Primary Cervical Intramedullary Lymphoma: A Case Report

Sameh ACHOURA, Mohamed AMINE RACHDI, Rahma YAICHE, Mohamed Déhmani YEDAES, Hichem AMMAR, Ridha CHIKILI

University Tunis El Manar, Tunisia

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ABSTRACT

Primary non-Hodgkin's lymphomas of the central nervous system represent 1.5% of all neuraxial tumors. Intramedullary localizations are very rare.

We report the case of primary intramedullary lymphoma. The patient was a 46-year-old man with no previous history of lymphoma. He was admitted to our department with walking difficulties, heaviness in all four limbs, spinal claudication, and vesico-sphincter problems such as urinary leakage, which had been present for 6 months.

The examination revealed spastic tetra paresis, a quadri-pyramidal syndrome, and cervical spinal syndrome. A spinal cord MRI showed an encapsulated intramedullary tumor extending from C4 to C6.

The patient underwent surgical excision, and the pathological examination concluded in favor of a small cell lymphoma type B. The patient was subsequently treated with chemotherapy and adjuvant radio therapy with good progression.

Keywords: lymphoma; intramedullary; cervical; surgery; histopathology.

INTRODUCTION

Spinal lymphomas, whether primary or secondary, have long been an unexplained disease. They account for approximately 1-2% of extra nodal lymphomas and often arise as epidural or intramedullary lesions [1,2].

The diagnosis of primary spinal lymphoma is made if no other sites are affected at the time of diagnosis. Secondary spinal lymphomas, on the other hand, arise from direct hematogenous invasion or venous involvement via lymphatic diffusion or from droplet metastases with brain involvement [3].

When these tumors arise as intramedullary lesions, they are difficult to diagnose because they resemble primary spinal tumors and inflammatory lesions [5].

Based on our case and the results of a systematic literature review, we summarize the clinical, radiological, and pathological features associated with spinal lymphoma and present our approach to the treatment.

CASE PRESENTATION:

A 46-year-old male, with no past medical history, presented with intermittent fever and chills and a progressive history of right-sided weakness over the past eight months.

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He did not complain of back pain, loss of appetite, or weight loss. On multiple occasions during the course of these past months, steroids and pain treatment had partially relieved his weakness. He had no prior history of malignancy or family history.

He was a known smoker, 1 pack/day for the past twenty years. A lumbar puncture was done, and CSF was negative for infections. He was treated with high dose corticosteroids and discharged after his symptoms improved.

He then presented one month later, with worsening right-sided weakness, walking difficulties, heaviness in all four limbs, right C5 cervico-brachial neuralgia, spinal claudication and episodes of urinary leakage.

The patient was immediately admitted to our neurosurgical department.

Physical examination revealed a spastic tetraparesis, a motor deficit in all 4 limbs, a quadri-pyramidal, and a spinal cord syndrome.

The rest of the physical examination was within normal limits. He was restarted on corticosteroids, which considerably decreased his weakness over the course of the following days.

MRI of the brain done at the time showed no abnormality but MRI of the cervical spinal cord showed an encapsulated intracanal tissue mass extended from C4 to C6 with a low signal intensity on a T2 weighted image, associated to a T1 iso signal with homogeneous and intense enhancement. **Fig. 1**

After discussion with the patient and his family, we performed a bilateral laminectomy from vertebrae C3 to C7, the dura was opened along the midline, a midline myelotomy was completed and exposed a reddish, hemorrhagic and brittle, intramedullary tumor, attached to nerve roots.

In this case only subtotal resection was possible because of the invasive nature of the mass.

Pathological examination supplemented by an immunohistochemical investigation concluded to a small B cell CD 5 lymphoma, of non-specific phenotype. **Fig. 2**

Based on the diagnosis and the incomplete resection leaving residual tumor tissue, the patient was started on chemotherapy with adjuvant radiotherapy on the cervical region.

After six months, the patient remained clinically stable without any new neurological deficits. Follow-up evaluations were conducted every three months during the first year and every six months thereafter, including medical history, physical examinations, and MRI with gadolinium contrast.

At 6 months, brain and cervical MRIs showed a decrease in the lesion’s extent and no evidence of spinal cord expansion.

DISCUSSION

Primary intramedullary lymphoma (PIML)) occurs in middle-aged and older adults and mimics other causes of myelopathy. Delay to diagnosis is common due to its rarity, its similarity to other causes of myelopathy, and the difficulties in obtaining pathologic diagnosis [13].

CSF cytology has traditionally been diagnostic in only a small percentage of cases [4]. However, CSF is rarely normal, with 75% of patients exhibiting elevated protein levels and 50% showing mild pleocytosis in primary central nervous system lymphoma (PCNSL) [7] cases. Cellular immunophenotyping and repeated

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CSF examinations can enhance diagnostic accuracy [6]. Nevertheless, due to the high rate of false negatives and the diagnostic delays caused by multiple lumbar punctures, it is advisable to consider an early biopsy if PCNSL is suspected.

The MRI features of PIML may be indistinguishable from other causes of myelopathy especially early on, but certain characteristics may suggest lymphoma diagnosis; multifocal spinal cord lesions, persistent gadolinium enhancement and conus medullar or cauda equina involvement [4].

In this case, the tumor was localized on the cervical segment and was a monofocal lesion.

The role of spinal cord biopsy in diagnosing lymphoma is unclear. There is limited literature on the safety of spinal cord biopsies, and it is crucial to consider the high risk of irreversible spinal cord damage associated with the procedure. In our summary of the literature, which includes our case, only 4 out of 10 cases required a biopsy for a definitive diagnosis. Therefore, biopsies should be limited to patients with inconclusive imaging and laboratory findings [8].

Subjective improvement following initial treatment was common, but significant functional gains were rare. Methotrexate was the primary initial therapy, yet relapses typically occurred within 3 months. Most patients required additional treatments, such as R-CHOP, radiation, or peripheral blood stem cell transplants. Drawing conclusions about the relative efficacy of different treatment recommendations is challenging due to the small number of patients and the variability in treatments [9].

The role of radiotherapy in treating spinal cord lymphomas is not well-defined. Radiation can be employed for cases resistant to chemotherapy and serves as local therapy for palliative purposes in patients unable to tolerate high-dose chemotherapy. However, craniospinal irradiation carries significant risks of neurological and bone marrow toxicity [15].

The role of surgery in treating spinal cord lymphomas seems less critical compared to systemic chemotherapy. In a case reported by Guzik et al. [11], surgical resection of the spinal cord lymphoma was performed, but within four weeks, the tumor had grown larger than before diagnosis, necessitating chemotherapy with methotrexate and cytarabine to achieve remission. Consolidation was then completed with radiotherapy. In another case reported by Rao et al. [12], complete surgical resection was done, but the patient did not experience clinical improvement and passed away without further treatment. In both instances, the tumors were relatively large, and surgery was undertaken primarily for tissue diagnosis, with the possibility of spinal cord lymphoma not being anticipated prior to the procedure [14].

CONCLUSION

Lymphoma involving the spinal cord is a rare and challenging condition to diagnose. It should be considered in the differential diagnosis for patients who present with rapidly progressing motor and sensory deficits along with spinal cord lesions on imaging, even if there is no evidence of systemic lymphoma. Improvement in symptoms and tumor shrinkage following steroid treatment should raise suspicion of lymphoma. It is essential to confirm the diagnosis of lymphoma before proceeding with a spinal cord biopsy. Aggressive systemic chemo therapy is vital for survival.

CONFLICTS OF INTEREST

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

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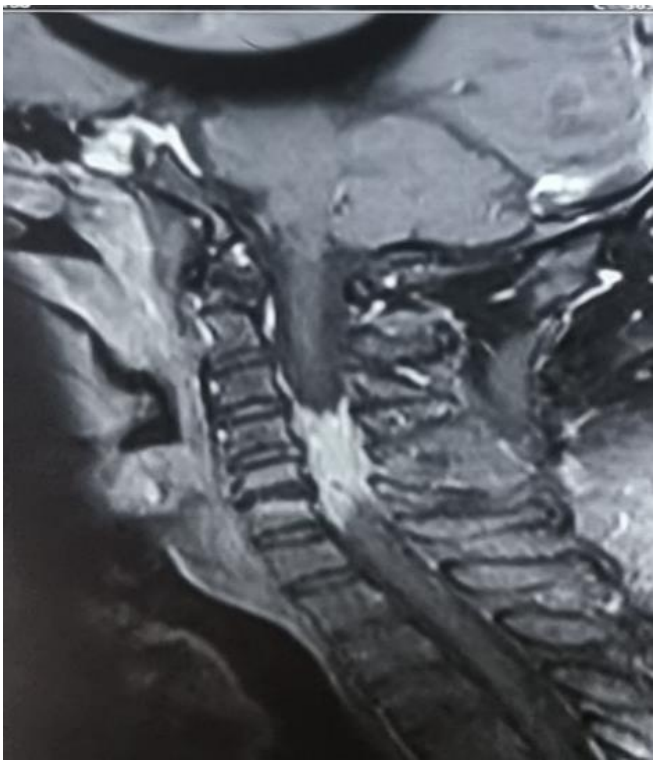
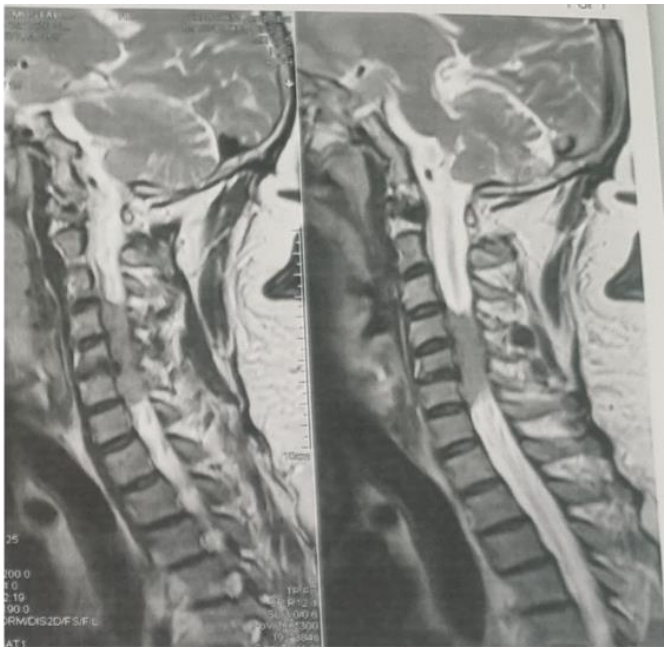
APPENDIX

Figure: 1



T1



T2

MRI of cervical region on sagittal section emphisized the extension of this tumoral mass from C4 to C6 With a low signal intensity on T2 weighted image associated to a T1 iso signal with homogeneous and intense enhancement

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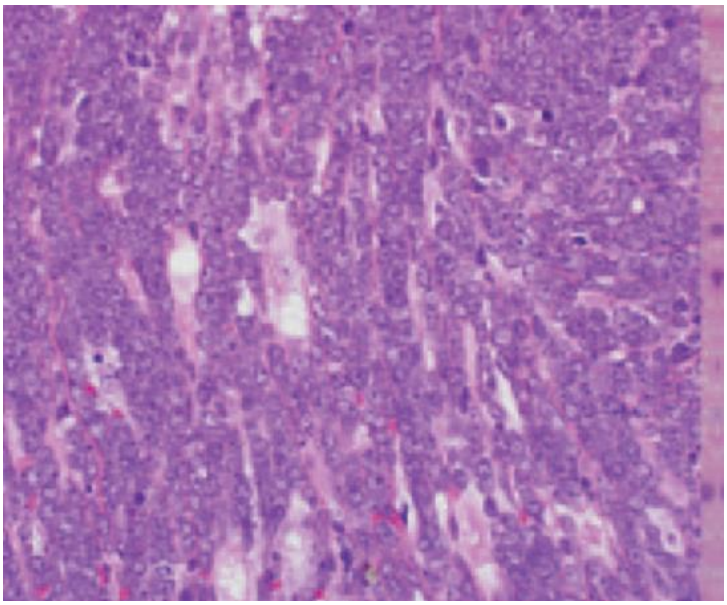
Figure 2



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