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Diffuse-primary-B-cell lymphoma of the cranial vault presenting as stroke

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ABSTRACT

A rare case of diffuse-primary-B-cell lymphoma was misdiagnosed on emergency computed tomography because of blurred findings and a sclerotic appearance of the right parietal bone. In spite of computed tomography, magnetic resonance imaging provided a higher diagnostic yield, revealing more extensive diploic alterations and indicating the involvement of all of the cranial vault compartments. Therefore, a histologic examination of the surgical specimen was conducted to reach a conclusive diagnosis.

Introduction

Primary cranial vault non-Hodgkin lymphomas (CVLs) have been very rarely reported. They continue to represent a diagnostic challenge for both clinicians and radiologists because of the heterogeneous clinical presentations and radiological features.

A primary CVL can easily be missed during an initial diagnosis, mainly in the emergency setting as it shows focal neurologic deficits and mild sclerotic bone appearances. Only 2 cases of primary CVL have previously displayed purely sclerotic bone patterns [1,2].

Case presentation

A 74-year-old woman came to our emergency room for acute aphasia, left limb weakness, and an acute confusional state. Her family members spoke of an episode of a syncopal episode 3 months earlier. The neurologic examination showed a mild left-sided hemiparesis (Muscle scale grades MRC 4/5). A cerebral computed tomography (CT) revealed a subcortical hypodense area in the temporal lobe (Fig. 1A), which was interpreted as a subacute phase of ischemia. A bone window view showed, on the right parietal diploic space, a focal area of sclerosis and a mild bulging scalp, which were deemed less relevant

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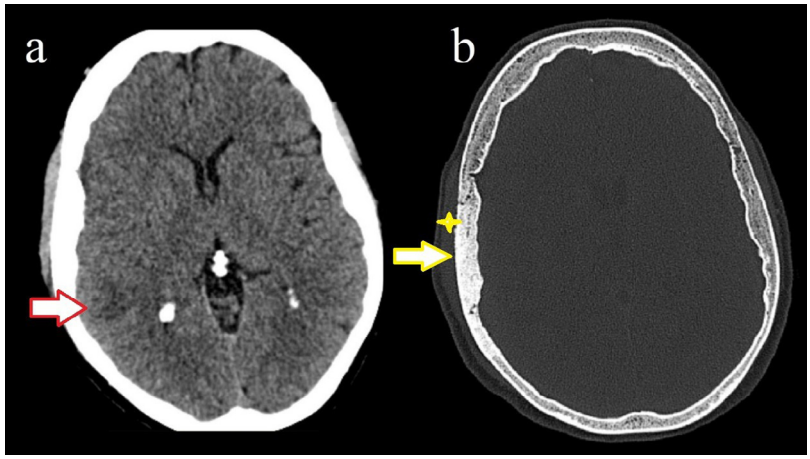

Fig. 1 – Computed tomography examination without contrast product administration. (A) The right hypodense subcortical temporal area is indicated by a white arrow with red border. (B) The sclerotic bone appearance of right parietal bone (white arrow with yellow border) with the imperceptible bulging (yellow star).

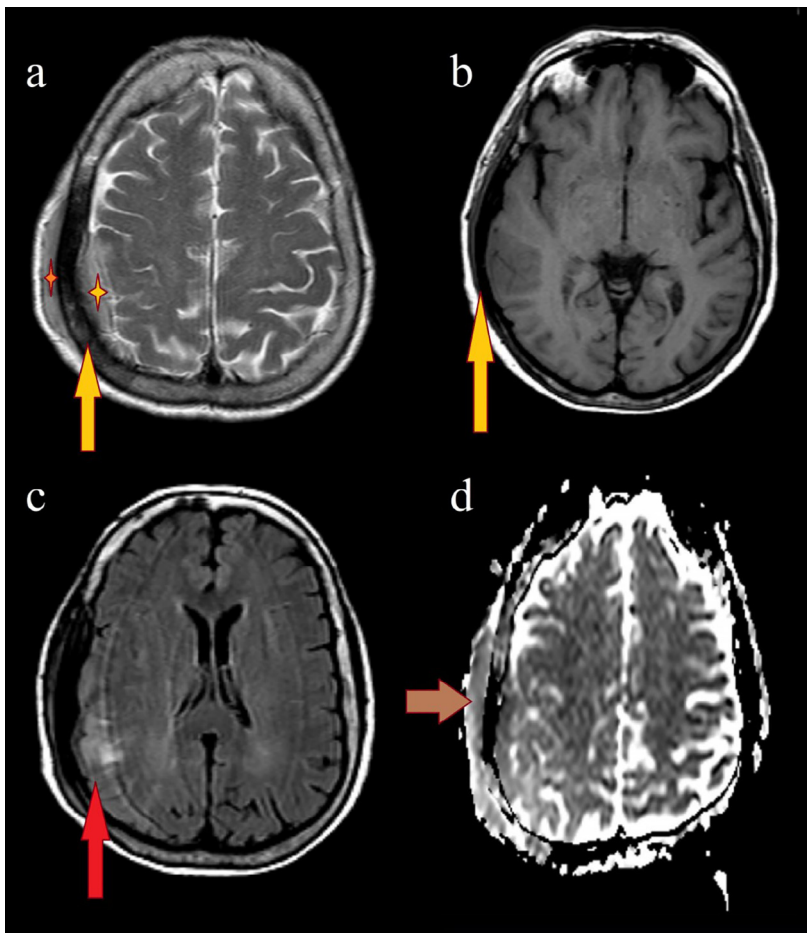

Fig. 2 – Morphologic magnetic resonance imaging sequences: (A) hypointense diploic signal of right parietal bone on T2 (long yellow arrow) with an isointense dural mass (yellow star) and swelling of the scalp (orange star); (B) the extended hypointense diploic signal on T1 (long yellow arrow); (C) the hyperintense edema on FLAIR (long red arrow); (D) the low apparent diffusion coefficient of soft tissue tumefaction (short orange arrow).

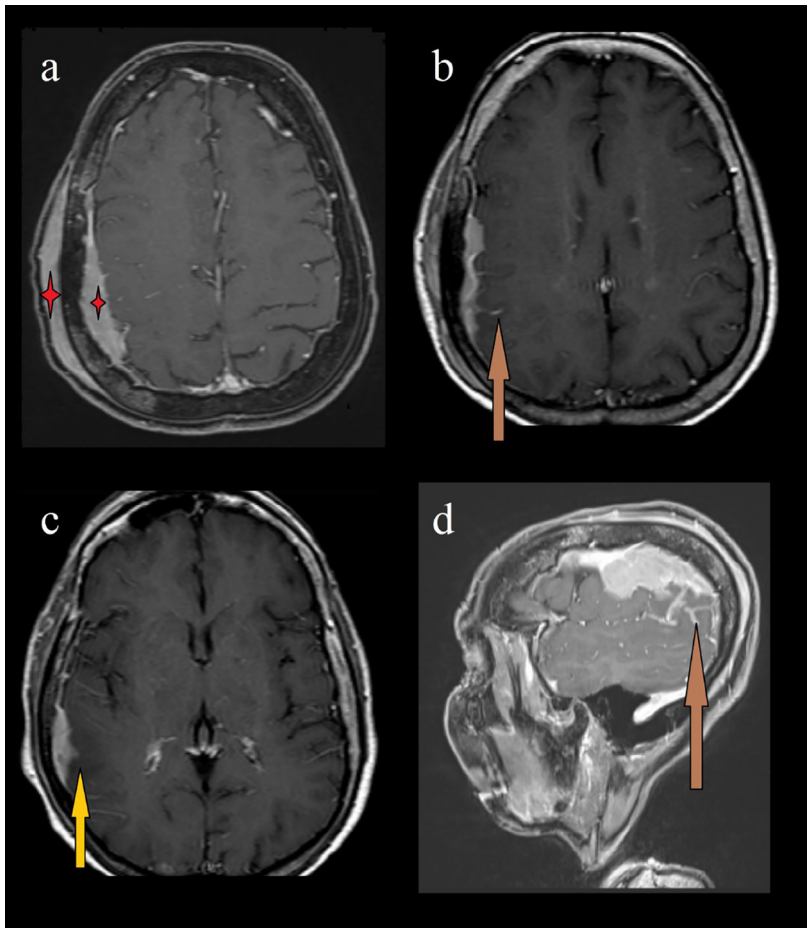

Fig. 3 – Magnetic resonance imaging with contrast product administration: (A) the homogeneous tumor's enhancement (red stars); (B) leptomeningeal involvement on axial plane (long orange arrow); (C) indistinguishable border from the meninges and brain cortex with subcortical edema (long yellow arrow); and (D) the leptomeningeal involvement on sagittal plane (long orange arrow).

(Fig. 1B). Therefore, an anticoagulant oral therapy (aspirin 300 mg/d) was commenced. However, the patient's confusional state continued to worsen and clinicians requested a magnetic resonance imaging (MRI) 48 hours later. The morphologic sequences highlighted in the right parietal-temporal diploe a more extensively altered signal, which resulted hypointense on both T2 and T1 sequences (Fig. 2A and B). A correlated dural-based hysointense mass on T1 and T2 (Fig. 2A) was also observed, with restricted diffusion and was surrounded by a hyperintense edema (Fig. 2C). Additionally, the near scalp showed restricted diffusion with low apparent diffusion coefficient (mean apparent diffusion coefficient value 0.784×10^{-3} mm^2/s) (Fig. 2D). The study was completed by gadolinium-based contrast agent administration and the same areas were enhanced homogeneously (Fig. 3A). An indistinguishable border from the meninges to the brain cortex was also described (Fig. 3C), with right parietal sulci leptomeningeal involvement (Fig. 3B and D). The initial diagnosis was a primary extracranic tumor with intracranic extension. The patient was initially treated with methylprednisolone (1000 mg/d intravenously) and levetiracetam (500 mg twice daily). An accurate workup showed a normal white blood cell counts (6.78×10^9/L), with 62% neutrophils, lymphocytes 26%, and a normal platelet counts. The hemoglobin level, hematocrit, mean cell volume, and other laboratory values, including electrolytes, creatinine, and liver enzymes, were in the normal range. The lactate dehydrogenase (LDH) value was 154 U/L, also normal. The patient tested negative for HIV virus and the thorax-abdominal CT showed no other primary tumoral localizations. However, when a craniectomy with both scalp bone and intracranial tumor excisions were carried out, a grayish white tumor with a soft consistency was found (Fig. 4A). The parenchymal infiltration and the involvement of right temporal muscle were confirmed. The histological examination of the surgical specimen revealed a diffuse-primary-B-cell lymphoma (Fig. 4B and C) with immunohistochemistry CD20+, CD10+/–, PAX5+, BCL6+, BCL2+/– and Cyclin D1–, MUM1–, CD23–, and high cell proliferation index (Ki67 > 60%) (Fig. 4D). A bone marrow biopsy from the posterior iliac crest resulted negative for lymphoma. The patient was then given a chemotherapy regimen consisting of 4 cycles of R-CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisolone) every 3 weeks. After 6 months of therapy, the patient showed no signs of systemic dissemination.

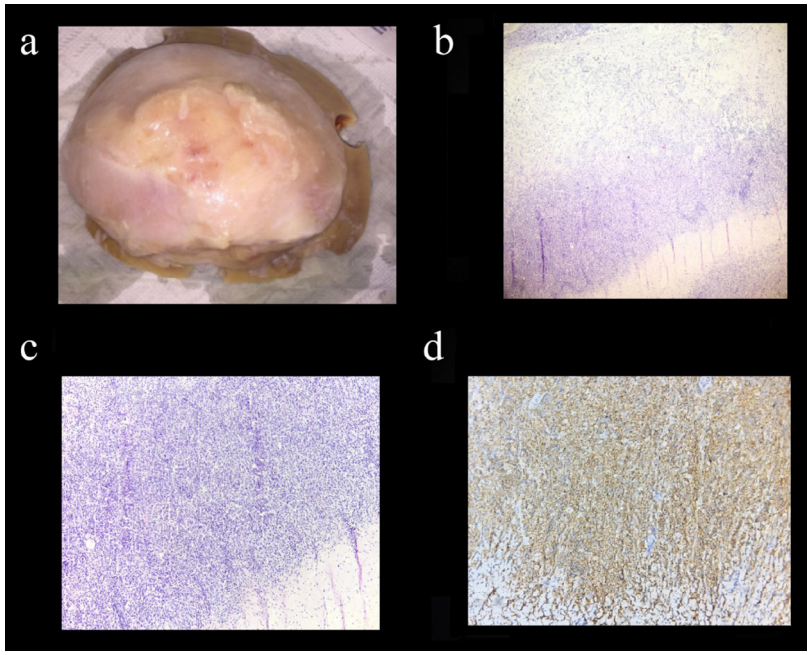

Fig. 4 – (A) The craniotomy flap containing tumor's grayish white appearance photographed at surgical examination. (B-D) Photomicrographs of a surgical specimen showing a diffuse proliferation of pleomorphic atypical cells on original magnification 4× (B) and original magnification 10× (C); strong CD20 staining in the tumor cells at immunochemistry (D).

Discussion

Primary bone lymphoma is defined as a lymphoma within a single bone, either with or without regional nodal metastasis, and lacks distal lesions within 6 months after diagnosis [3]. The cranial vault is rarely the primary location of non-Hodgkin lymphomas, usually with a predominance of diffuse large B-cell subtype. The typical pattern of CVL is the involvement of all of the cranial vault compartments with intra- and extracranic extensions. Lymphoma cells spread along the surrounded soft tissue through the emissary vein and tend to infiltrate the brain cortex by leptomeningeal involvement or by direct infiltration [1,2,4,5]. Clinical presentations vary from tumors discovered by chance in asymptomatic patients to a history of painless scalp tumefactions and headaches [1,2,4–6]. Focal neurologic deficits, seizures, and acute confusional states are less common and are usually associated with brain cerebral cortex infiltration [1,4,5]. The typical and more frequent bone pattern is bone destruction, characterized by a moth-eaten or a permeative lytic appearance [1,3,7,8]. Bone hyperostosis and blastic-sclerotic patterns that appear as mixed lytic or sclerotic lesions have also been observed [1,3,4,8,9]. Purely sclerotic lesions, local or diffuse, are rarely found in CVL. They are more common in bone metastatic lymphomas [1,3,8,9]. It has been suggested that sclerotic to lytic transformations are associated with disease progression [1]. In this case, the CT examination was misleading as the sclerotic bone appeared blurred and due to the fact that the radiologists were more focused on vascular ischemia since the patient displayed stroke-like symptoms. Only the MRI, used to double check the vascular ischemia, was able to spot the advanced tumor mass with intracranic and extracranic involvement. Furthermore, with unclear clinical presentation and inconspicuous swelling and tumefaction of the scalp, it is common to overlook, underestimate, or misdiagnose bone sclerosis, frequently as benign bone tumors such as meningioma or osteoma [2]. MRI is an accurate tool to study bone marrow abnormalities [3]. In fact, in this case, the discovered diploic alterations were more significant in size on the MRI, appearing hypointense on T1- and T2-weighted sequences. In most of the previous cases, CVL usually showed an isointensity signal on unenhanced MRI [1,3,7]. Hypointensity tumor signal on T2-weighted MRI has been reported only in 9% of cases and this feature may be related to fibrosis content [1,3]. Administering a contrast product is useful in establishing lesion extensions and dural and leptomeningeal enhancements. An indistinguishable border from the meninges to the brain cortex associated with subcortical edema is a strong indication of brain invasion [1], although many other different diseases can mimic CVL on MRI. Anaplastic meningioma, for instance, could appear identical to a CVL on an MRI [10]. Other differential diagnoses include Ewing sarcoma, which usually occurs in younger patients with florid aggressive periosteal reaction, or calvarial metastases, especially from thyroid and renal cancers, which can manifest itself in the form of extended lesions with bone destruction that invade the soft tissue [11] and osteomyelitis, although this is usually associated with external otitis and systemic symptoms.

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