Rare presentation of a spinal lymphoma: case report and diagnosis by magnetic resonance imaging

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ABSTRACT

A primary central nervous system lymphoma is a form of extranodal disease originating in the brain, leptomeninges, spinal cord, or eyes. Spinal tumors are low-prevalence neoplasms and can cause considerable neurological morbidity and mortality. An isolated lymphoma emerging within the dural canal is the rarest form of primary central nervous system lymphoma: it accounts for approximately 1% of cases occurring more often in the context of secondary dissemination than as the primary site of origin. Symptoms are nonspecific and depend on the spinal level involved. The presentation is insidious and includes dorsalgia, weakness, and progressive difficulty in ambulatory function. MRI is the modality of choice to search for lesions within the spinal/rachial canal in patients presenting with neurological symptoms. Surgical treatment is not helpful, and the main objective of surgery is to know the histological diagnosis.

Key words: primary central nervous system lymphoma, intramedullary, magnetic resonance imaging, case report.

INTRODUCTION

Among the various oncologic pathologies, spinal tumors are low-prevalence neoplasms but can cause considerable neurological morbidity and mortality. Historically, we can distinguish three main groups of spinal tumors: extradural, intradural-extramedullary, and intramedullary[1].

Primary central nervous system (CNS) lymphoma is a form of extranodal disease originating in the brain, leptomeninges, eyes, or spinal cord.

We describe the case of a patient with primary intradural intramedullary lymphoma due to the infrequent presentation of this disease and highlight the role of diagnostic imaging in decision-making regarding management and follow-up.

CLINICAL CASE

Male patient, 83 years old, with no relevant history and immunocompetent, who consulted for progressive weakness in the left leg and low back pain of 4 weeks of evolution, exacerbated during the practice of a sport. On neurological examination, he presented bilateral crural paresis 4/5 and diminished osteotendinous reflexes in both legs, with preserved sensitivity. Laboratory tests were within normal limits.

We indicated analgesic treatment and requested, in the first instance, an X-ray which showed signs of spondyloarthrosis without evidence of acute pathology, and a magnetic resonance imaging (MRI) of the lumbosacral spine on an outpatient basis.

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MRI shows in intradural topography a focal lesion with an isointense-hypointense signal on T2 regarding the neural tissue covering segments L2-L3. It spares the bony structures and, after the injection of gadolinium, shows marked intradural homogeneous enhancement with leptomeningeal contact and enhancement of the nerve roots of the cauda equina at that level (Fig. 1).

The patient persists with progressive lumbago and paraparesis, so he is readmitted to the Emergency Department 5 days after the initial consultation, with no changes in his neurological examination. He comes in for poor pain management and to complete the study of the spinal lesion. Laboratory tests report mild thrombocytopenia with no other noteworthy findings. We requested an MRI of the brain and cervicodorsal spine to complete the evaluation of the neuroaxis but found no associated lesions to the known lesion at the lumbar level. We mention an incidental finding of a left parietal meningioma (Fig. 2).

The positron emission tomography (PET/CT) with F18-FDG (fluorodeoxyglucose) subsequently performed showed increased metabolism of the known intradural lesion with SUV (Standardized uptake value) of 28.8, with no other particularities in FDG distribution.

Because of these findings, we decided to operate. We performed a bilateral laminectomy exposing thickened nerve roots at the L3-L4 level, with pathological appearance (Fig. 3). Those roots that did not show neurophysiological response were sectioned and sent for histopathological analysis. The surgical specimen culminated in 4 fragments of whitish tissue, the largest of which was 1.2 cm.

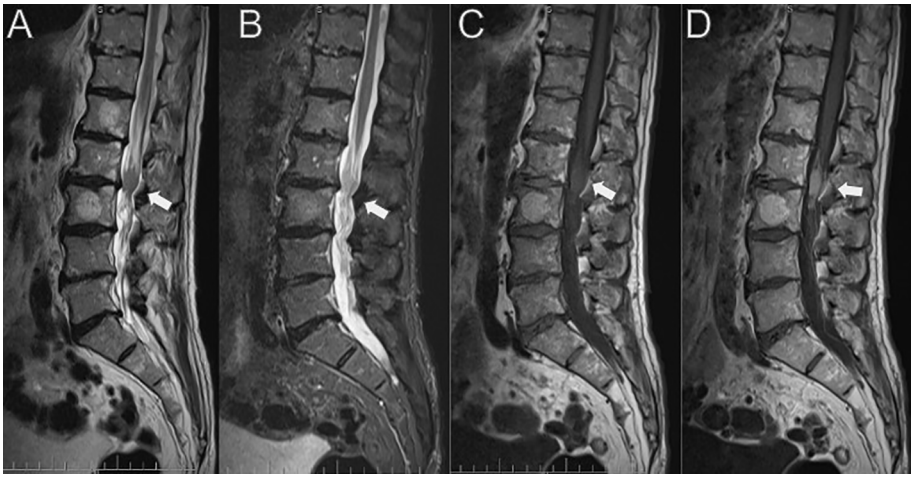

Figure 1. MRI of lumbar spine. Sagittal T2 (**A**), STIR (**B**), T1 without contrast (**C**), and T1 postcontrast (**D**). Intradural lesion observed at L2-L3 level (arrows), hypointense in T2 and T2 STIR, isointense in T1 regarding the neural tissue. The lumbar osseous duct diameters are within normal limits. After gadolinium administration, it shows marked enhancement with involvement of the roots of the cauda equina above and below the level of the lesion.

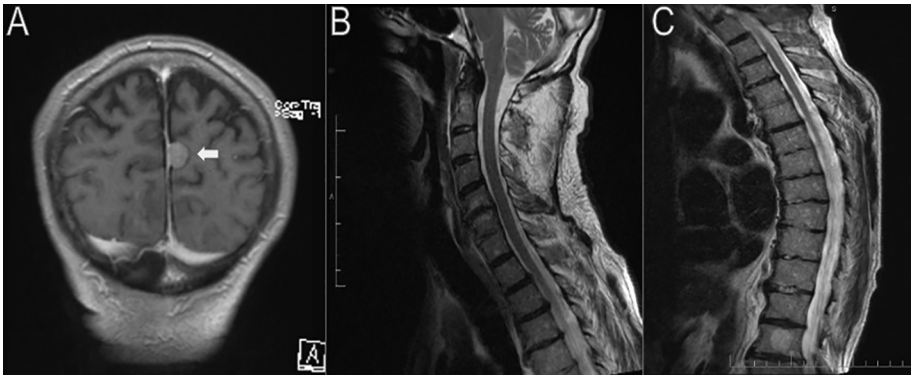

Figure 2. MRI of the brain and cervicodorsal spine. T2-weighted images of the rest of the neuroaxis, (**A**) coronal brain, (**B**) sagittal cervical spine, (**C**) sagittal dorsal spine, where there are no other findings related to the lesion under study; it only shows an image compatible with meningioma (arrow) in the left medial parietal region in close contact with the brain sickle.

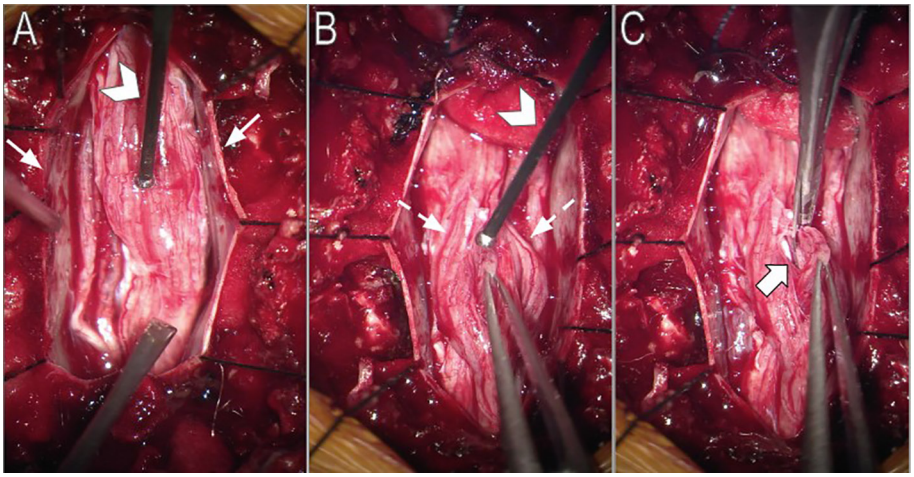

Figure 3. Surgical specimen. Access to the intramedullary cavity at the level of the cauda equina. **A**) Linear opening of the dura mater and its anchorage (white arrows). **B**) Dissection of the arachnoid (dashed arrows). **C**) Access to the intradural space (thick arrow). Physiological monitoring assesses the response to the motor roots in each segment (arrowhead).

The histopathological analysis shows the spinal neural fragment with infiltration by lymphoid proliferation constituted by enlarged cells, while immunohistochemistry was positive for CD20, BCL6, and MUM1 and negative for CD3, BCL2, CD10, and Cmyc. Our final diagnosis was infiltration by large B-cell lymphoma, non-centrogerminal phenotype.

DISCUSSION

Primary intradural lymphomas are extremely rare, comprising only 1-2% of patients with primary CNS lymphoma. They, in turn, constitute only 1% of all lymphomas in the body, with a slight male preponderance. As with other CNS lymphomas, the predominant known risk factor is immunosuppression. The average age of presentation in immunocompetent patients tends to be 62.5 years, whereas immunocompromised patients tend to be younger. About 90% of spinal lymphomas arise from B-cells, while the remainder are T-cell lymphomas[1].

Primary spinal involvement by lymphoma usually compromises, in order of frequency: the vertebral body, the epidural space, and the intradural space, either extramedullary or intramedullary, as the least affected sites[2]. Primary lymphoma of the bone, including the spine, is rare and accounts for less than 5% of extranodal lymphomas and less than 1% of non-Hodgkin's lymphomas. A primary spinal epidural lymphoma diagnosis requires isolated, nondisseminated disease and comprises less than 1% of extranodal lymphomas. Isolated lymphoma within the dural canal is the least common form of primary CNS lymphoma: it accounts for approximately 1% of cases occurring more often in the context of secondary dissemination than it does as the primary source site[3].

This lymphoma often arises in the upper thoracic or lower cervical regions along the spinal cord, with the lumbar region being the least frequently affected; symptoms are nonspecific and depend on the spinal level involved. The presentation is insidious and includes dorsalgia, weakness, and progressive difficulty in ambulation[4].

MRI is the modality of choice in the search for lesions within the spinal/rachial canal in patients presenting with neurological symptoms. If the vertebral bodies are involved, CT is an adequate method to assess the extent and pattern of bone involvement[5]. MRI is the most accurate method when the lesion involves the spinal canal. Lymphoma within the duct usually ranges from isointense to hypointense on T1-weighted images and hyperintense on T2-weighted images. There may be a surrounding hyperintense signal in association with vasogenic edema. The vast majority of spinal cord neoplasms show avid enhancement after contrast administration. The areas with enhancement probably represent more active portions of the tumors and may indicate potential sites for biopsy if resection is not feasible[6]. DWI sequence diffusion and the apparent diffusion coefficient (ADC) map reflect the macromolecular movement of extracellular water and are very useful in distinguishing lymphoma from other tumors. The high cellularity of the lymphoma decreases the extracellular space and restricts the normal random movement of water molecules. That is what sets it apart from most intradural lesions. It translates as hyperintensity on diffusion imaging and hypointensity

on ADC mapping. These sequences are also helpful for assessing response to treatment[2].

FDG-PET aims at determining other pathological focuses to rule out the presence of systemic lymphoma (in up to 7% of these patients). Detection of other neoplasms and false positives have also been reported in 5% and 13% of cases, respectively[4].

Treatment of lymphoma begins at diagnosis due to the aggressive nature of the tumor. Chemotherapy based on corticosteroids and methotrexate, with or without radiotherapy, is the first line of treatment[1]. Surgical treatment is not helpful, and the first goal of surgery is to obtain a histological diagnosis. The prognosis is worse for intradural lymphoma than for vertebral or extradural lymphoma, with less than 50% of patients surviving two years after diagnosis[3].

The principal differential diagnoses include ependymomas, astrocytomas, and metastatic disease. Myxopapillary ependymomas of the conus medullaris and filum terminale are relatively common spinal intradural neoplasms, predominantly seen in children and young adults, but can also occur at an older age. There is a slight male predominance. They appear as isointense or slightly hyperintense masses in the spinal cord on T1-weighted images, whereas on T2-weighted images, they appear hyperintense. They are usually extramedullary and present as a cauda equina syndrome. There may be posterior vertebral scalloping as well as intravertebral foraminal widening[1]. Astrocytomas prevalence in adults is second only to that of ependymomas. The mean age of presentation is 29 years, and the most common site of involvement is the thoracic spinal cord. It manifests most frequently with pain and sensory deficits. On MRI, these neoplasms usually have poorly defined margins and are isointense to hypointense relative to the spinal cord on T1-weighted images and hyperintense on T2-weighted images. The average length of involvement is seven vertebral segments, and virtually all astrocytomas show at least some enhancement after contrast material administration[6]. The most common solid tumors associated with spinal metastases are melanoma, breast carcinoma, and small cell lung carcinoma, while leukemia and lymphoma are the most common among hematologic malignancies. The clinical history in these cases is most relevant. Lower motor neuron weakness, dermatomal sensory changes, and bowel and bladder dysfunction are important clinical clues to leptomeningeal disease. Whole spine gadolinium MRI has been considered an accurate imaging test to document leptomeningeal dissemination in cancer patients.

Nodular lesions with enhancement after contrast administration, which usually predominate in cauda equina, strongly suggest a diagnosis in the appropriate clinical context.[6]

The present study was conducted within the guidelines laid down by the modified Helsinki Declaration.

CONCLUSION

Spinal lymphoma is an uncommon entity with various forms of localization. The clinical presentation is usually nonspecific. In our case, it is an extramedullary intradural lesion, one of the least frequent. MRI with gadolinium is the method of choice for evaluating these patients. We highlight the usefulness of postcontrast sequences and diffusion for differential diagnosis with other neoplasms and spinal pathologies. Treatment is limited to chemotherapy, so surgery remains reserved for biopsy for diagnosis. That is why diagnostic imaging is fundamental to alert the surgeon of the possibility of this etiology and thus avoid unnecessary surgical procedures and eventual complications.

Conflicts of interest: the authors declare no conflicts of interest.

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