Primary Diffuse Non-Hodgkin’s Lymphoma of the Scalp, Cranial Vault with Intracranial Extension- A Case Report

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Abstract:
Primary Non Hodgkin’s lymphoma (NHL) of the cranium with extra and intracranial extension without systemic or skeletal manifestation in a non immunocompromised patient is extremely rare. These lesions are most of the time misdiagnosed because they mimic other conditions like meningioma. Here we report a case in a 38 years old man. He presented to us with lobulated bulky scalp mass over bilateral frontoparietal region more over right side for 6 months. It was progressively enlarging in size. CT scan of Brain with Contrast Imaging showed involvement of scalp, cranial vault, meninges and the brain parenchyma, mimicking a meningioma. After total resection and biopsy of the scalp masses showed an intermediate grade of diffuse Non-Hodgkin’s lymphoma.

Key words: Meningioma, non Hodgkin’s lymphoma, scalp mass.


Introduction:
Primary extra lymph node lymphomas are not uncommon, but they often cause difficulty in diagnosis. Primary lymphoma arising from the cranial vault can mimic clinically and radiologically a meningioma.¹,² We present a case of lymphoma arising from the cranial vault mimicking as a meningioma and also review the literature.

Case Report:
A 38 years old man presented with progressively increasing unsightly lobulated bulky scalp mass over bilateral frontoparietal region more over right side with a linear healed scar mark over centre of the forehead for 6 months duration. He had occasionally mild headache but no other signs and symptoms. There was no history of trauma, fever, or any other illness in the past. But he had a history of surgery with biopsy that revealed nothing significant. After that he referred to Neurosurgeons. On examination, he was conscious and without any neurological deficit. There was non tender lobulated bulky mass over bilateral frontoparietal region, with a firm, ill defined, and bosselated surface. There was no local rise of temperature, and skin over it was healthy [Figure 1]. His blood investigations were normal. X ray chest, ultrasonography of the abdomen, and peripheral smear were normal. CT scan of brain with contrast

Fig.-1: Bulky lobulated scalp mass over bilateral frontoparietal region of scalp
showed large enhancing destructive lesion of bilateral frontoparietal bone with overlying scalp mass and intracranial extradural and intradural components infiltrating superior sagittal sinus causing mass effect and edema of underlying brain parenchyma and effaced frontal horn of lateral ventricle [Figure 2, 3, 4, 5]. For inconvenience CT angiography and MRV could not be done. He underwent bifrontal craniotomy and total excision of the lesion was done. Involved bone was removed. Dura was opened that expose the intradural portion of tumor. Proximal third of Superior Sagittal Sinus found invaded by the tumor that was ligated and removed with Dura and intradural portion of tumor. Duroplasty done by temporal fascia. Tumor was moderately vascular, fleshy in consistency, and infiltrating the scalp, bone, and brain parenchyma [Figure 6, 7]. Postoperatively on 5th POD patient develop CSF leak. Lumbar drain was given but not improving. Twenty days later reexploration done. Dural defect was detected and duroplasty again done with artificial Dura (G-patch) [Figure 8]. Drain tube keep in situ that was removed on 8th POD. CSF leak stopped. Histopathology revealed a diffuse intermediate grade Non-Hodgkin’s lymphoma (NHL). Post operative CT scan of brain was done that revealed clearance of tumor [Figure 9].

Fig.-2: CT scan of brain with contrast showed large enhancing lesion with extradural and intradural components causing mass effect and edema of underlying brain parenchyma and effaced frontal horn of lateral ventricle

Fig.-3: Axial and coronal view of tumor
**Fig.-4:** Contrast CT scan of brain showing the Tumor with intra and extra cranial extension

**Fig.-5:** Bony window showing extensive bony erosion of frontal

**Fig.-6:** Pre operative photo (a) fleshy tumor with moderate vascularity (b) infiltration of scalp and bone (c) Involved bone removed by craniotomy (d) Intradural part of tumor with involved SSS
Fig.-7: (a) After Dura resection with proximal third of SSS (b) Duroplasty (c) Cranioplasty (d) after skin closure.

Fig.-8: In re-exploration Dural defect detected and CSF leak repaired by artificial Dura.
Discussion:
NHL represents only 3%-4% of all neoplasms in the general population and it occurs more frequently in patients with AIDS. Direct involvement of the CNS occurs only in 1%-2% of patients with lymphoma.2-5 These lesions are commonly reported in the seventh and eighth decades.2,6-9 But our patient present at young age (38 years of age) in comparison to the literature. The incidence of CNS lymphoma has increased in both immunocompromised and immunocompetent persons.3 Malignant lymphoma originating from the skull may initially extend outside the cranium, extension within the cranium occurring only subsequently. The initial symptoms and signs of lymphoma in the skull include a painless scalp lump, headache due to bone destruction or tumor infiltration of meninges, seizures, and focal neurological deficits resulting from infiltration of the cortex.6,8,10 In our case, painless subcutaneous scalp mass was not associated with any neurological deficit.9 The disease can involve the pericranium, underlying meninges, and subcutaneous tissue. Pathologically, the spread of the disease to the meninges suggests that the lymphoma cells grow through the diploic spaces along the emissary veins and nerves that pass through the Dura to the leptomeninges. Because of the characteristic permeating growth pattern of lymphoma, there was large soft-tissue component along with bone destruction in our case.6,8,10 CT scan head generally shows extra-intracranial extent, bone and Dura mater invasion.7 On plain CT scan and MRI, these lesions are isodense and isointense in nature, respectively, which enhance after contrast administration.2,9,11 MRI is helpful in showing diffuse primary cutaneous lymphoma of the cranial vault with orbital and brain invasion, and can thus aid in the decision making regarding different treatment strategies by revealing the invasion of tumor.2,11 The angiographic findings of these lesions include mild vascularity in the periphery of the tumor and displacement of neural and vascular structures unlike meningiomas.2 When there is diffuse vault, meningeal and parenchymal infiltration, an intraoperative frozen section is recommended since the identification of a lymphoma is likely to influence the decision about the extent of the surgical excision.2,6 These lesions are effectively treatable by surgery and radiotherapy, with a good outcome in most of the cases.10,12,13 This can be followed by systemic chemotherapy with cyclophosphamide, vincristine, and prednisolone (CVP).7 Present case was managed by wide excision of tumor and involved bone. The prognosis of a lymphoma appearing in the skull vault is uncertain, but any involvement of the cerebral structures by direct invasion or by leptomeningeal seeding indicates a less favorable prognosis.14,15

Conclusion:
Although this condition is rare, NHL diagnosis must be considered in the differential diagnosis in any patient with a scalp mass extending through the skull.

References:
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