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CASE REPORT

Primary malignant large B-cell non-Hodgkin's lymphoma of the scalp and cranial vault: a case report and an overview

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Abstract

Primary non-Hodgkin's malignant lymphoma of the bone is a rare entity, accounting for 4% of all non-Hodgkin's malignant lymphomas (NHL). Cranial vault involvement is rarer, found only in 0.2% of cases (1)(2). Only few cases have been reported previously in the literature (3)(4).

Here, the authors are reporting a new case of primary malignant large B-cell non-Hodgkin's lymphoma of the cranial vault because of its rare occurrence.

Key words: nonHodgkin's malignant lymphoma, scalp, surgery

1 | INTRODUCTION

Primary non-Hodgkin's malignant lymphoma of the bone is a rare entity, accounting for 4% of all non-Hodgkin's malignant lymphomas (NHL). Femur, tibia and pelvis are the most common sites. Cranial vault involvement is rarer, found only in 0.2% of cases (1) (2). The absence of involvement of the cerebral parenchyma and systemic or skeletal manifestation is also rare. Only few cases have been reported previously in the literature (3) (4). Primary malignant lymphoma of the bone is defined as an isolated bone lesion with no signs of other sites involvement and no blood-borne spread within six months of diagnosis (5).

Here, the authors are reporting a new case of primary malignant large B-cell non-Hodgkin's lymphoma of the cranial vault because of its rare occurrence.

Case report

A 30-year-old young lady with no medical or surgical history was admitted from our outpatient department of neurosurgery for progressively growing of a painless scalp lump at the level of the parieto-occipital right bone. There were no signs of raised intra cranial pressure mostly without headache. The patient confirmed no prior head trauma, fever, or general condition impairment. Upon neurological examination, she was awake, alert and well oriented with no neurological disorder. Local examination of her scalp swelling revealed a hard and well-defined nontender immovable solitary subcutaneous painless lump, measuring 7cm in diameter. She had no lymphadenopathy or hepatosplenomegaly and the rest of her body check-up showed no abnormalities including her lymph node areas. The complete blood count showed hemoglobin level of 11.4 g/dL, white blood cell count of 8500/mm3 with a platelet count of 331000/mm3. Both blood serum ionogram and hemostasis assessment and evaluation were normal.

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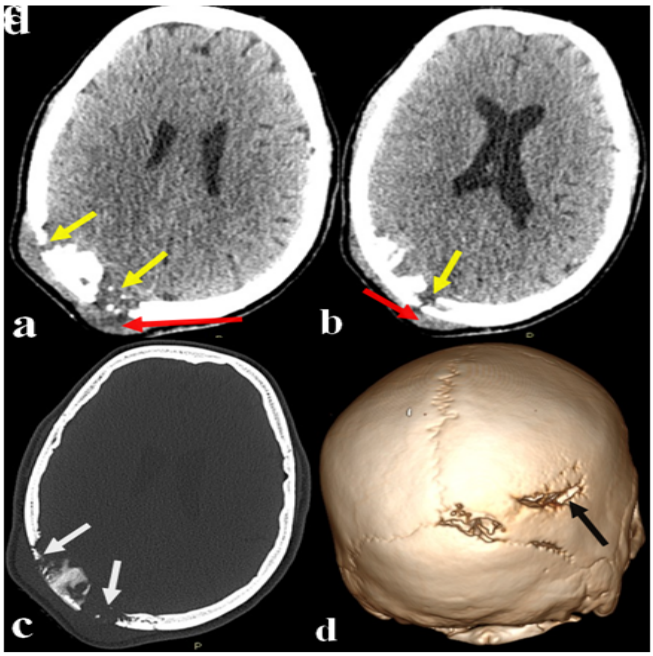

Fig. 1: Axial brain computed tomography (CT) scan in parenchymal window (**a**, **b**) and bone window (**c**) showing expansive osteolytic lesion (**a**, **b**; yellow arrows) (**c**: white arrows) in the right parieto-occipital bone measuring 3.6 * 1 cm in diameter without perilesional edema or a midline shift; d: three-dimensional CT reconstruction showing bone lysis (black arrow). Note the subcutaneous lump clearly visible (**a**, **b**; red arrows).

A brain computed tomography (CT) scan (Figure 1) was thus performed showing an expansive osteolytic lesion in the right parieto-occipital bone measuring 3.6 * 1 cm in diameter without perilesional edema or a midline shift.

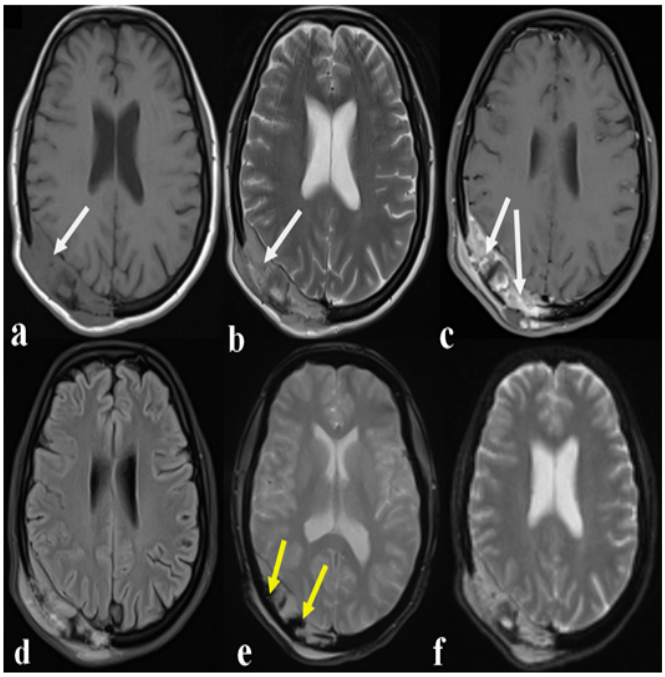

Fig. 2: Axial brain magnetic resonance imaging showing a fairly lytic bone mass inisosignal on T1-weighted image (**a**; white arrow) and discreet hypersignal on T2 sequence (**b**; white arrow) with homogenous enhancement after Gadolinium chelates injection (**c**; white arrows). Fluid attenuated inversion recovery sequence showed discreet hypersignal (**d**) and GRE T2* weighted image revealed low intensity signal in favor of bone signal (**e;** ; yellow arrows). Diffusion weighted image showing a significant restricted signal (**f**).

Further investigation through magnetic resonance imaging (MRI) (**Figure 2**) confirmed the fairly lytic bone mass in isosignal on T1-weighted image and discreet hypersignal on T2 sequence with homogenous enhancement after Gadolinium chelates injection. The lesion comes into contact with the superior sagittal venous sinus without infiltrating it. Diffusion weighted image showed a significant restricted signal with low apparent diffusion coefficient (ADC) value on ADC mapping. As part of the extension assessment, the thoraco-abdominopelvic CT, breast and thyroid ultrasound, as well as bone scintigraphy were without abnormalities. All these finding suggested thus a primary bone scalp lymphoma.

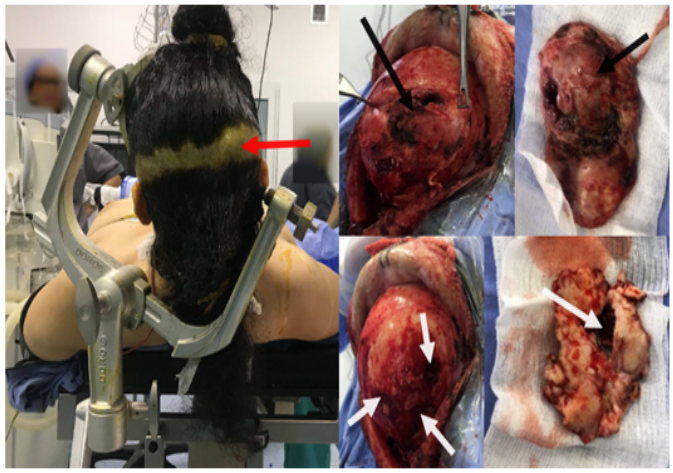

Fig. 3: Preoperative (left image) and intraoperative (right image) photographs in the operating room showing the supine position with patient's head fixed in Mayfield head clamp turned slightly to the left side biparietal skin incision (red arrow) was performed and an excision of her bone mass through. Preoperatively the mass was subcutaneous, hard, grayish, and easily cleavable (black arrows). The dura mater was intact and the bone scalp was found to be moth eaten (white arrows).

The patient underwent, under general anesthesia and in supine position with his head fixed in a Mayfield head clamp turned slightly to the left side (Figure 3), an excision of her bone mass through a large biparietal skin incision. The mass was subcutaneous, hard, grayish, and easily cleavable. The dura mater was intact and the bone scalp was found to be moth eaten. By the end a cranioplasty was performed covering the tumor bed. The mass along with the involved bone were sent for histopathological examination.

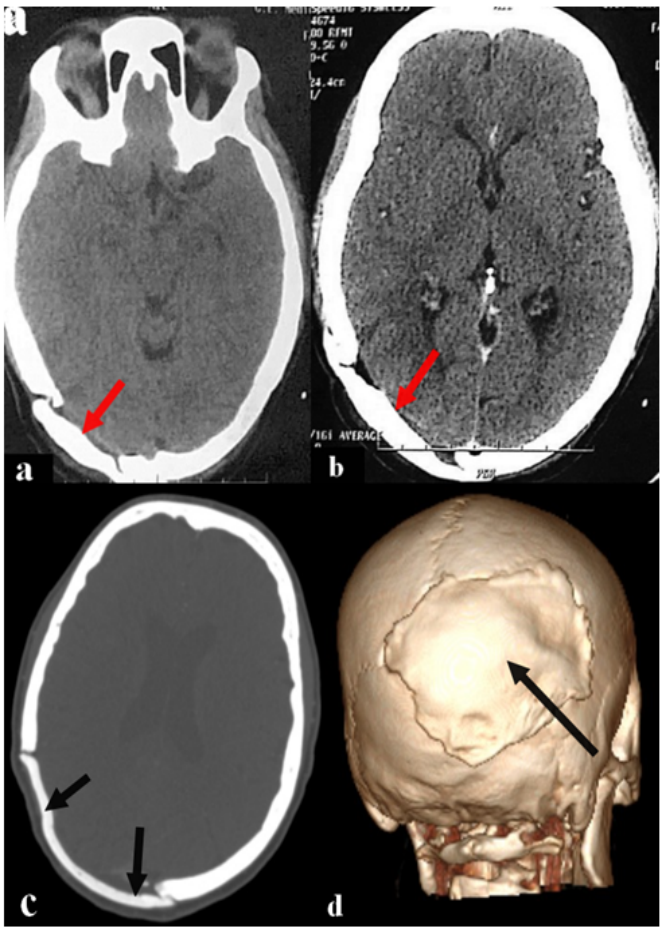

Fig. 4: Immediate axial postoperative CT scan in parenchymal window before (**a**) and after enhancement (**b**) showing the absence of tumor residue. Axial CT in bone window (**c**) and three-dimensional reconstruction (**d**) correct placement of the cranioplasty (**e**).

Postoperative course was uneventful. Immediate postoperative CT scan (Figure 4) showed the absence of tumor residue as well as correct placement of the cranioplasty.

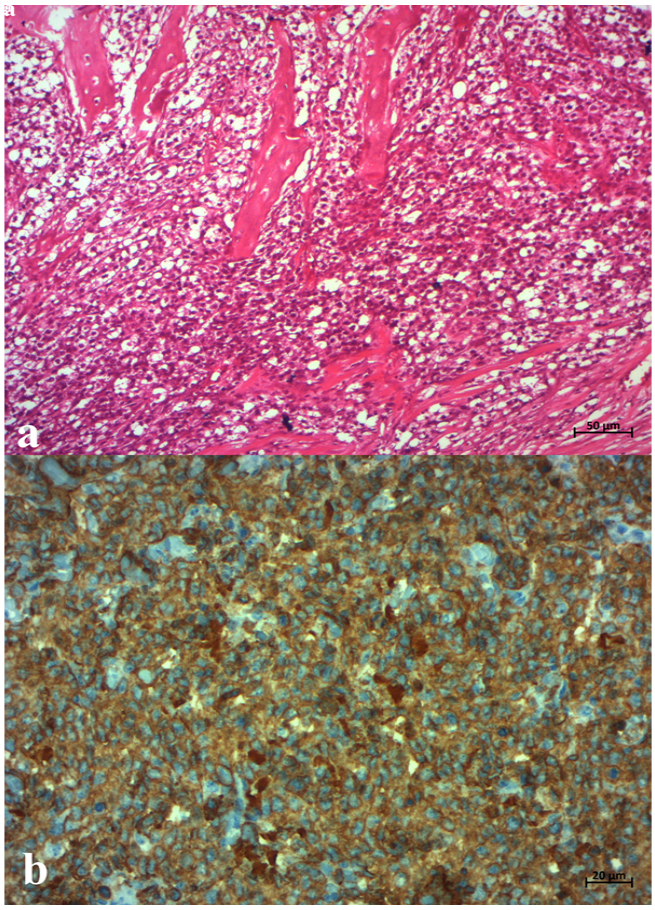

Fig. 5: Photomicrographs of a surgical specimen showing tumor proliferation of similar morphology; tumor cells were round, of large size, with round nucleus, lobulated vesicles and small nucleoli, more or less prominent; mitoses were very numerous; this proliferation massively infiltrates the bone tissue; it was in places delimited by a small cell lymphoid sleeve (**a**; HE x 100). Immunohistochemistry (**b**) showing diffuse immunoreactivity for leukocyte CD20.

Histopathological examination (**Figure 5**) was con-ducted showing tumor proliferation of similar mor-phology on all samples; it is a dense proliferation in diffuse, partly lobulated layers; the tumor cells were round, of large size, with round nucleus, lobu-lated vesicles and small nucleoli, more or less promi-nent; mitoses were very numerous; this proliferation massively infiltrates the bone tissue; it was in places delimited by a small cell lymphoid sleeve. There were small areas of necrosis. On immunohistochem-istry tumor cells were massively immunoreactive for leukocyte common antigen (LCA), CD20, Oct 2, and Bcl-6 and they

were weakly positive for CD 10. The proliferation index Ki 67 was estimated to be 90%.

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The patient was discharged of our department at day 4 postoperatively and addressed to the oncology's department for further management. She received 3 cycles of systemic chemotherapy. The cycles were repeated after 3 weeks.

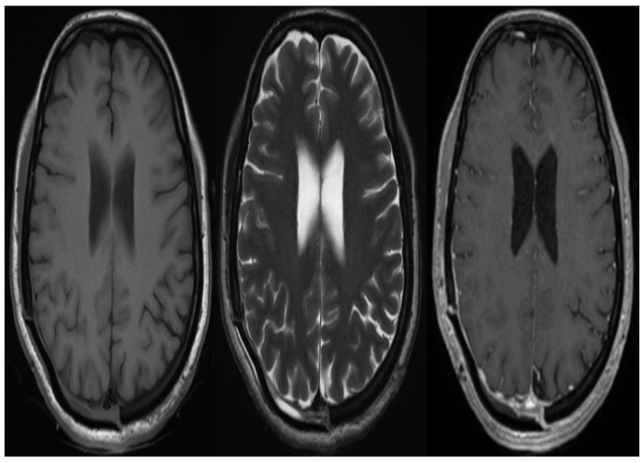

Fig. 6: Brain MRI in axial plane at 01 month postoperatively showing no signs of tumor recurrence on T1 and T2 weighted images as well as after enhancement.

Our patient underwent, at our out-patient clinic, a physical examination, complete hemogram, and brain MRI (**Figure 6**). All these investigations did not revealed no signs of tumor recurrence.

2 | DISCUSSION:

Primary bone lymphoma is a rare malignant tumor, which accounts for only 4% of non-Hodgkin lymphomas (1). This entity is defined by the isolated presence of lymphoma in the bone without other distant localization within six months of diagnosis (5). Bone involvement primarily affects the femur, humerus, pelvis, spine, mandible, and scapula. Primary localization to the cranial vault remains extremely rare (6). It has been reported in immunocompromised or trauma patients. However primary NHL with extra- and/or intra-cranial extension without systemic or skeletal involvement in a nonimmunocompromised or nontraumatic context is extremely uncommon (4). To our knowledge, only 19 cases had been reported previously in the literature (1) .

The inaugural clinical signs revealing the disease are painless lump in the scalp observed in 90% of patients, headache due to bone destruction or tumor infiltration of the meninges in 30% of cases, and signs of focal neurological palsy secondary to tumor infiltration of the cerebral parenchyma in 10% of cases (1) (6). Lymphoma also may infiltrate the spaces within the diploe and along the emissary veins reaching the soft tissues on either side of the bone. In our patient, the painless scalp lump was the revealing mode with no other neurological symptoms. The parieto-occipital bone destruction as well as the sparing of the dura were both obvious peroperatively.

Iconographic data is not sufficient to confirm the diagnosis. Thus, on MRI, these lesions are relatively nonspecific, appearing in hypointense on T1-weighted sequences and hypersignal on that of T2 (2). Possible differential diagnoses are

Iconographic data is not sufficient to confirm the diagnosis. Thus, on MRI, these lesions are relatively nonspecific, appearing in hypointense on T1-weighted sequences and hypersignal on that of T2 (2). Possible differential diagnoses are osteomyelitis, metastatic or primary skull tumors, and intraosseous meningioma. These lesions are accompanied by significant cortical bone destruction unlike lymphoma.

Histologically, most cranial vault lymphomas are non-Hodgkin's large B-cell lymphomas expressing CD20 but not CD5, CD10 and CD23 (6). In our patient, immunohistochemistry showed a tumor cells immunoreactivity for LCA (CD 45), CD20, Oct 2, and Bcl-6 with less important immunoreaction for CD 10.

Due to the rarity of cases described in the literature, the optimal treatment of primary lymphoma of the cranial vault has not been well established and no definite conclusion can be drawn. Some authors rec-ommend surgery followed by radiotherapy in case of localized disease, and chemotherapy in case of metastatic progression (7). It must be remembered that first of all, since primary lymphoma of the cen-tral nervous system (CNS) is multifocal and infiltrat-ing, the role of surgery, unlike other brain tumors, is purely diagnostic. The place of surgery in primary bone lymphomas located in the cranial vault remains limited to the diagnosis of histopathology (bone biopsy). This dogma was recently challenged by a second analysis of the largest randomized trial of pri-mary CNS lymphomas, which included 526 patients and found a survival benefit from first surgery (8) . Further tests are needed to confirm these results and to this day it remains confined to its diagnostic role.

In view of the low incidence and the paucity of the reported cases, it is still difficult to establish a definitive prognosis for this disease. El Asri et al. (6) reported that 13 were still alive among 16 patients followed over a period of 12 months. In a review published by Aquilina et al. (9), And among 16 patients with vault lymphoma, 6 had no systemic involvement at the time of diagnosis and continued to be symptom-free after treatment with follow-ups ranging from five months to 72 months.

Although it remains a rare entity, primary non-Hodgkin's malignant lymphoma should be cited in the differential diagnoses of expanding scalp lesions. The treatment must necessarily involve a multidisci-plinary therapeutic approach, including several spe-cialties including neurosurgery, radiotherapy and oncology

3 | CONCLUSION

Primary lymphoma of the cranial vault remains an extremely rare tumor. Its treatment is still not well standardized. The most optimal therapeutic option is based on the combination of surgery and / or radio-therapy and chemotherapy. Therefore, a thorough search is needed to improve the management of this disease as well as its prognosis.

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