PRIMARY MALIGNANT NON-HODGKIN'S LYMPHOMA OF CRANIAL VAULT: A CASE REPORT

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Abstract:

Primary Non-Hodgkin's Lymphoma of the scalp and cranial vault is a rare disease. We are describing a case of the same in a 70-year-old lady who presented with a rapidly enlarging diffuse swelling in the right frontoparietal region of scalp for 2 months. Imaging showed diffuse infiltration of the skull vault with extra and intra cranial soft tissue masses. FNAC and biopsy of the scalp mass showed B-cell Non Hodgkin's lymphoma. Further investigations did not reveal any other evidence of systemic lymphoma. Till 2010 only seventeen cases were reported ,among which 10 cases were purely primary skull vault lymphoma . We are reporting this case due to its aggressiveness and rarity.

Key Words: *Non Hodgkin's Lymphoma(NHL), Skull vault*

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Introduction

Non Hodgkin lymphoma (NHL) is an uncommon neoplasm with an incidence of only 3% to 4% of prevalence in general population. It occurs more frequently in patients with acquired immunodeficiency syndrome (AIDS). Primary involvement of the central nervous system (CNS) occurs only in 1% to 2% of patients with lymphoma (1). NHL originating primarily in bones is rare, occurring only in 3-4% of patients with lymphomas (1), with a predilection for the long bones of the upper and lower extremities, the pelvis and the spine. NHL presenting with primary involvement of cranial vault and dura mater is an unusual event and have rarely been reported in literature. Out of 17 cases reported, 7 cases were associated with systemic disease on presentation (2). In the other 10 cases, the disease was restricted to the cranial vault and these cases were therefore described as primary cranial vault lymphomas.

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Case Presentation:

A 70 year old female presented with a painless, rapidly growing swelling of scalp just behind the forehead on right side and over vertex for last two months. She had a history of minor trauma over the same location one & half yrs back. The patient was old and debilitated but didn't give any history of recent weight loss, cough, chest pain or haemoptysis. Despite occasional headache she didn't give any history related to raised ICP or seizure. She was not suffering from any other infectious or non infectious co morbidities. On examination, there was a lump over right fronto-parietal region crossing the midline about 10 cm x 8 cm size with irregular surface, non-tender, non-fluctuant, fixed to the overlying skin or underlying structure. (Figure 1) A small tiny reddish nodule was present over skin of forehead. No palpable lymph node was found in cervical chain, axilla or anywhere in the body.

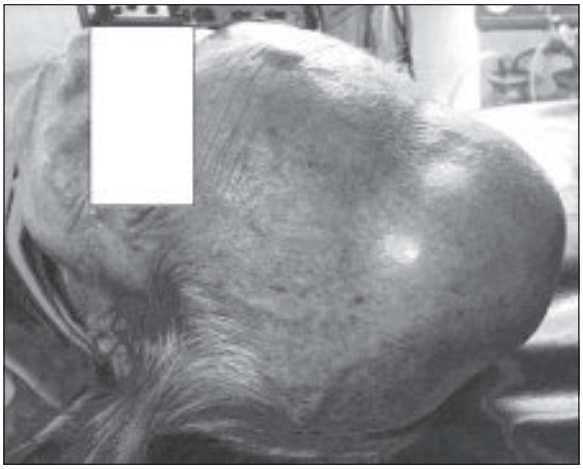

Fig.-1: *Pre operative picture of the patient showing the skull lesion.*

CT Scan (Figure 2) & MRI of Brain (Figure 3) revealed osteolytic lesion with external soft tissue mass in the right fronto-parietal region. Mild enhancement of soft tissue is seen after I/V contrast. MRV (Figure 4) shows that anterior part of surperior sagittal sinus was compressed but there was no tumor infiltration within the sinus. FNAC of the mass showed features, consistent with Non-Hodgkin's Lymphoma.

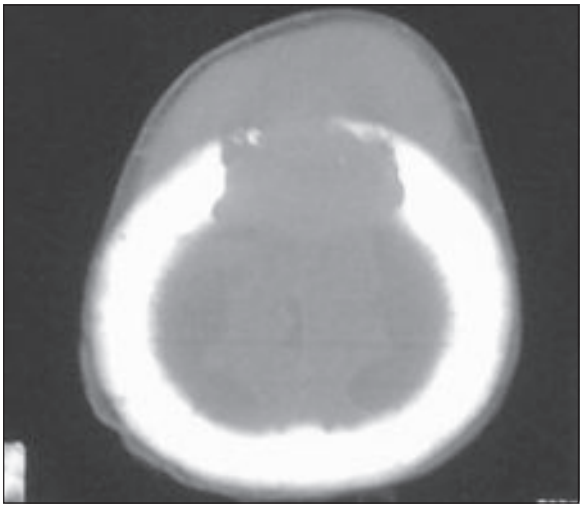

Fig.-2: *Pre operative CT scan of head showing erosion of skull bone with tumor extension.*

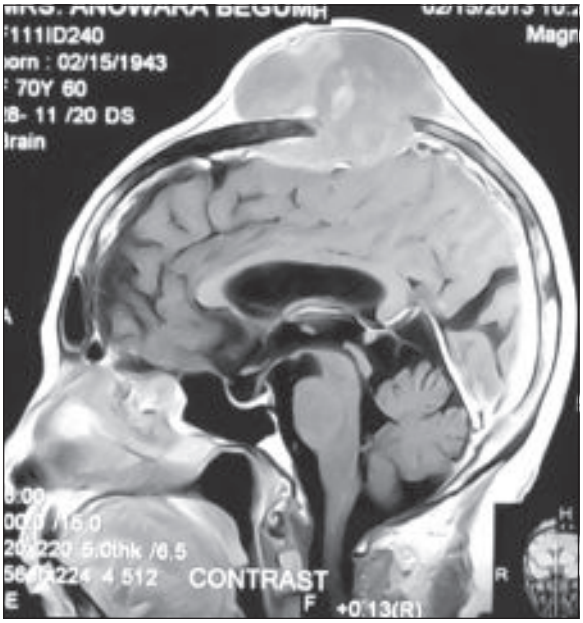

Fig.-3: *Contrast MRI of brain showing both intra and extra cranial extension of tumor with dural attachment.*

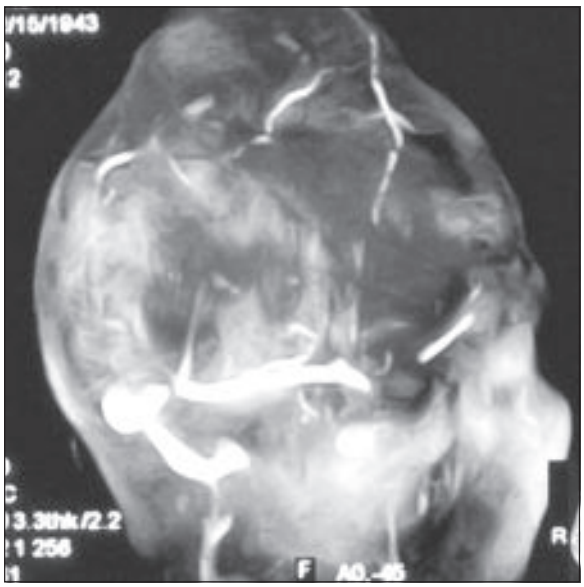

Fig.-4: *MRV of brain showing compression of the superior sagittal sinus.*

Under general anaesthesia, with the patient in supine position, a midline linear incision was given over the swelling and the tumor was explored. A moderately vascular and firm tumor was found, which was free from skin but was adherent to the outer surface of the dura eroding the frontal bone. (Figure 5) The margin of the bony gap was thin and friable. The tumor was totally dissected out from dural surface by bipolar coagulation. The superior sagittal sinus was

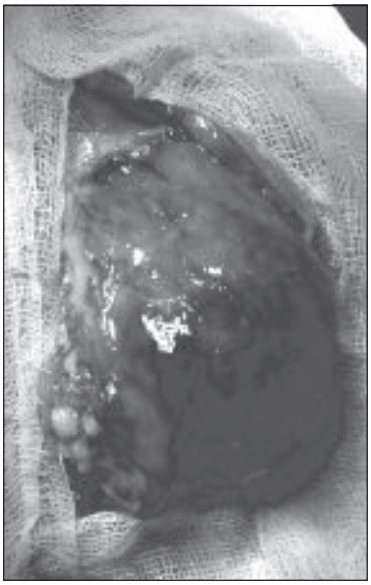

Fig.-5: *Per operative view of the tumor just after exposure.*

compressed but free from tumor invasion. The unhealthy bony margins were removed circumferentially till macroscopic healthy margin was reached. Cranioplasty was done.

Histopathology was suggestive of Non-Hodgkin's lymphoma (diffuse large cell type).

The post-operative period was uneventful. Chemotherapy (CHOP: Cyclophosphamide, Hydroxydaunorubicin, Oncovin, Prednisosolone) followed by radiotherapy was planned following consultation with an oncologist. Prior to the scheduled time, on the 11th post-operative day, it appeared that, the pre-existing tiny reddish nodule over the forehead about the size of a rice grain which rapidly started to increase in size. The tiny nodule assumed a size of about 4 cm x 4 cm within one week of time. (Figure 6) Chemotherapy was started immediately and the lesion disappeared after about 1 week. At 3 weeks interval, 3 cycles of chemotherapy was given but the patient became so debilitated that she could not tolerate further treatment and she succumbed to her illness in a few days' time.

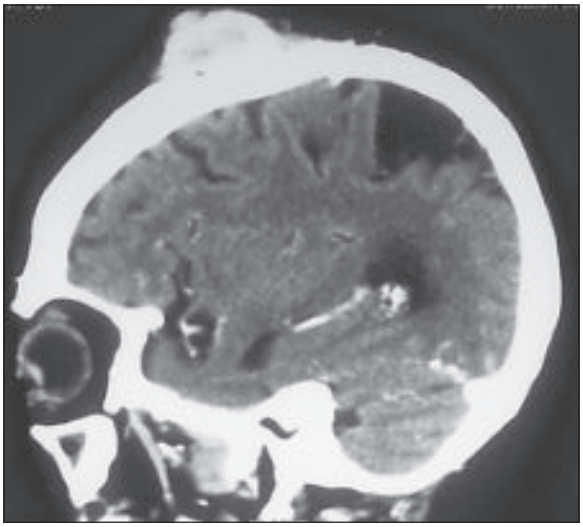

Fig.-6: *Post operative contrast CT scan showing flare up of preexisting smaller skin nodule.*

Discussion:

Primary lymphoma involving the cranial vault, pericranium and dura are extremely rare[1-4].

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[1, 5, 6].

Radiologically, most primary lymphoma of bones are osteolytic and often permeative with rare cases of sclerosis[3]. There may be destruction of cortical bone. Pathologically, the spread of the disease to the meninges suggests that the lymphoma cells grow through the diploic spaces along the emissary veins [1,6,7]. In our case, it seems that, the tumor originated from skull bone and by erosion of bone it extended both extra and intracranially.

The treatment of primary non-Hodgkin lymphoma of bone is local radiotherapy and chemotheraphy (8). We started chemotherapy earlier to prevent systemic dissemination of the disease.

The prognosis of lymphoma involving the cranial vault is uncertain but any involvement of cerebral structures by direct invasion or by leptomeningeal seeding is associated with less favourable prognosis. Clinical suspicion, early diagnosis and aggressive treatment preferably with both chemotherapy and radiotherapy are indicated (1). In the present case it seems that, though the tumor was attached with dura , but post operatively the preexisting skin nodule flared up enormously.

Involvement of the cranial vault is an unusual manifestation of aggressive non-Hodgkin's lymphoma. The 5 and 10-year survival rate for multifocal bone lymphomas were 42% and 35% respectively (1, 8). In patients without HIV infection, it is usually a localized disease. Ten patients with non-HIV related primary cranial vault lymphoma have been reported in the literature (1-9). Complete remission was achieved in 8 of 10 patients with median remission duration of 7+ months (range, 1-72+). Three of these patients were still disease free after 2 years. Of the 10 patients, 5 received whole brain radiation alone, 4 received systemic chemotherapy plus whole brain radiation, and one received whole brain radiation and intrathecal methotrexate. Complete remissions were achieved in 3/5, 4/4, 1/1 and patients respectively (9). In comparison to previous cases our case seems to be more aggressive because few days after surgery, preexisting skin nodule became a big mass which responded well with chemotherapy and disappeared.

While combined modality treatment including anthracycline-based systemic chemotherapy and involved field radiation therapy is considered the standard for localized aggressive non-Hodgkin's lymphoma, these reports suggest that durable

remission of primary cranial vault lymphoma can be achieved by radiation therapy alone. In patients with primary cranial vault lymphoma who are unlikely to tolerate systemic chemotherapy, single modality treatment with radiation is a reasonable initial therapy (9). Due to aggressiveness , we started chemotherapy initially after surgery and it was planned to start radiotherapy later on . But we failed to give RT as patient passed away. We recommend neoadjuvent CHOP therapy followed by adjuvant CHOP and radiotherapy in more aggressive cases.

Conclusion:

Primary Non Hodgkin's Lymphoma is a rare but aggressive disease. Early diagnosis and aggressive management with surgery, radiotherapy and chemotherapy is therefore important.

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