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# PRIMARY SKULL VAULT LYMPHOMA-A RARE PRESENTATION OF SCALP MASS : A CASE REPORT.

**Radiodiagnosis**

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## ABSTRACT

Primary skull vault lymphoma is a rare malignant bone tumor involving the skull vault presenting usually as a growing scalp mass with no systemic dissemination involving the skull bone. It usually missed as initial diagnosis in a scalp mass therefore it should be considered as differential of rapidly growing scalp mass. We present a case report of a 45 year old male with non-tender scalp swelling showing MRI features of an extra-axial and extraclavarial tumor involving the skull bone which on histopatholohy and IHC was confirmed to be primary skull vault lymphoma.

## KEYWORDS

Primary lymphoma of the bone (PLB), Primary Cutaneous B cell lymphoma (PCL), Non-Hodgkins lymphoma (NHL).

## INTRODUCTION:

Non-Hodgkin lymphomas represent only 3% to 4% of all neoplasms in the general population. [1]. Primary lymphoma of the bone (PLB) is uncommon, accounting for only 7% of all malignant bone tumours and less than 1% of all non-Hodgkin's lymphomas, and the majority of them involve the pelvis or limb bones [2-3]. Primary cranial vault involvement is rarely reported [4]. Here we report a case of a cranial vault lymphoma that was found as a frontal scalp mass in an elderly man.

## CASE REPORT:

A 45 years male patient came with the chief complaint of gradual swelling over scalp in the anterior aspect since past 2-3 months with intermittent on and off headache. There was no history of any trauma, fever, ear discharge, TB, syphilis or HIV. Physical examination revealed a non-tender, non-fluctuant subcutaneous scalp lump. There were no enlarged axillary or cervical lymph nodes noted. There were no neurological signs and symptoms. Also the Liver and spleen were within the normal limits. All the routine blood investigations were within normal limits.

The patient underwent MRI Brain with Gadolinium enhanced contrast scan and limited CT cuts were taken, which revealed:- An intracranial ,extra-axial , moderately enhancing, T2 heterogeneously hyperintense and T1 iso-hypointense lesion (measuring approx.10.0 x 6.5cm) was noted in bilateral frontal regions with involvement of entire frontal bone, anterior parts of parietal bones, frontal sinus walls & roof of orbits with large extracalvarial enhancing soft tissue component (measuring approx.11.6 x 8.7 x 2.0 cm) noted. There was an intraorbital extraconal extension of the soft tissue in both the orbits. Diffusion restriction was noted in the intracranial as well as in the extracalvarial components. The Intracranial component was causing smooth indentation over underlying bilateral frontal lobes with no obvious intra-parenchymal extension. The intraorbital component was causing indentation over bilateral superior rectus and superior oblique muscles and over right eye globe. Optic nerves, retro-bulbar spaces and rest of the extraocular muscles were normal. Based on these findings following differential were given–1.Granulocytic sarcoma (Extra-medullary leukemic tumour)/ Extramedullary hematopoiesis. 2, Atypical En plaque meningioma

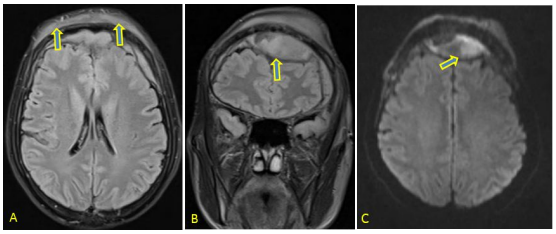


**Figure1. Pre-contrast MRI Brain.**

A: Axial T2/FLAIR WI shows an isointense extraclavarial mass lesion in the bilateral frontal regions (arrow).
B: Coronal T2/FLAIR WI shows the extra-axial component of the lesion is indenting the underlying parenchyma (arrow) with no obvious intraparenchymal extension.
C: DWI (Diffusion weighted Image) shows restriction (arrow) in the extraaxial component.

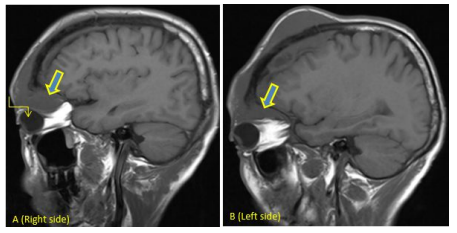


**Figure2. Pre-contrast MRI Brain.**

A & B: The sagittal T1 WI shows the intraorbital extension (arrow) which is causing indentation over bilateral superior rectus and superior oblique muscles and also over right eye globe (curve arrow in Figure A).

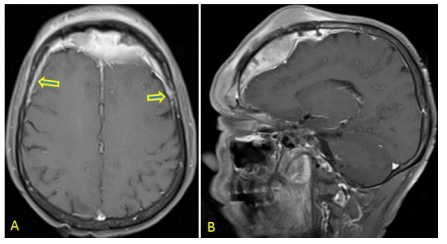


**Figure3. Post-contrast MRI Brain.** A, Axial and B, Sagittal T1 FS Images shows diffuse enhancement in the tumor lesion with enhancement of the adjacent dura (arrow).

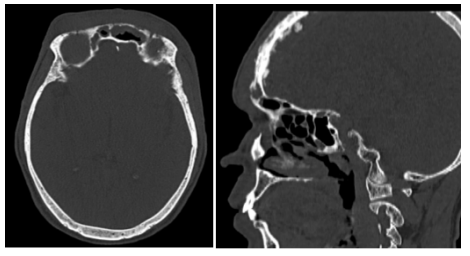

**Figure4. Computed tomography (CT) imaging. A:** Axial B: Sagittal bone window shows, ill-defined borders with osteolytic and sclerotic changes of the underlying frontal bone and also of the roof of the orbits.

After the scan the patient was subjected to biopsy. The initial biopsy report from the cranial lesion was reported as a round cell tumor. A repeat biopsy with IHC from swelling on the scalp revealed histology of NHL, diffuse large B-cell type with immunohistofluorescence showed that the lymphoma cells were positive for CD20 and CD79a but negative for CD30 suggesting a diffuse large B-cell lymphoma. Ki-67 staining was positive in over 70-80% of nuclei.

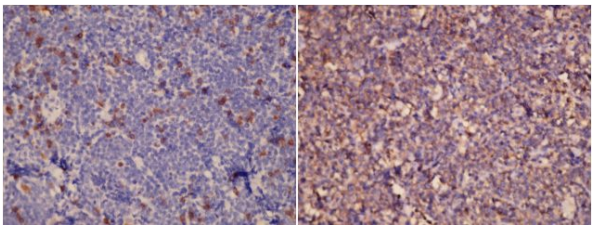

**Figure6. Immunohistofluorescence and Ki-67 staining.**

## DISCUSSION-

A true primary malignant lymphoma of the bone is defined as a solitary mass lesion with no evidence of disease at others sites and no systemic dissemination within 6 months of detection of tumor [5, 6]. The diagnostic criteria of PLB are 1) a primary focus in a single bone, 2) a positive histological diagnosis, 3) metastasis to only regional areas on presentation or the primary lesion preceding metastasis [3]. This patient had all of these diagnostic criteria. Causes of PLB have been suggested to be inflammation, trauma, or viral infections, but our patient had revealed any such specific history.

Although incidence of primary skull lymphoma is very rare, its possibility must be kept in mind in the differential diagnosis of primary skull lesions, irrespective of immune status. Patients can present in various manners ranging from solitary skull lesion to diffuse lesion with or without focal neurological deficit. In our case the patient presented only with the scalp mass and no neurological complaint. The most common clinical manifestations of calvarial lymphomas are painless scalp masses [3, 7, 8], headaches [7, 8], convulsions [2], or focal neurologic signs [2]. But these symptoms and signs are nonspecific, so further radiological and histological studies are needed for a conclusive diagnosis.

Most primary cutaneous B-cell lymphomas (PCL) have been reported involving the head & neck region and therefore PCL of the scalp need to be differentiated from primary malignant non-Hodgkin's lymphoma of cranial vault which are more frequently associated with intracranial (extradural) extension [9]. The main clinico-radiological differences between PCL of the scalp and primary NHL of the cranial vault include a shorter duration of symptomatology and early onset of focal neurological deficits, large soft-tissue mass and extensive osteolytic lesions in primary NHL of the cranial vault whereas PCL runs an indolent course. Other important clinicopathologic and radiological differential diagnoses of the scalp masses include protruding tumorous angiolymphoid hyperplasia with eosinophilia (ALHE), Kimura's disease of the scalp, pseudolymphoma, cranial vault meningioma, osteomyelitis, turban tumor, and metastasis.[10,11]

Primary lymphoma of the cranial vault presenting as a scalp mass can easily be missed at the initial diagnosis because of its extreme rarity. However, because of its rapid growth and invasion to the brain, an early diagnosis and active treatment are very important. During the early stages of calvarial lymphoma, bony change is minimal but gradually extends outward and finally destroys the bone completely [4]. Our patient showed mild bony changes, suggesting that the lesion was in an early stage. The tumour grows rapidly and invades the dura and brain, so early diagnosis is important [4]. Chemotherapy and radiation therapy are the basic treatment modalities for primary calvarial lymphomas, but there are still no standard treatment protocols [4]. Surgery followed by local radiation is the preferred treatment for a single lesion of extra nodal disease, but if the lesion is completely removed, radiological follow up without additional treatment may be another option [2].

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