

Case Report

Primary cauda equina lymphoma with an isolated lesion: a case report and review of literature

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Abstract: We present a case of a 59-year-old man with backache, weakness and numbness of both lower extremities, magnetic resonance imaging (MRI) showed a round isolated intradural extramedullary lesion between L3 and L4 of T1 isointensity, T2 mild hyperintensity, which was markedly enhanced with gadolinium while cauda equina nerve was slightly enhanced. L3 laminectomy was performed and the pathological diagnose confirmed it was a diffuse large B-cell lymphoma. Then he received chemoradiotherapy. This is the second case of primary cauda equina lymphoma with an isolated lesion. In addition to the case report, we review the relative literature in English to describe the clinical features, radiology findings as well as treatment of primary cauda equina lymphoma.

Keywords: Primary cauda equina lymphoma, isolated lesion, surgery, chemoradiotherapy

Introduction

Lymphoma of the spinal cord is uncommon, accounting for 3.3% of all central nervous system (CNS) lymphoma, which constitutes only 1% of all lymphomas in the body [1]. In addition, primary cauda equina lymphoma is extremely rare, without an accurate incidence. There are 18 cases of primary cauda equina lymphoma reported in English literature previously, most of which an isolated lesion cannot be found [2-19]. Here we report a rare case of primary cauda equina lymphoma with an isolated lesion.

Case report

A 59-year-old man previously healthy, with complaint of backache and numbness of both lower extremities for 5 months was admitted. The symptoms occurred as a backache at first, and then numbness arose in both lower extremities, more severe on the left. As it developed, 2 months before admission, he felt weakness of the lower extremities, also left more severe than right.

There was no peripheral lymphadenopathy or hepatosplenomegaly. Neurological examination of the cranial nerves and the upper limbs

was normal. The examination revealed atrophy of left crus, hypoesthesia of both lower extremities, especially the lateral crus and foot on the left, weakness of both extremities with myodynamia of Grade III on the left and Grade IV on the right, deep tendo reflexes were hypoactive on both lower extremities.

Routine hematological and biochemical tests, as well as human immunodeficiency virus serology, were normal.

MRI scan showed an intervertebral disc herniation of L2 and a round isolated intradural extramedullary lesion between L3 and L4 of T1 isointensity, T2 mild hyperintensity, which was markedly enhanced with gadolinium while cauda equina nerve was slightly enhanced (**Figure 1A-C**).

The diagnosis then was made as intradural extramedullary occupation, intervertebral disc herniation. The differential diagnoses included neurilemmoma and ependymoma. To confirm the property of the lesion, we performed a surgery, hoping to excise the isolated lesion and clear the herniated disc at the same time. The thecal sac looked like in high pressure, nerve roots of the cauda equina appeared thickened

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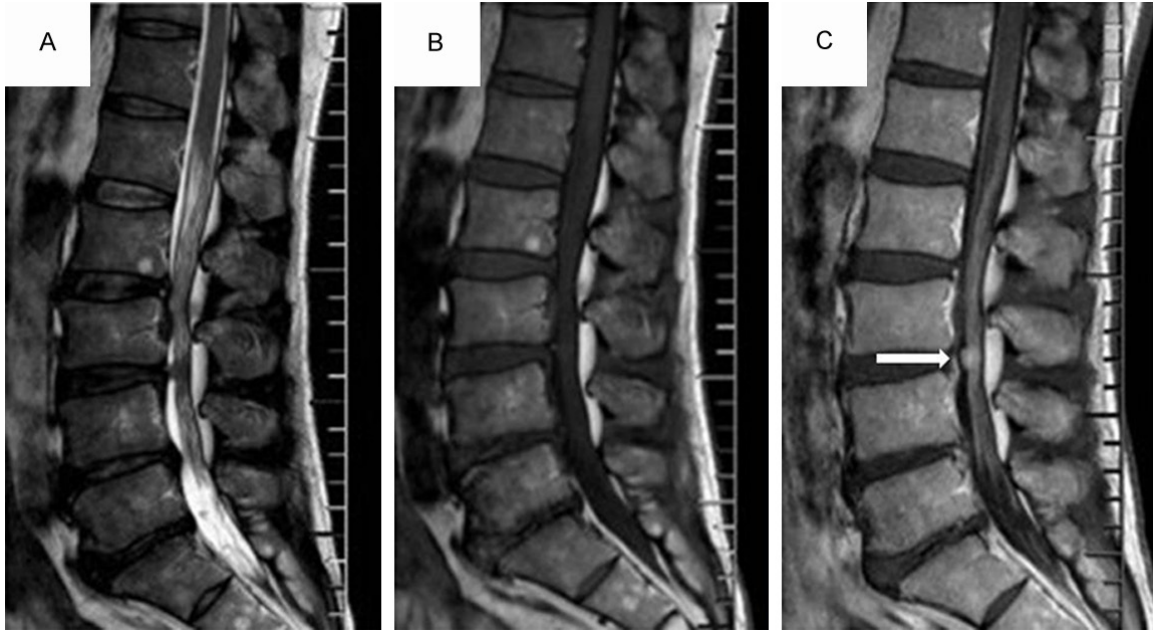


Figure 1. Preoperative Magnetic resonance imaging (MRI) of the patient. A: T2-weighted sagittal image; B: T1-weighted sagittal image; C: Contrast enhanced T1-weighted sagittal image, a round isolated lesion (arrow) was markedly enhanced while cauda equina nerve was slightly enhanced.

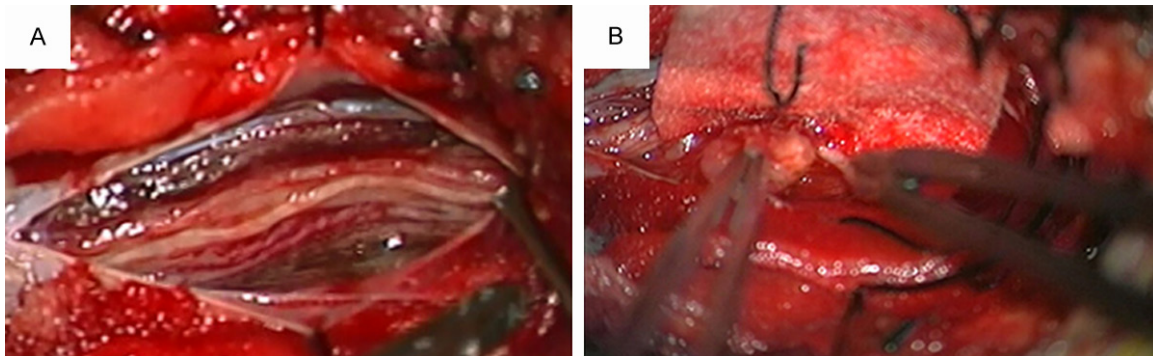


Figure 2. Intraoperative image. A: Nerve roots of the cauda equina appeared thickened and swelling with blood vessels dilated; B: A fishlike and fragile isolated lesion with clear margin attached tightly to a ventral nerve root.

and swelling with blood vessels dilated (**Figure 2A**). A round isolated fishlike tumor with clear margin was found adhered tightly to a ventral nerve root and it was excised completely (**Figure 2B**). Because the cauda equina was so swelling that it was too difficult to clear the disc. After the operation, slight improvement of weakness presented on his left lower extremity.

Histopathology examination revealed it was a diffuse large B-cell lymphoma. Immunohistochemical analysis showed it was positive for CD20 with high proliferative index (ki-67 80%)

(**Figure 3**). Then he received whole-body fludeoxyglucose (FDG)-positron emission tomography. There was no accumulation of FDG elsewhere other than cauda equina. Bone marrow biopsy of the iliac bone did not show abnormalities. Thus he was diagnosed as primary cauda equina lymphoma.

MRI scan after surgery showed no isolated lesion anymore and the cauda equina was slightly enhanced (**Figure 4A-C**). Then he was transferred to hematology department for further treatment with chemotherapy of intravenous methotrexate (MTX) and intrathecal MTX

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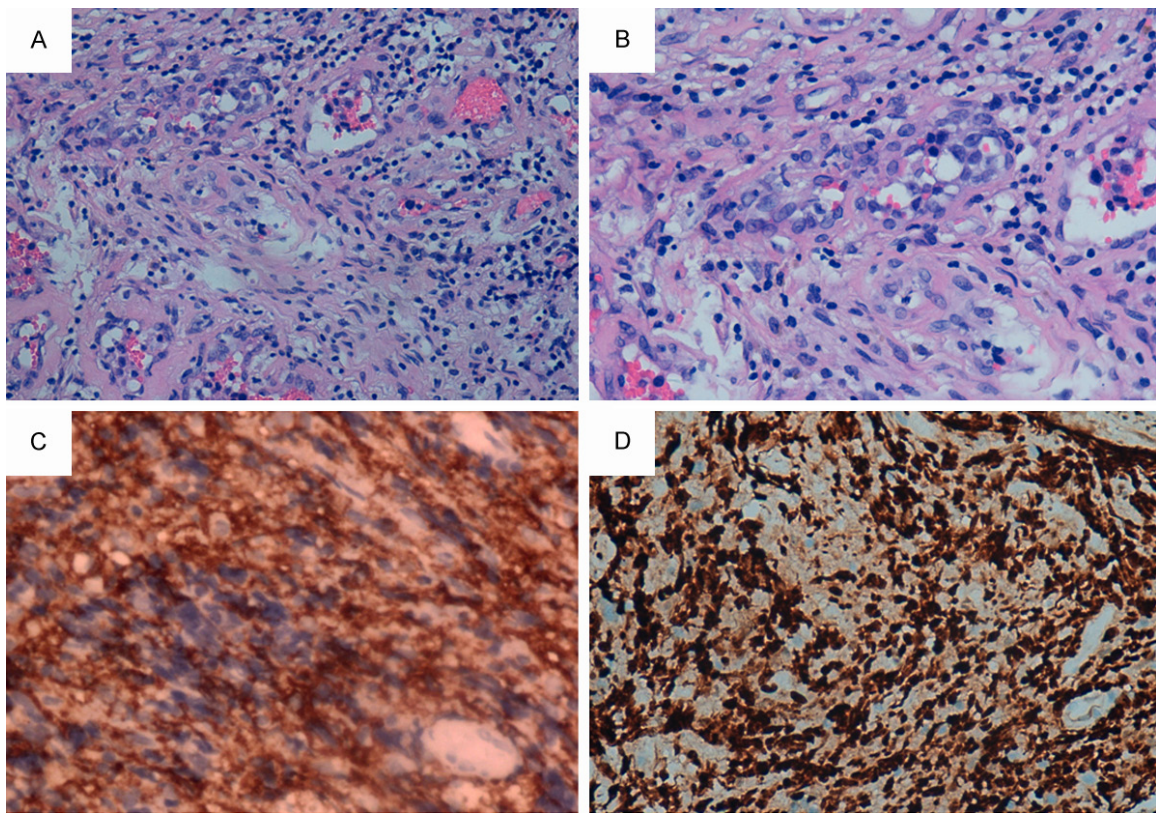


Figure 3. Haematoxylin-Eosin (H&E) staining and immunohistochemical analysis of the lymphoma. A: H&E staining ($\times 200$); B: H&E staining ($\times 400$); C: Immunohistochemistry is positive for CD20; D: Immunohistochemistry shows high proliferative index (ki-67 80%).

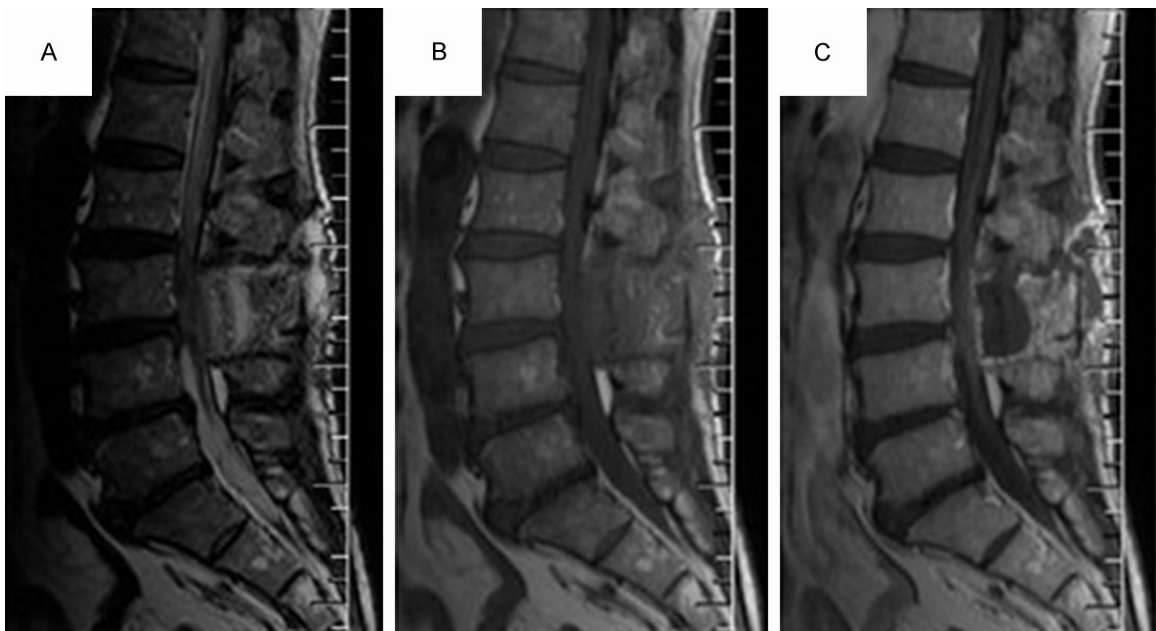


Figure 4. Postoperative MRI of the patient. A: T2-weighted sagittal image; B: T1-weighted sagittal image; C: Contrast enhanced T1-weighted sagittal image, no isolated lesion was found, cauda equine was slightly enhanced.

combined with cytarabine (Ara-C) and lumbar radiotherapy.

12 months after the operation, he can walk with the help of a cane and the numbness of

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both low extremities shows no exacerbation.

Discussion

Lymphoma of the spinal cord is uncommon, accounting for 3.3% of all CNS lymphoma, which constitutes only 1% of all lymphomas in the body [1]. Furthermore, primary cauda equina lymphoma is extremely rare. There are all together 19 cases including ours reported in English literature [2-19]. Only 2 cases have found an isolated lesion [14]. Most primary CNS lymphomas are often associated with immunocompromised patients [19]. However, of all cases of primary cauda equina lymphoma, only 2 (10.5%) were reported with immunodeficiency because of acquired immune deficiency syndrome (AIDS) [4, 13]. Of all the patients, there are 11 males and 8 females, aging from 11 to 71, average 53.1.

As the lymphoma involved cauda equina, of all the 19 cases, 14 (73.7%) experienced numbness, weakness of both lower extremities; 3 (15.8%) had bladder dysfunction as an subacute cauda equina syndrome within 3 months. 5 (26.3%) suffered a history of more than 5 months, the longest for 15 months before being admitted. As a whole, the symptoms progress rapidly.

MRI can be a useful accessory examination [6]. Primary cauda equina lymphoma presents with expanding cauda equina, generally poorly defined, isointense or hypointense on T1 with homogeneous contrast enhancement and hyperintense on T2 [20]. In our case, besides these findings, a round isolated lesion was so apparent that we took it for a neurilemmoma. However, the expanding cauda equina with slight enhancement reminded us it might be a malignant tumor. Cugati reported another case with a well-defined sausage shaped lesion isointense on T1 and T2. But they didn't perform a contrast examination and they took it for an ependymoma according to MRI [14]. Because of the non-specific MRI findings, it's not easy to establish an accurate diagnose according to MRI scan. Maybe the most useful information on MRI should be expanding cauda equina with homogeneous contrast enhancement. And for laboratory test, the serum soluble interleukin-2 receptor (sIL-2R) levels may be helpful [21].

Surgery plays an important role in either nerve root biopsy or excision of the lesion, if there is one, which can finally confirm the pathological diagnose and relieve symptoms. During the operation, we found a fishlike and fragile isolated lesion with clear margin attached tightly to a nerve root, which resembled a neurilemmoma, except for the swelling cauda equina. In all the 19 cases, 12 carried out surgeries. Only in 2 cases an isolated lesion was found, so Teo et al, recommends an intra-operative smear if the cauda equina appears marked thickness and greyish with fine telangiectatic blood vessels to avoid excessive operation [13]. Nishida et al, achieved a definite diagnose with the help of cerebrospinal fluid (CSF) in which they found malignant lymphocytes with CD20 positive [16]. However, it's usually hard to get CSF through lumbar puncture because the swelling cauda equina may cause obstruction and it's even harder to find malignant lymphocytes in CSF [17, 18].

As there are few cases of primary cauda equina lymphoma, the therapy strategy varies in different cases. 15 cases reported have chemotherapy of different regimes. 1 case received high dose of intravenous MTX (3.5 g/m²) and 1 case received high-dose of intravenous MTX (1.0 g/m²) with cytarabine, both patients got complete remission for a survival of 1.5 years [15, 16]. It appears that high-dose MTX can be effective, while combination chemotherapy has good oncologic rational and has proven effective [22]. Intrathecal (IT) chemotherapy increases the concentration of drug in CSF, thus decrease the total dose of drug and minimize systemic side effects. But neurotoxicity shouldn't be ignored. Even the cauda equina syndrome may exacerbate [23]. 13 cases performed radiotherapy, most of which were local radiation, total doses ranging from 30 Gy to 50 Gy, consistent with the dose of 40-50 Gy for primary CNS lymphoma [24]. 12 cases received chemoradiotherapy, they got a mean survival of 16.3 months. The 5-year overall survival (OS) rate of primary CNS lymphoma could be 30-50% [24], and that of primary cauda equine lymphoma needs longer follow-up.

Conclusion

Primary cauda equina lymphoma with an isolated lesion is extremely rare. A differential diagnose of primary cauda equina lymphoma

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should be taken into consideration if MRI scan finds expanding cauda equina with enhancement, which can be a main feature of primary cauda equina lymphoma. Surgery is helpful in either excision or biopsy. The expanding cauda equina might be an indicator for an intraoperative smear to avoid excessive surgery. Early intervention of chemoradiotherapy will be necessary for a better prognosis.

Disclosure of conflict of interest

None.

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