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Case Report

Primary diffuse large B-cell lymphoma of dura: two cases report and brief review of the literature

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Abstract: Primary diffuse large B-cell lymphoma (PDLBL) of dura is a subtype of central nervous system lymphoma and is rare in clinical practice. The MRI features of PDLBL are still lacking, and this tumor is easily misdiagnosed as other cerebral disorders before operation, such as meningioma. Therefore, the accurate diagnosis of PDLBL is important before treatment. In this study, we reported 2 cases of PDLBL. The 2 cases had similar MR findings including the presence of vasogenic edema, hemorrhage, destruction of the adjacent skull, the antenna-shaped prominence in the inner margin and parenchymal brain invasion with a fuzzy tumor brain interface. These MRI features may help us to make a diagnosis of dural PDLBL.

Keywords: Lymphoma, central nervous system, magnetic resonance imaging

Introduction

Primary central nervous system (CNS) lymphomas is an extranodal non-Hodgkin lymphoma that involves the brain, leptomeninges, intraocular structures, or spine cord in the absence of systemic disease. Primary leptomeningeal lymphoma (PLML) originates from the meninges without any brain involvement. Primary dural lymphoma (PDL) is described as dura matter involvement, a reported subentity of PLML, and comprises 0.6% to 3% of all brain tumors [1, 2].

Primary diffuse large B-cell lymphoma (PDLBL) presenting as PDL is extremely rare. The PDL is usually a low-grade marginal zone lymphoma (MZL), whereas other types of PCNSLs are usually high-grade, diffuse, large B-cell lymphomas. PDLBL of the dura is easily misdiagnosed as other meningeal tumor before operation. It is very valuable and important to position it and qualitatively diagnose it before operation. In this study, we reported 2 cases of dural PDLBL with clinical data and MR imaging findings, and we reviewed the relevant literature.

Case report

Case 1

A 71-year-old woman presented to our hospital with left limb weakness for 20 days. Her symptom was aggravated progressively and accompanied with headache. She had no previous medical or surgical history. No significant physical examination was found except for 3/5 motoricity in the left upper and lower limbs. No abnormal blood tests were found. An extra-axial spindle mass was found at the right temporoparietal region in the MR examination before surgery. The lesion located near the endocranium. The size of lesion was 3.2 cm (lateral) × 7.2 cm (anteroposterior) × 8.3 cm (vertical), with well defined boundary. It appeared heterogeneous isointense on T1-weighted image (**Figure 1A**) and slightly heterogeneous hyperintense on T2-weighted image (**Figure 1B**), and was obviously homogeneously enhanced with dural tail sign after administration of gadolinium (**Figure 1C** and **1D**). Nodular hematoma was found in the rim of the mass. There was bone destruction in the adjacent skull.

The patient underwent a surgical excision. During the surgery, it was found that the tumor was firm, dural-based, involving the adjacent skull. There was no clear boundary between the tumor and the normal brain. The tumor had rich vascular supply. The final histopathological diagnosis was diffuse large B-cell lymphoma, with positive CD20 (+++) and CD79a (+++).

Case 2: A 44-year-old woman presented to our hospital with a frontal mass which was acciden-

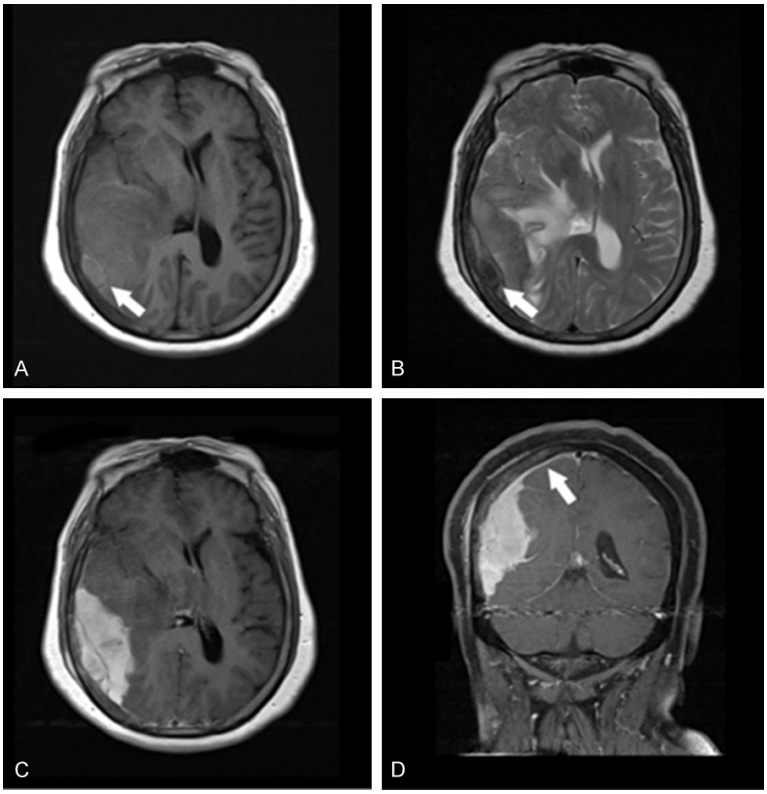

Figure 1. An extra-axial mass appeared heterogeneous isointense on T1-weighted image (A) with a nodule with hyperintense, suggestion of hematoma, could be seen in the mass (white arrow), and heterogeneous slightly hyperintense on T2-weighted image (B) in the temporal region, with surrounding vasogenic edema. Postcontrast T1-weighted image (C and D) showed the mass was homogeneous enhanced with clear dural tail (white arrow) and antenna-shaped inner margin (black arrow). The destruction of adjacent skull was seen.

tally found 5 days ago. The patient had no limbs powerless or activity obstacle. Preoperative MRI examination revealed a well-defined mass, locating at the right frontal-parietal, with wide dural base. The size of lesion was 3.1 cm (lateral) × 4.3 cm (anteroposterior) × 2.9 cm (vertical). Homogeneous isointense and mild hyperintense were found in brain parenchyma on T1-and T2-weighted sequences, respectively (**Figure 2A** and **2B**). The lesion enhanced homogeneously with a classic dural tail, and extended into left sulci like an "antenna" (**Figure 2C** and **2D**). The destruction of adjacent skull was seen (**Figure 2D**).

The patient underwent a surgical excision. The tumor was a dural-based soft and grey tissue, involving the adjacent skull. The postoperative pathological diagnosis was diffuse large B-cell lymphoma, with positive CD20 (+++) and CD79a (+++).

Discussion

PCNSL is an uncommon variant of extranodal NHL that involves the brain, leptomeninges, eyes, or spinal cord without evidence of systemic disease [1, 2]. PCNSL accounts for 1% of non-Hodgkin's lymphomas (NHL) and is indistinguishable from NHLs that occur at other body sites [3].

The PDL is usually a low-grade marginal zone lymphoma (MZL) and lesser is diffuse large B-cell lymphoma histologically. The two types are indistinguishable from the radiological imaging. The dural PDLBL expresses B-cell-associated antigens such as CD20 and CD79a. Primary CNS lymphoma occurs more often in males, but PDL has a female predilection [4]. The 2 cases we reported were both female. Primary CNS lymphomas has been reported to be related with immunosuppression, while PDL has no correlation with immunocompromised conditions [5]. The symptoms of the PDL are variable and non-specific, usually depending on the location of the tumor. The most common clinical presentations are headaches, seizures, focal sensory or motor deficits, and visual disturbances [4]. The first case of our study showed the deficits of motor function, but the second case in our study had no specific symptom.

The pathogenesis of PDL remains unclear because lymphoid tissue is absent in the dura. The following hypotheses may explain the pathogenesis of PDL [6]. Firstly, dural-based lymphoma may result from meningeal seeding from an undiagnosed systemic lymphoma. Secondly, meningothelial cells are embryologically analogous to epithelial cells at other sites in which lymphomas arise, and these cells can be found within the arachnoid membrane and dural venous sinuses. Thirdly, inflammatory con-

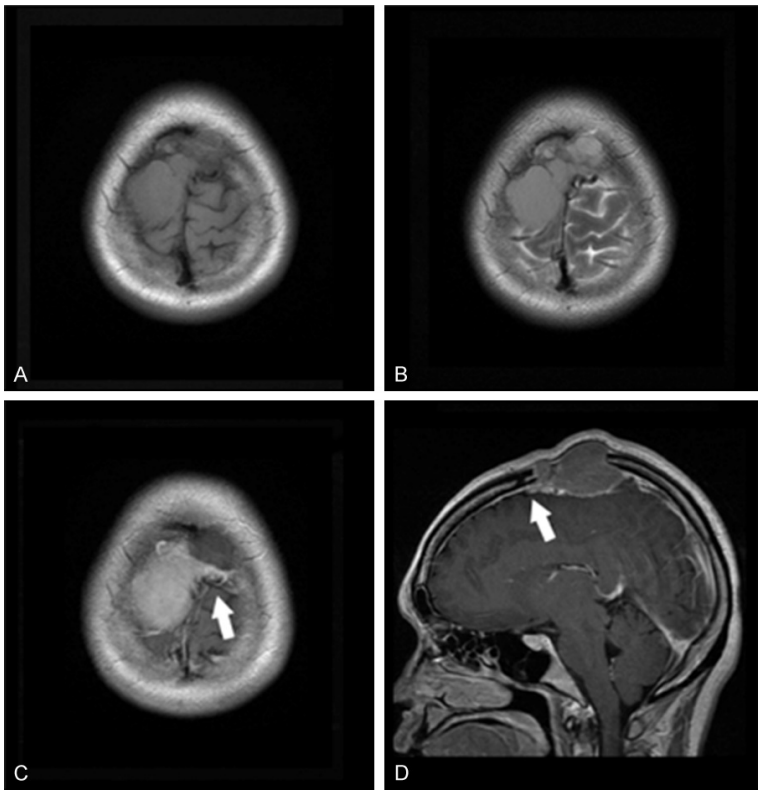

Figure 2. An extra-axial lesion appeared homogeneous isointense on T1-weighted image (A) and slightly hyperintense on T2-weighted image (B) in the frontal region. Postcontrast T1-weighted image (C and D) showed intense homogeneous enhancement of the lesion, with antenna-shaped inner margin (white arrow) and dural tail sign (black arrow). The adjacent skull was damaged (D).

ditions involving the dura could attract polyclonal lymphocytes from which lymphoma could arise.

MRI is sensitive in detecting the dural-based extra-axial lesions. The features of PDL on MRI examination are isointensity-slight hypointensity on T1-weighted image, isointensity-slight hyperintensity on T2 weighted image, and diffusely enhanced with a 'dural tail' sign after administration of gadolinium contrast. The tumor could have hemorrhage and remarkable vasogenic edema. The presence of adjacent skull invasion and antenna-shaped prominence in the inner margin of lesion with a fuzzy tumor-brain interface is valuable MRI findings.

The main differential diagnosis is meningioma [7]. PDL and meningioma share many similar clinical and radiographic and features, including female predilection, age of onset and MRI findings. Both tumors could appear isointensity-slight hypointensity on T1 weighted image and isointensity-slight hyperintensity on T2 weighted image. MRI often demonstrates extra-axial lesions with a 'dural tail' sign in both tumors [8]. However, the degree of enhancement in PDL was less than that in meningioma after the administration of gadolinium. The presence of vasogenic edema and parenchymal brain invasion with a fuzzy tumor-brain interface may favor a diagnosis of PDL [9]. One of our cases showed a specific hemorrhage in the tumor, but the hemorrhage was rare in meningioma. The different changes in adjacent skull are important clue to differentiate PDL and meningioma. Destruction of adjacent skull is usually seen in PDL, while meningioma is typically associated with hyperostosis of adjacent skull.

In conclusion, we reported 2 cases of primary malignant B-cell dural lymphoma, a very rare subtype of primary CNS lymphoma. Although the MR findings of PDLs are similar to meningioma's, the presence of vasogenic edema, hemorrhage, destruction of the adjacent skull, the antenna-shaped prominence in the inner margin and parenchymal brain invasion with a fuzzy tumor brain interface favors a diagnosis of PDL.

Disclosure of conflict of interest

None.

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