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Primary Cranial Vault Lymphoma with Extra- and Intracranial Extension: A Case Report

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Abstract **Case Report**

Primary cranial vault lymphoma with extra- and intracranial extension in an immunocompetent patient is extremely rare. Given its rarity, the optimal management of this entity remains uncertain. We report an atypical case of a 21-year old immunocompetent man, who presented with an 8-month history of enlarging parieto-occipital mass. CT and MR imaging revealed a large mass with extra cranial and cranial components with sagittal sinus involvement. Unusually, angiography showed a significant tumor blush with large feeding vessels. After a pre surgical embolization, the patient underwent a complete tumor resection followed by radio and chemotherapy with satisfactory results.

Keywords: Primary cranial vault lymphoma; MRI, CT.

INTRODUCTION

Primary extra-lymph node lymphomas are not uncommon, but they often cause difficulty in diagnosis. The most common skeletal locations of non-Hodgkin lymphomas (NHL) are the spine, pelvis, rib, and long tubular bones, especially the lower extremities. Primary NHL of the skull with extra- and intracranial extension without systemic or skeletal manifestations in an immunocompetent patient is extremely rare. Because of the rarity of this entity and the lack of evident literature, its optimal management is still unknown. We describe here a case of primary cranial vault lymphoma (PCVL) in an immunocompetent young male that presented with extracranial, cranial, extra- and intra-axial components with high degree of vascularity and sagittal sinus involvement. The patient underwent a total resection after preoperative embolization.

CLINICAL CASE

A 21-year old immunocompetent man presented with an 8-month history of enlarging parieto-occipital mass without history of trauma. Physical examination showed a firm, non-tender and non-pulsatile subcutaneous mass in the left parieto-occipital region, measuring 9 x 6 cm. There was no local rise of temperature, and the skin over the lesion was healthy. The remainder of the examination was unremarkable; in particular no lymphadenopathy and no abdominal masses were felt.

Neurological examination was normal. The blood investigations were all within normal limits. X-ray chest and ultrasonography of the abdomen were normal.

Computed tomography (CT) showed a large hyperdense mass occupying both intra- and extra cranial spaces. Bone windows showed a significant bone lysis of the parietal bone with rupture of the internal and external table and without periosteal reaction opposite (Figure 1). Magnetic resonance imaging (MRI) revealed an isointense mass on T1 and T2- weighted images that homogeneously enhanced after Gd-DTPA (figure 2). The lesion destroyed the cranial vault of the parietal region and invaded the epidural space, causing a diffuse thickening of the dura. The imaging also showed an enhancing intra-axial component of the lesion responsible for mass effect on the parietal lobe with cerebral edema and involvement of the superior sagittal sinus.

Angiography was performed (figure 3) and showed feeding arteries originating mainly from the bilateral occipital arteries, and incidentally from the left superficial temporal artery and the left middle meningeal artery. After a preoperative embolization, a parietal craniectomy was performed at the limit of the

extra- cranial part which has made it possible to remove the extra-cranial part en bloc and to minimize the bleeding. The tumor was moderate vascular, firm, dense and infiltrating the scalp and the bone (Figure 4 A and B). The dura involvement was diffuse and it was removed with wide margins, with the extra-axial part of the tumor and the invaded part of the superior sagittal sinus. Duraplasty was performed using autologous fascia lata. Eventually, the patient underwent a total mass removal with cranioplasty using acrylic cement. Postoperative period was uneventful.

Histopathology revealed a diffuse, large B-cell type of non-Hodgkins lymphoma (NHL). Bone marrow biopsy from the iliac crest was normal.

The diagnosis was Primary Cranial Vault Lymphoma (CVL). The patient was later referred to the oncology department to receive subsequent radio and chemotherapy. At a follow-up of 3 months (Figure 4C), he had remained well with no evidence of a tumor recurrence. He is followed-up regularly in consultation.

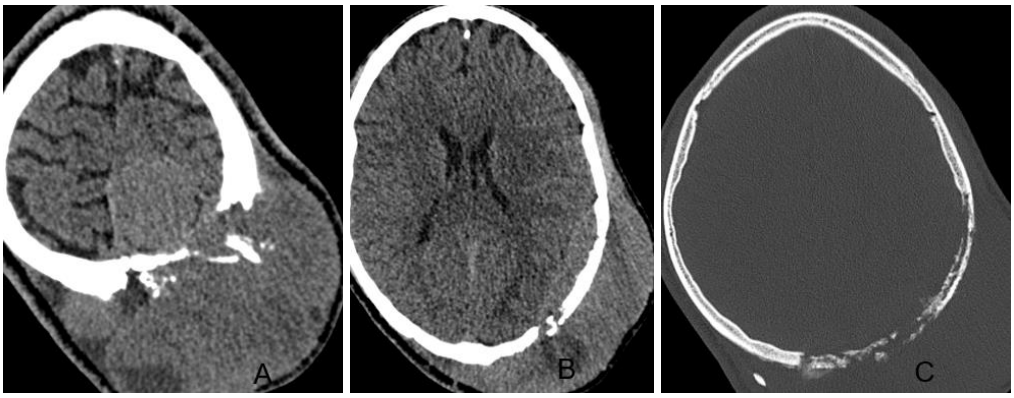

Figure 1: Brain scan with axial acquisition before (A) and after PDC (B) injection and bone window (C). Large hyperdense mass which enhanced after the injection of the contrast product, occupying both intra- and extracranial spaces. The bone window (C) revealed significant bone lysis of the parietal bone with rupture of both the internal and external tables, without any opposite periosteal reaction

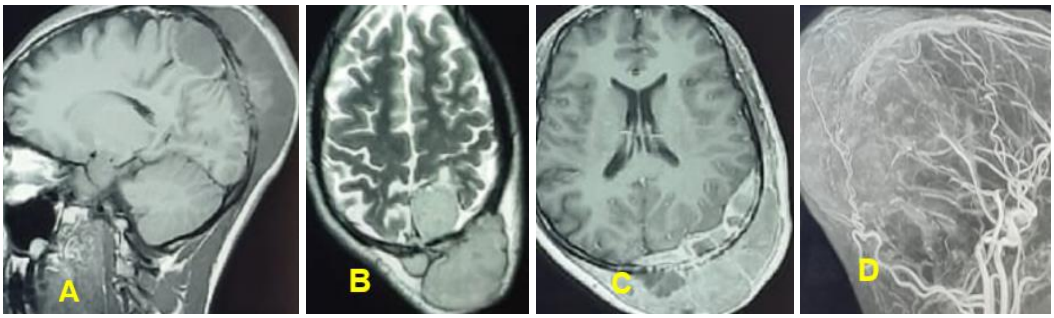

Figure 2: Cr Brain MRI with axial T1 sequence after gadolinium injection (A) and T2 (B), sagittal T1 sequence without gadolinium injection (C) , and an angiography sequences (D). Isointense mass with contrast enhancement on left parieto-occipitlal region invading the cranial vault, infiltrating the dura mater and the parietal lobe in postcentral gyrus and superior parietal lobule. Magnetic resonance imaging (MRI) with MR angiography demonstrated involvement of posterior part of superior sagittal sinus

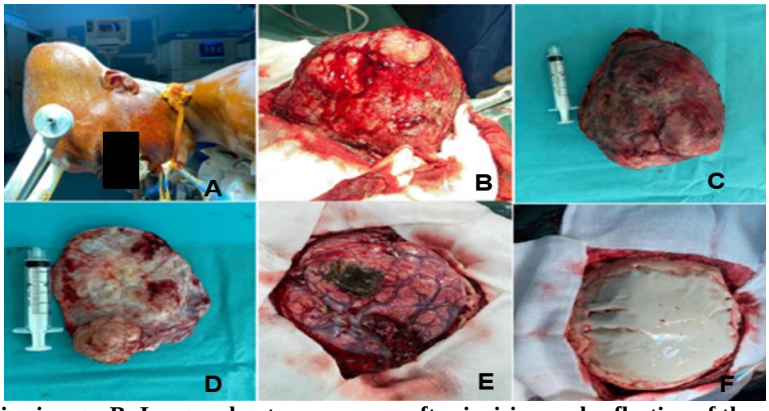

Figure 3: A: pre-operative image. B: Large subcutaneous mass after incision and reflection of the scalp. C: Specimen after removal on bloc of extra-cranial part and the bone invaded. D: Specimen after removal of the dura mather invaded and the intra-cranial part. E: Final results after total removal of the tumor. F: cranioplasty using cement acrylic

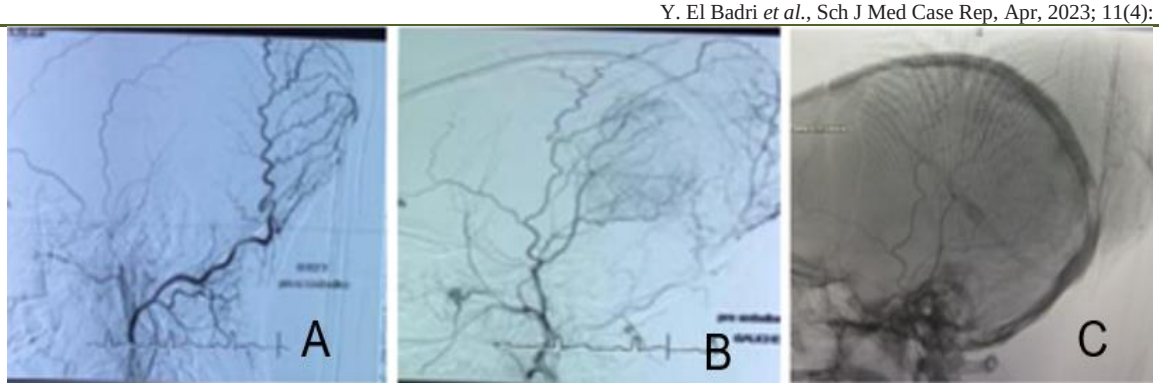

Figure 4: Lateral angiography showing tumour blush. The tumor is fed by the left superficial temporal artery, the left middle meningeal artery and principally the bilateral occipital artery (A and B). (C) Post-embolization control reveals a good result of obliteration of tumor blush.

DISCUSSION

Primary extra-lymph node lymphomas are not uncommon, but they often cause difficulty in diagnosis. A progressive increase in the incidence of lymphomas has been observed in the last decade, both in individuals affected by immunodeficiencies and in the general population [1]. PCVL occurs in all ages, but is much more common in older patients, with a mean age of 60.5 years and 60% of patients older than 60 years. The sex ratio was 1:1 M/F, showing no significant gender difference [2]. It mostly affected the femur or pelvis (50%), and humerus (20%), with the remaining 30% occurring in the spine, ribs, mandible, and scapula [1]. Primary lymphoma of the skull vault is extremely rare [3].

Primary cranial vault lymphoma is rarely different from typical metastatic bone lymphomas, and it is defined as solitary mass lesion without any evidence of disease at another site and no systemic dissemination within 6 months of tumor detection [1, 4]. The clinical presentation of CVL can vary depending on the location and extent of the lesion. Intracranial symptoms, such as seizures and hemiplegia, have also been observed [5, 6]. Cranial vault lymphoma involves cranial bone destruction. Although bone change is initially minimal, these tumors can ultimately infiltrate the skull and destroy it completely [7]. In our case, the patient presented the same pattern and had a bulky subcutaneous mass without associated neurological deficits. Our patient had a significant bone lysis of the parietal bone with rupture of the internal and external table and without periosteal reaction opposite. These results are likely due to a delay in seeking medical consultation, estimated to be around 8 months.

The clinical presentation of CVL can vary depending on the location and extent of the lesion. Commonly, it presents as a painless, palpable mass that can lead to bony erosion and invasion of adjacent structures, as seen in this case. In some instances, patients can also present with neurological symptoms such as seizures, headaches, or focal deficits [8].

Imaging plays a crucial role in the diagnosis and management of CVL. CT and MRI are the preferred imaging modalities that can provide detailed information about the location, extent, and invasion of adjacent structures [9]. The CT findings in this case of primary cranial vault lymphoma are consistent with previous literature reports which have described hyperdense masses occupying both intra- and extracranial spaces, with bone lysis and rupture of the internal and external tables of the skull without periosteal reaction opposite [10]. This is because the bone destruction in primary cranial vault lymphoma is typically caused by tumor invasion rather than periosteal reaction. A study by Wu *et al*., (2016) reported that hyperdense masses were present in 86% of primary cranial vault lymphoma cases, and that bone lysis with or without periosteal reaction was observed in 92.9% of cases [11]. The study also noted that the presence of hyperdense masses on CT scans may help distinguish primary cranial vault lymphoma from other diseases such as meningioma, which typically appears as a low-density mass on CT scans [12]. Another study by Ma *et al*., (2015) reported similar CT findings. The authors noted that the presence of bone destruction on CT scans may be helpful in differentiating primary cranial vault lymphoma from other diseases such as scalp or skull metastases, which may appear as soft tissue masses without bone destruction [6, 7].

In this case, MRI showed an isointense mass on both T1 and T2-weighted images, which enhanced homogeneously after Gd-DTPA injection. The lesion destroyed the cranial vault of the parietal region and invaded the epidural space, causing thickening of the dura. The MRI findings in this case are consistent with those reported in the literature for lymphomas [2]. However, given the nonspecific nature of these imaging findings, a definitive diagnosis of lymphoma was made only after histopathological examination of the tumor [5,9]. Other diseases can mimic the radiographic features of lymphomas, and therefore, a multidisciplinary approach involving radiologists, pathologists, and clinicians is essential for the accurate diagnosis and management of these tumors [2, 10].

Histopathological examination of the lesion confirmed the diagnosis of diffuse, large B-cell type of NHL, which is the most common subtype of NHL. The treatment of CVL usually involves a combination of surgery, chemotherapy, and radiation therapy. In this case, the patient underwent a parietal craniectomy with total mass removal and cranioplasty using acrylic cement, followed by subsequent radio and chemotherapy. The patient remained well with no evidence of tumor recurrence during the 3-month follow-up.

CONCLUSION

Primary cranial vault lymphoma is an extremely rare entity. It should be considered in the differential diagnosis of a lesion involving all three compartments of the cranial vault including the scalp, skull, and pachymeninges. The optimum management of PCVL remains uncertain; however, surgical removal, followed by radiotherapy and chemotherapy can be recommended.

REFERENCES

1. Adams, H., Tzankov, A., d'Hondt, S., Jundt, G., Dirnhofer, S., & Went, P. (2008). Primary diffuse large B-cell lymphomas of the bone: prognostic relevance of protein expression and clinical factors. *Human pathology*, *39*(9), 1323-1330.
2. Aquilina, K., O'brien, D. F., & Phillips, J. P. (2004). Diffuse primary non-Hodgkin's lymphoma of the cranial vault. *British journal of neurosurgery*, *18*(5), 518-523.
3. Ferreri, A. J. M., & Reni, M. (2007). Primary central nervous system lymphoma. *Crit Rev Oncol Hematol*, 63, 257–268.
4. El Asri, A. C., Akhaddar, A., Baallal, H., Boulahroud, O., Mandour, C., Chahdi, H., ... & Boucetta, M. (2012). Primary lymphoma of the cranial vault: case report and a systematic review of the literature. *Acta neurochirurgica*, *154*, 257-265.
5. Kosugi S, Kume M, Sato J, Sakuma I, Moroi J, Izumi K, Sato Y, Nakamura N, Takahashi M & Miura I: Diffuse large B-cell lymphoma with mass lesions of skull vault and ileocecum. J Clin Exp Hematop 53: 215–219.2013.
6. Martin, J., Ramesh, A., Kamaludeen, M., Ganesh, K., & Martin, J. J. (2012). Primary non-Hodgkin's lymphoma of the scalp and cranial vault. *Case Reports in Neurological Medicine*, *2012*, 616813.
7. Duyndam, D. A. C., Biesma, D. H., & Van Heesewijk, J. P. M. (2002). Primary non-Hodgkin's lymphoma of the cranial vault; MRI features before and after treatment. *Clinical Radiology*, *57*(10), 948-950.
8. Parker, F., & Jackson, H. (1939). Primary reticulum cell sarcoma of bone. Surg Gynecol Obstet, 68, 45-53.
9. Dai, M. S., Ho, C. L., Chen, C. Y., Chen, T. M., Yu, C. P., & Chao, T. Y. (2000). Lymphoma of bone with initial presentation as a calvarial mass. *Annals of hematology*, *79*(12), 700-702.
10. Fadoukhair, Z., Lalya, I., Amzerin, M., Elkhanoussi, B., Sbitti, Y., Boutayeb, S., ... & Errihani, H. (2011). Successful management of primary non Hodgkins lymphoma of the cranial vault. *Pan African Medical Journal*, *8*(1), 50.
11. Mohindra, S., Gupta, R., Sharma, T., Das, A., & Radotra, B. D. (2007). Calvarial lymphoma. *British Journal of Neurosurgery*, *21*(6), 629-630.
12. Maruyama, D., Watanabe, T., Beppu, Y., Kobayashi, Y., Kim, S. W., Tanimoto, K., ... & Tobinai, K. (2007). Primary bone lymphoma: a new and detailed characterization of 28 patients in a single-institution study. *Japanese journal of clinical oncology*, *37*(3), 216-223.