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LETTER TO THE EDITOR

A fatal back pain: report of two cases of diffuse large B cell/Burkitt-like nervous system lymphoma

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Dear Editor-in-Chief,

Primary central nervous system lymphoma (PCNSL) is an uncommon variant of the extranodal non-Hodgkin lymphoma which involves different structures of the nervous system. It accounts for 2.7% of all nervous system malignancies [1]. Most PCNSL are diffuse large B cell lymphomas (DLBCLs). Diffuse large B cell lymphoma/Burkitt-like (DLBCL/BL) is a term assigned to an aggressive B cell lymphoma with features intermediate between the diffuse large B cell lymphoma and the Burkitt-like lymphoma in the updated WHO classification of tumors of the hematopoietic and lymphoid tissues [2]. There are no data on the incidence and prognosis of the nervous system DLBCL/BL since there has been only one report of DLBCL/BL with cerebral and involvement of the cauda equina which suggested DLBCL/BL could be PCNSL with a very aggressive nature [3]. We present a case of PCNSL and a case of primary lymphoma of peripheral nerve (PLPN), both DBCL/BL, with an initial presentation as lower back pain.

Case 1

A 47-year-old, previously healthy woman was admitted to the emergency room (ER) complaining of a dull pain localized in the lumbar region lasting for five days. Neurological examination was normal and the patient was discharged with a recommendation of analgesia and bed rest. In the next seven days, the patient returned two more times complaining of the same symptoms. The neurological examination was normal and a plain X-ray of the lumbar spine followed by a multislice computed tomography (MSCT) of the lumbar spine was also normal. Four days after the last examination, the patient presented to the ER with leg weakness and reported obstipation. Neurological examination revealed moderate to heavy flaccid paraparesis and hypoesthesia of the left groin with urinary retention (1600 mL). Urgent magnetic resonance imaging (MRI) revealed pathological process inside the spinal canal exceeding from Th7 to L4 segment with compression and ventral dislocation of the medulla (Fig. 1). Sub-acute epidural hematoma was diagnosed and the patient underwent an urgent operation. After the surgery, the patient experienced a prompt relief of the pain and the motor strength recovered. Four days later, flaccid paraplegia developed. The control MRI showed tumor mass at the level of L1. Pathological analysis detected a diffuse proliferation of medium- to large-size lymphocytes. Immunohistochemistry showed cells which were strongly positive for CD20, CD10, PAX5, BCL2, and MUM1; negative for BCL6, CD3, CD5, cyclin D1, and CD21; and more than 90% positive for Ki-67 proliferation index. Multiple scattered macrophages were detected with a starry-sky appearance. The DLBCL/BL was diagnosed. Additional findings excluded infiltration of the bone marrow and lymph nodes. Radiotherapy was performed without any regression of the neurological deficit, and chemotherapy regimen (R-DA-EPOCH, rituximab with dose-adjusted etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin) was initiated. The patient died a few days after the fourth cycle of chemotherapy regimen, due to an infection.

Case 2

A 50-year-old, previously healthy woman was admitted to the ER due to a dull back pain and left sciatica followed by left

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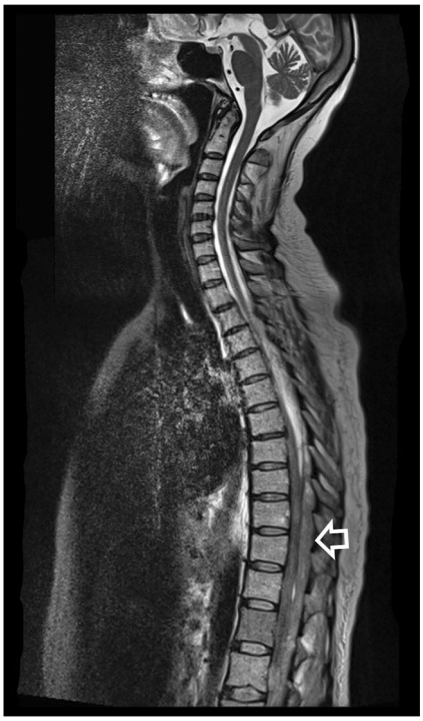

Fig. 1 Sagittal T2-weighted MRI of the cervical and thoracic spine showing process inside the spinal canal causing compression and ventral dislocation of the medulla

foot paralysis. The pain had lasted for twenty days and, a week prior to our consultation, the diagnostic workup for sciatica was performed in another hospital. The MRI of the lumbar spine revealed a mild protrusion of the L4/L5 intervertebral disc without any indication for neurosurgical intervention. Two days prior to our consultation, the patient developed left foot paralysis. Neurological examination showed left foot paralysis, hypoesthesia of the L5 and S1 dermatomes, and positive Lasegue sign at 40 degrees on the left side. MSCT of the lumbar spine revealed swollen left S1 root from which tumor expanded towards to the retroperitoneum and invaded left piriformis muscle (Fig. 2). The patient underwent an urgent surgery during which was detected that a tumor initiated around the left sciatic nerve, expanded to the left piriformis muscle and towards the pelvis. Pathological analysis showed multiple medium- to large-size atypical cells which were highly positive for PAX5, MUM1, BCL2, and CD10; partially positive for BCL6 and CD19; and negative for CD20, CD3, CD5, TdT, cyclin D1, and CD79a with nearly 100% for positive Ki-67 proliferation index. DBCL/BL lymphoma was diagnosed and chemotherapy regimen (R-DA-EPOCH) initiated. No recovery of neurological deficit was observed. The patient died after the third cycle of chemotherapy regimen, due to an infection.

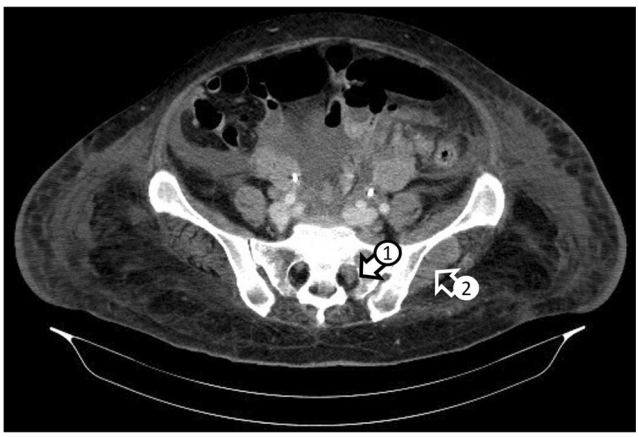

Fig. 2 MSCT of the lumbar spine showing swollen left S1 root (arrow 1) and tumor invading left piriformis muscle (arrow 2)

Discussion

We presented two patients, middle aged, previously healthy women, with nervous system DLBCL/BL initially presenting as usually benign neurological condition—lower back pain. The rapid progression of symptoms and neurological deficit in the first case mislead towards the diagnosis of a spinal hematoma. As the DBCL/BL was diagnosed, the size of the spinal tumor mass and the absence of typical B symptoms, which are not usually seen in patients with extra nodal localizations of lymphomas, indicate that this was the PCNSL [4]. The aggressive nature of DBCL/BL was observed in our second patient in whom MRI of the lumbar spine performed two weeks prior to our consultation had not revealed any pathological process and the MSCT of the lumbar spine performed in our ER revealed a huge tumor expanding from the left S1 root. The failure of the detachment of the tumor from the sciatic nerve together with the absence of B symptoms is highly indicative that the primary site of the tumor was also the nervous system, but in this case the peripheral nervous system. PLPNs are extremely rare. The majority of reported patients presented as mononeuropathy, with most frequently involved sciatic nerve. Almost all cases were found to be of B cell lineage, with different clinical outcomes [5]. Our second patient presents the first report of DBCL/BL as PLPN, with very aggressive nature.

Conclusion

In our patients, DLBCL/BL lymphoma had an aggressive nature, as it was previously observed [4]. Fortunately, this entity is a very rare nervous system malignancy. However, in certain cases when medical history and nature of signs and symptoms are not typical for intervertebral disc disorder, despite normal initial radiological findings, an underlying malignancy should be suspected.

Compliance with ethical standards

Informed consent Informed consent was obtained from both patients included in this paper.

Conflict of interest The authors declare that they have no conflict of interest.

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