Case Report

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Primary Diffuse Large B Cell Lymphoma of the Cranial Vault: A Rare Entity

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Abstract

Primary Non-Hodgkin's Lymphoma of cranial vault is a rare entity. We came across an interesting case of a 30 year old immunocompetent male who presented to us with a scalp swelling in the left parietal region. Computed tomography (CT) and MRI (Magnetic resonance imaging) of head showed contrast-enhancing lesion in left parietal region. CT scan of patient's abdomen, chest and pelvis did not reveal any lymphadenopathy or organomegaly. After excision, the histopathology and immunohistochemistry of the lesion confirmed the diagnosis of Diffuse Large B cell Lymphoma. Patient was further treated with chemotherapy and radiotherapy with complete resolution of the lesion. The rarity of the condition with a good response to treatment combining surgery, chemotherapy and radiotherapy prompted us to report this case.

Keywords: Primary Diffuse Large B-Cell Lymphoma; Cranial Vault; Chemotherapy; Immuno-Histochemistry.

Introduction

Non-Hodgkin lymphoma represents only about 3%-4% of all lymphomas in general population. Bone involvement is very uncommon and few cases involving femur, tibia and pelvis have been reported. Primary involvement of cranial vault has been rarely reported, and to the best of our knowledge only fourteen immunocompetent patients with primary cranial vault lymphoma have been reported in the literature [1-5]. We report a case of cranial vault lymphoma in a young immunocompetent male patient who presented to us as with a left parietal scalp swelling.

Case Report

A 30 year old immunocompetent male patient presented with a 6 month history of a painless mass involving the left parietal region. There was no history of trauma to head, fever or any other systemic symptoms (weight loss, loss of appetite etc). His general physical examination did not reveal any pallor, lymph node enlargement, bony tenderness or hepatospleenomegaly. Local examination revealed a 10x8 cm firm mass in left parietal region which was non-tender, non-movable, and fixed to underlying skull [Figure 1]. The overlying skin was normal. The patient's neurological examination including higher mental function was unremarkable.

Non-contrast CT of the head revealed a 10x8 cm hyper dense mass arising from calvaria and extending both intra and extracranially with bone destruction. On administration of contrast the lesion showed homogenous enhancement. His MRI revealed that the lesion was isointense to hypointense on T1 and T2 and was enhancing well on gadolinium administration [Figure 2]. In view of these findings, a preoperative diagnosis of meningioma with transcalvarial extension was considered. The patient underwent craniotomy and total excision of the lesion was done. Both extracranial and intracranial part of the tumour along with involved bone was removed. The tumour was grayish white, mildly vascular that was easily dissected from surrounding tissue. There was diffuse involvement of dura and the involved dura was also excised along with the tumor. There

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was no involvement of underlying brain. Post operative period was uneventful and a CECT scan on post operative day 1 revealed complete excision of the tumour [Figure 3].

Fig. 1: Figure showing scalp mass in left parietal region

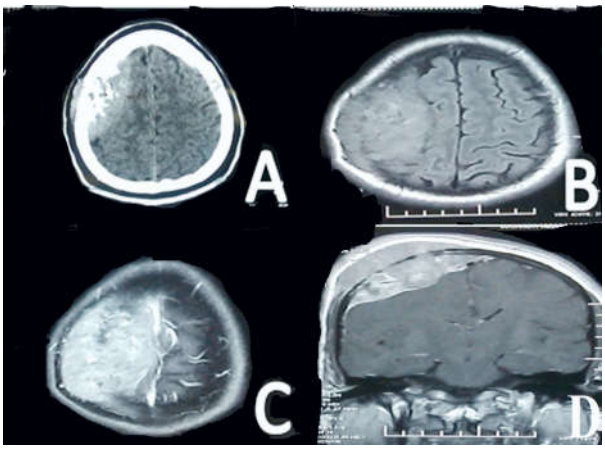

Fig. 2: A. Plain CT showing hyperdense lesion with bony destruction in left frontoparietal region. B. T1W image showing isointense lesion. C. T1 contrast image showing brilliant contrast uptake. D. T1 coronal image showing transcalvarial extension of the lesion.

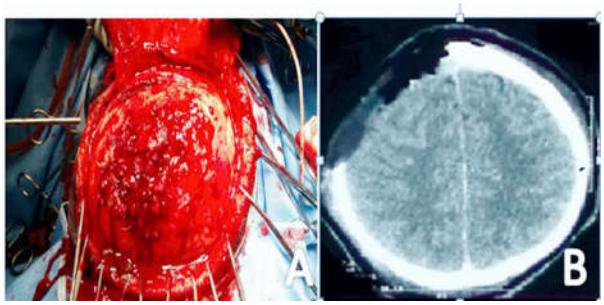

Fig. 3: A. Per operative photograph showing bony destruction by tumour. B. Post operative CT showing excision of tumour along with overlying bone.

Histopathology

The histopathological examination of the specimen showed diffuse infiltration of bone and dura by atypical medium to large lymphoid cells with cells displaying high nucleocytoplasmic ratio and hyper chromatic nuclei, inconspicuous nucleoli and scant cytoplasm.The proliferation fraction, as detected by Ki67 was high 78%. On immuno-histochemistry, tumour cells displayed diffuse cytoplasmic expression of CD45 along with CD20 and were negative for GFAP and Synaptophysin [Figure 4]. Based on results of immunohistochemistry, morphological diagnosis of diffuse large B-cell lymphoma of cranial vault was established.

In view of the histopathology and immuno-histochemistry report, the patient was further investigated. His Hb was 11.0gm%, TLC was 8400/ mm^3 and DLC was N-62%, L-37% and E-1%. His HIV, HB_SAg and HCV test was negative. The CSF cytology was negative for malignant cells. CT scans of the abdomen, chest and pelvis did not pick any significant lymph node enlargement or hepatosplenomegaly.

The patient was then referred to haemato-oncology department, where he received four cycles of chemotherapy with CHOP (cyclophosphamide, adriamycin, vincristine and prednisolone). Twenty four months after the follow-up the patient is doing well, without any signs of recurrence or dissemination.

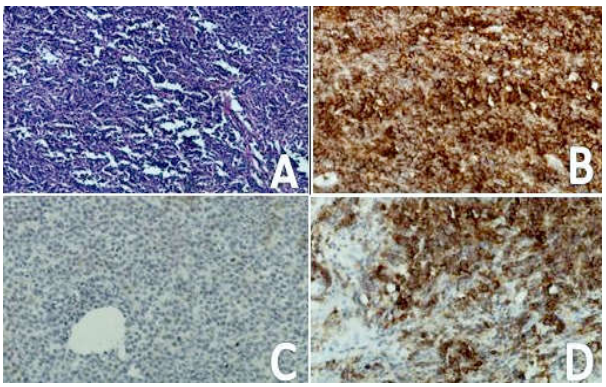

Fig. 4: A. H&E 10X-Diffuse infiltration by small round cells with high nucleo-cytoplasmic ratio. B. Diffuse membrane positivity for LCA(CD45). C. Tumour cells showing GFAP negative. D. Tumour cell showing membrane positivity for CD20.

Discussion

Primary bone lymphoma is very rare and was first described by Oberling in 1928. Cranial vault lymphoma is often associated with intra and extra cranial extension with intervening bone destruction. The most common clinical manifestations of calvarial lymphomas are painless scalp masses [10,11] headaches [10],convulsions [12], or focal neurological deficit [12]. Our patient also presented with painless scalp mass without any neurological deficit.They have male preponderance, and a median age of presentation in fifth or sixth decade.

Involvement of cranial vault is an unusual manifestation of Non-Hodgkin's lymphoma in patients without HIV infection. Our patient was HIV negative and presented at younger age in comparison to the literature.

MRI in lymphomas is non-specific and shows variable signal intensities on T1 and T2WI, and shows good enhancement on contrast administration. Differential diagnosis includes metastasis, meningioma, osteomyelitis, primary skull tumours and, haemangiopericytoma.

In early stages of cranial vault lymphoma, bony destruction is minimal but gradually extend to involve the bone completely [13,14,15]. Our patient also showed extensive bony destruction ,suggesting the lesion was in advanced stage. The tumour grows rapidly to involve dura and brain parenchyma, so early diagnosis is necessary. Besides, to confirm a diagnosis of cranial vault lymphoma, an IHC is mandatory. Majority of cranial vault lymphomas are of B-cell origin, as in our case [10]. Chemotherapy and radiotherapy are the treatment modalities for the calvarial lymphomas, but no standard protocol exists. The CHOP regimen was among the first combinations to produce complete response and long-term survival. For patients with advanced diffuse large B-Cell lymphoma, addition of a monoclonal antibody against CD20 (rituximab) to CHOP is beneficial. Our patient showed a good clinical outcome with complete response to a combination of sequential chemotherapy and radiotherapy.

Conclusion

Although extremely uncommon, cranial vault lymphoma should be kept in differential diagnosis of patients who presented with a scalp swelling. Various clinical presentation exists ranging from solitary skull lesion to diffuse lesion with or without focal neurological deficit. Chemotherapy followed by involved field radiotherapy, appears to be an established treatment. Generous reporting of such type of cases help us in better understanding of such a rare entity.

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