approximately 0.1% - 6.5% of all lymphoma cases and about 9% of spinal epidural tumors. They are defined as lymphomas confined to the epidural space without evidence of systemic disease at the time of diagnosis. These tumors are believed to originate from paraspinal lymphoid tissue, with secondary extension into the spinal canal.

The thoracic spine is the most frequently affected region, with a predominance in male patients. Clinical manifestations typically include back pain, weakness of the lower limbs, sensory disturbances, and, in some cases, dysfunction of urinary or anal sphincter control.

In this report, we describe the case of a 46-year-old man who presented with upper limb sensory disturbances and mild hand weakness due to a spinal extradural lesion at the T1 level, which was subsequently diagnosed as germinal center-type diffuse large B-cell lymphoma.

Case Report

A 46-year-old man was admitted to the emergency department of the Military Hospital of Rabat with upper thoracic pain associated with sensory disturbances in the upper limb. He reported progressive onset of numbness and paresthesia along the medial aspect of the arm and forearm.

Neurological examination revealed mild weakness involving the intrinsic muscles of the hand, with hypoesthesia in the T1 dermatome distribution. Deep tendon reflexes were normal, and no sphincter dysfunction was noted. No motor deficit of the lower limbs was observed.

A magnetic resonance imaging (MRI) was ordered as the patient was suspected to have thoracic spinal involvement due to a primary spinal tumor or metastasis. MRI revealed an intraspinal extradural lesion at the T1 level, occupying and obliterating the posterior epidural space with left foraminal extension. The lesion was isointense on both T1-weighted and T2-weighted imaging and showed homogenous enhancement after gadolinium administration. It exerted a significant mass effect on the spinal cord, which was displaced anteriorly. Associated involvement of the T1 vertebral body, its spinous process, and adjacent soft tissues was also noted (Figure 1-4).

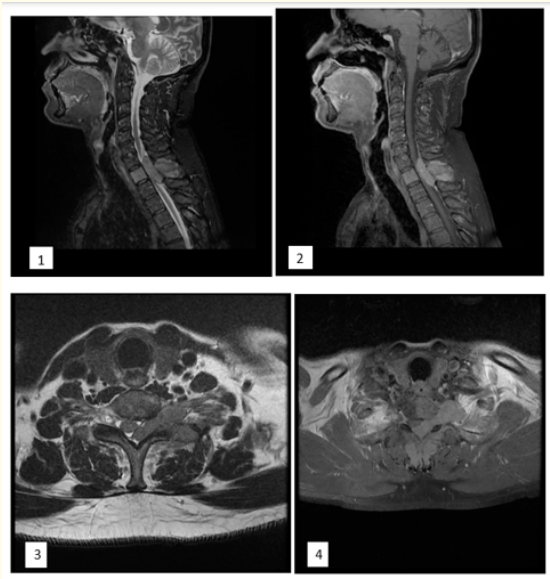

Figure 1-4

The patient was admitted to the neurosurgery department, and surgical intervention was planned to obtain a diagnostic biopsy as well as to achieve urgent spinal cord decompression. The patient subsequently underwent surgery, and decompression was performed via a T1 laminectomy with tumor resection with careful handling to avoid traction on the spinal cord.

A firm, grey-pink mass with associated bone involvement was excised and sent for histopathological analysis, which revealed diffuse large B-cell lymphoma.

Immunohistochemical analysis showed that the neoplastic cells were positive for CD20, with a high proliferative index demonstrated by Ki-67 positivity in nearly all large tumor cells. These findings supported the diagnosis of B-cell lymphoma with a germinal center phenotype.

The patient was subsequently investigated to rule out other possible sites of lymphoma involvement.

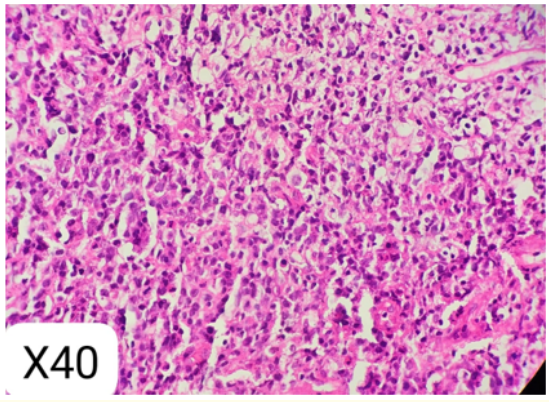

Figure 5: *An infiltration of atypical lymphoid cells is observed. The infiltrating cells are irregularly shaped or lobulated lymphoid cells with clear cytoplasm and large nuclei. Lymphocytes are also present in areas distant from the trabeculae.*

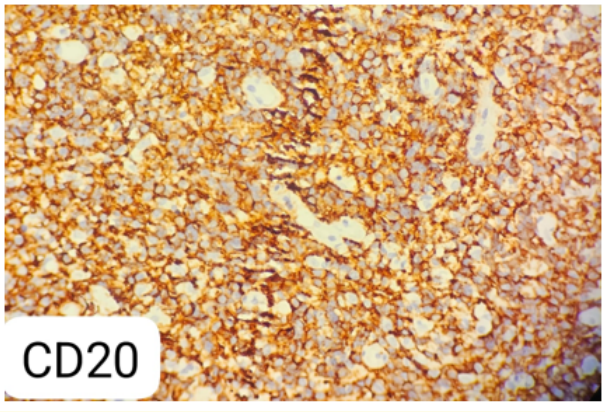

Figure 6: *Lymphoid infiltration shows strong and diffuse staining with CD20 immunohistochemically.*

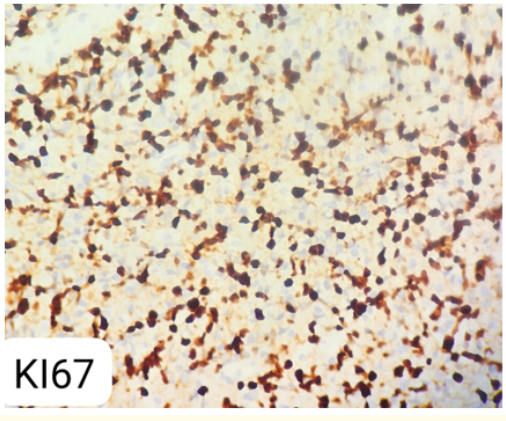

Figure 7: *Ki-67 immunostaining demonstrating a high proliferative index, with positivity in more than 70% of malignant lymphoid cells.*

No other primary site of lymphoma was identified on whole-body bone scintigraphy, single-photon emission computed tomography (SPECT/CT), or positron emission tomography (PET). The final diagnosis was primary spinal epidural lymphoma.

The patient was evaluated by the hematology department, and a bone marrow biopsy was performed. Pathological examination revealed mild small B-cell proliferation with rare CD20-positive large B-cells, and a normocellular bone marrow that was suboptimal for full evaluation. The definitive diagnosis was B-cell lymphoma. The patient completed six cycles of chemotherapy. High-dose methotrexate was administered for central nervous system (CNS) prophylaxis. Post-treatment PET-CT demonstrated a complete metabolic response.

Discussion and Conclusion

Extranodal non-Hodgkin lymphoma (NHL) accounts for approximately 24% - 48% of all NHL cases. Spinal epidural lymphomas represent about 9% of all spinal epidural tumors and 0.9% of all extranodal NHLs [5]. Primary NHL of the spinal epidural region is rare, with epidural involvement itself comprising around 9% of spinal epidural tumors [5,6]. Subdural and intramedullary localizations are even less frequently reported [7].

This disease shows a male predominance (approximately 7:2), and typically presents between the fourth and seventh decades of life. The mid-thoracic spine is most commonly affected (69%), followed by the lumbar (27%) and cervical regions (4%) [2,5,8,9]. The main differential diagnoses include metastatic disease, primary bone tumors, multiple myeloma, and infectious processes [4].

Primary spinal epidural non-Hodgkin lymphoma (NHL) is diagnosed only after excluding systemic disease and other potential pathological conditions. In our case, all other etiological possibilities were ruled out, and the definitive diagnosis was established following surgical intervention and histopathological examination.

MRI is a valuable imaging modality for detecting spinal cord compression and for surgical planning. However, systemic primary lymphoma must first be excluded. Typically, lesions appear hypo- to isointense on T1-weighted images and iso- to hyperintense on T2-weighted images, although T2 signal may occasionally be low due to variable tumor cellularity [2,4]. In our case, the lesion appeared isointense on T1-weighted images and iso-hyperintense on T2-weighted images, with homogeneous enhancement, consistent with previously reported imaging characteristics in the literature.

Since non-Hodgkin lymphoma (NHL) is highly sensitive to both radiotherapy and chemotherapy, surgical intervention is generally reserved for patients with localized disease to obtain a definitive diagnosis and guide subsequent treatment. Surgical decompression combined with biopsy allows for maximal tumor debulking and is therefore considered the preferred approach in cases of spinal cord compression.

Rapid progression of motor deficits, particularly when associated with loss of urinary or anal sphincter control, necessitates emergency decompression. In cases of spinal instability, surgical stabilization may also be required [5,10]. This strategy enables histopathological confirmation in patients without a prior diagnosis related to the epidural lesion and facilitates timely planning of chemotherapy and radiotherapy when indicated.

Primary spinal epidural lymphoma should be considered in patients presenting with spinal cord compression caused by an extradural soft-tissue lesion, with or without associated bone involvement. The prognosis and functional recovery are generally more favorable in cases of spinal cord compression due to non-Hodgkin lymphoma. Therefore, early recognition and timely diagnosis are essential to prevent irreversible neurological deficits, including paraplegia.

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