Clinical Case Report

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Head trauma complicated with primary cranial vault lymphoma

A case report

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Abstract

Rationale: Primary cranial vault lymphoma (PCVL) is an extremely rare extranodal lymphoma in the skull. This case study investigates the clinical features, so as to improve the understanding of the diagnosis and therapy.

Patient concerns: A 31-year-old male presented painful scalp mass at the site of 1-month-old head trauma.

Diagnosis: The final diagnosis was plasma cell lymphoma, which is a rare subtype of diffuse large B-cell lymphoma based on biopsy and immunohistochemistry findings.

Interventions: The patient received total tumor resection in combination with chemotherapy

Outcomes: The patient survived without signs of systemic dissemination for 12 months after surgery at the time of last follow-up.

Lessons: Trauma may be one of the factors that induce PCVL. The final diagnosis of PCVL depends on pathology and immunohistochemistry findings. A combined treatment of surgery, chemotherapy, and radiotherapy can achieve favorable outcomes.

Abbreviations: NHL = non-Hodgkin lymphoma, PCVL = primary cranial vault lymphoma, PLB = primary lymphoma of the bone.

Keywords: magnetic resonance imaging, primary cranial vault lymphoma, trauma

1. Introduction

Primary lymphoma of the bone (PLB) is a rare type of tumor, accounting for 3% to 5% of extrahepatic lymphoma. About 75% of PLB occurs in the pelvis and limbs.[1,2] PLB in the skull, that is, primary cranial vault lymphoma (PCVL), is particularly rare.[3,4] Some researchers have suggested that trauma may be related to PCVL, but the relationship between head trauma and PCVL remains controversial.[5–8]

To the best of our knowledge, only 8 cases of trauma-related PCVL cases have been reported in the literature to date.[5–12] Herein we report a case of a 31-year-old man with a growing scalp mass at the site of a previous trauma involving the right side of the occipital and parietal bone.

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2. Case report

This study was approved by the ethics committee of The Second Affiliated Hospital of Dalian Medical University. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained. The approval number was 2,018,077.

A 31-year-old male was admitted to the Second Affiliated Hospital of Dalian Medical University (Dalian City, China) for treatment of a painful scalp mass at the site of head trauma induced by an accidental blow on an iron door handle 1 month previously. An immediate head computed tomography (CT) scan at his local hospital showed no scalp hematoma or fracture signs after the trauma.

Twenty-five days after the trauma, the patient noticed a fist-sized scalp mass on his right parietal occipital bone where he experienced volatile pain. He went to his local hospital and received a second head CT scan that showed skull destruction below the scalp mass. The patient was then transferred to our hospital for further treatment.

Physical examination confirmed a scalp mass at the right parietal occipital bone of the patient; the mass induced volatile, persistent, and paroxysmal pain. Physical examination detected no other positive signs. His medical history was otherwise unremarkable, and he had no family history of cancer and genetic disease. Laboratory tests showed an elevated white blood cell count (12.55×10^9/L). Hepatitis virus and HIV virus tests were negative. Cerebrospinal fluid examination showed no abnormalities. Considering the patient's age, history of head trauma, and the volatile pain associated with the scalp mass, a diagnosis of arteriovenous fistula was made.

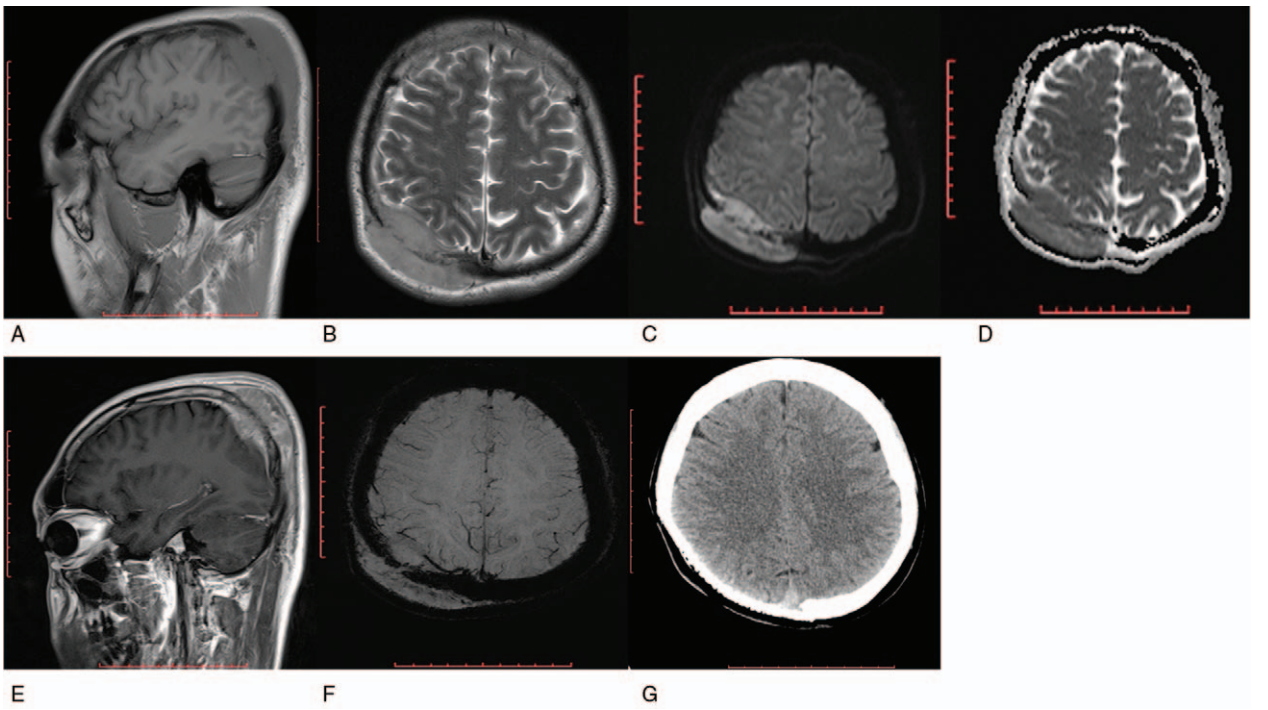

Figure 1. (A–D) A non-enhanced MRI scan revealing a solid mass on the right parietal occipital bone area with osteolytic erosion and intracranial and extracranial involvement. The mass was homogenous isointense on T_1WI (A) and T_2WI (B); and slight hyperintense on DWI (C) and hypointense on ADC map (D). (E) The mass was enhanced heterogeneously and a dural tail was seen. The neighboring brain tissue was compressed. (F) SWI showed no bleeding signals or vascular malformations in the mass. (G) There was no sign of recurrence in the CT image 12 months after surgical treatment.

A head magnetic resonance imaging (MRI) scan after admission revealed a solid mass on the right parietal occipital bone with osteolytic erosion and intracranial and extracranial involvement. The mass showed a homogenous isointense signal on T_1WI (Fig. 1A) and T_2WI (Fig. 1B), a hyperintense signal on DWI (Fig. 1C), and a hypointense signal on ADC map (Fig. 1D). The mass area was enhanced after contrast agent administration (Fig. 1E), showing a sharp boundary with brain parenchyma and a "dural tail sign." The right parietal occipital bone showed "sieve-like" destruction, while the general shape of the skull was preserved (Fig. 1E). The mass showed growth toward the intracranial space and compressed the brain parenchyma. The mass showed no definite bleeding signs or vascular malformations on SWI (Fig. 1F). Cerebral angiography also showed no positive signs of arteriovenous fistula or other vascular malformations.

The above radiologic findings did not support the diagnosis of arteriovenous fistula, but instead suggested malignant tumor. Meanwhile, the patient's condition deteriorated rapidly, and his headache aggravated and was not alleviated by analgesics. Thus tumor resection was performed under general anesthesia. During the operation, skull destruction and dura mater lateral violations were observed, while the medial surface of the meninges was found to be smooth. Intraoperative biopsy suggested malignant tumor. Hence, tumor total resection was performed and the dura was repaired.

Postoperative standard pathological results suggested the mass to be non-Hodgkin lymphoma (NHL), favoring plasmablastic lymphoma (Fig. 2Aand B). Immunohistochemistry further confirmed the classification of plasmablastic lymphoma (Fig. 2C–E). Specifically, cells were found to be immune-positive for LCA, CD30, MUM-1, EMA, and vimentin; partially positive for CD38 (Fig. 2C), CD138 (Fig. 2D), Bcl-6, and CD68; and negative for CD20 (Fig. 2E), CD79α, PAX-5, CD3, CD5, S-100, CD1α, MPO, HMB45, and AE1/AE3. Lysozymes were expressed in a few scattered cells; Ki-67 was positive in 40% of the tumor cells.

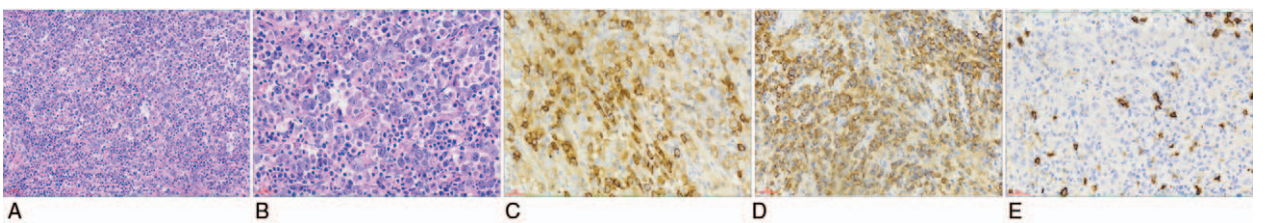

Figure 2. (A) HE staining showed large immunoblast-like cells with monoclonal proliferation (HE, ×200). (B) HE staining showing large and round tumor cells with thick chromatins and very small nuclei (HE, ×400). (C–E) Immunohistochemistry staining showed that CD38 (C) and (D) CD138 were partly positive, while CD20 (E) was negative (×200).

Two months after the operation, the patient received 2 regimes of velcade and hyper-CVAD, and 6 courses of intrathecalcytarabine and methotrexate. Twelve months after the operation, the patient received another head CT scan which showed no sign of recurrence (Fig. 1G), and a complete remission of the disease was determined.

3. Discussion

The diagnostic criteria for PLB include that the tumor mass is isolated from the bone, and there is no lesions in other parts of the body, or no spread systemic lesions for at least 6 months.[6,13,14] The tumor found in our case met these diagnostic criteria of PLB. According to most previous literature, PCVL more often occurs in elderly populations.[3,15] However, our patient presented with PCVL at a younger age. The mass in our patient was first misdiagnosed as arteriovenous fistula and then confirmed as PCVL basing on MRI and pathological findings. We herein summarize the clinical features of PCVL, and discuss the strategies to avoid misdiagnosis and the possibility of early diagnosis for timely treatment.

PCVL usually starts as a painless subcutaneous mass. When the mass gradually invades the cranial skull, dura mater, or infiltrates the cortex, corresponding focal neurologic signs appear such as headache, limb numbness, hemiplegia, and convulsions.[3,6,16,17] In our case, the patient had a history of head trauma and his scalp mass gradually increased to a large size. According to the literature, the differential diagnoses of PCVL include meningioma, metastatic infiltration, histiocytosis, subdural hematoma, plasma cell tumors, and osteomyelitis.[3,18]

Our patient was misdiagnosed with arteriovenous fistula because of his young age and history of head trauma. However, bone destruction appeared on the patient's MRI scan, and even meningeal invasion or cortical infiltration. Considering the rapid growth of his scalp lump, we suspected an alternative diagnosis.

Previously, Curty et al[9] reported a 19-year-old girl who had aggravated headache accompanied by nausea and vomiting 6 weeks following head trauma due to a motorcycle accident. Her condition showed no improvement after 1 week of treatment for subdural hematoma until her skull lump was surgically removed. The lump was pathologically confirmed as PCVL. Dai et al[10] reported a 21-year-old male patient who developed a mass in the forehead 4 months following head trauma. The final pathological diagnosis of the mass was diffuse large B-cell lymphoma.

Basing on these cases and our patient, we suspect that trauma may be one of the pathogenic factors of primary lymphoma in the skull. The causal relationship between trauma and primary lymphoma in the skull had not been well studied. As mentioned in,[8] prolonged or repeated information due to head trauma may lead to abnormal cell proliferation and the formation of neoplasm; the information might also attract circulating lymphoma cells to lodge and accumulate at the site of head trauma.

When a patient is admitted with scalp mass and consistent pain at the site of previous head trauma, the skull should be imaged to determine its completeness, and to determine whether the mass continues to grow. If the mass induces bone destruction and shows intracranial growth, PCVL should be suspected, although it is very rare.

Although the imaging features of PCVL are not characteristic,[3,14,16] CT and MRI scans play essential roles in judging the severity of PCVL. A tumor involving both sides of the cranial vault with minimal bone destruction might be one of the radiological characteristics of PCVL.[18] Head CT scans can accurately show extra-intracranial extent, and detect early bone and dura mater invasion. Head MRI can more accurately define the boundaries of PCVL lesions, and detect the invasion of the meninges and brain parenchyma, providing valuable information for targeted treatment strategies.[17] A whole-body CT scan or PET-CT helps to exclude the involvement of other parts of the body.

The final diagnosis of PCVL is dependent on the pathologic biopsy. Almost all of the pathologic types of PCVL are NHL, which account for most large B-cell lymphomas.[5] The pathology consistently confirmed that the tumor mass of our case was also B-cell-derived NHL.

Immunohistochemistry findings are particularly important for diagnosing PCVL. Diffuse positive anti-CD20 staining supports the diagnosis of diffuse large B-cell lymphoma, negative anti-CD30 staining suggests a low possibility of Hodgkin's lymphoma, and positive anti-CD3 staining often supports the diagnosis of B cell lymphoma.[6]

The current case was diagnosed as plasma cell lymphoma, which is a rare subtype of diffuse large B-cell lymphoma, based on the following immunohistochemistry findings. Briefly, CD38, CD138, and Mum-1 were partly positive, while CD20, CD19, and PAX5 were negative. To the best of our knowledge, this is the first report on the pathological type of skull lymphoma.

At present, there are no uniform standards for the treatment of PCVL.[6,13,16] Radiotherapy is an effective means of treating PCVL because it is a radiosensitive tumor.[6] It has been reported that the combined use of surgical treatment and radiotherapy can achieve good results[19,20]; for most cases, surgical treatment was the initial treatment.[21] A separate systemic chemotherapy can also achieve good therapeutic effect.[21] CHOP regiment is the most common chemotherapeutic regimen used.[13]

Our patient received total tumor resection and dural repair; then he was given chemotherapy regimen for adjuvant therapy. The patient survived without signs of systemic dissemination for 12 months after surgery at the time of last follow-up. In general, combined treatment consisting of surgery, chemotherapy, and radiotherapy is considered optimum treatment for PCVL.

4. Conclusion

The site of trauma complicated by PCVL can easily be misdiagnosed as scalp or subdural hematoma because of its extreme rarity. Our case and previously reported cases suggest that trauma may be one of the factors that induce PCVL. Although it is difficult to diagnose PCVL by radiographic imaging alone, radiographs can help determine the extent of the lesion and provide help for the choice of treatment.

The final diagnosis of PCVL depends on pathology and immunohistochemistry findings. According to the present case and a literature review of previous studies, it only takes a few weeks from the self-perceived mass to the emergence of neurological symptoms; therefore, early diagnosis and correct treatment are particularly important. In general, a combined treatment of surgery, chemotherapy and radiotherapy can achieve favorable outcomes.

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