Frontal convexity primary lymphoma masquerading meningioma: A case report and review of literature

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Abstract

Primary non-Hodgkin’s lymphoma of the skull with extra- and intracranial extension without systemic or skeletal manifestation in a nonimmunocompromised patient is extremely rare. These lesions often cause difficulty in diagnosis because they mimic other conditions. We report a case of primary lymphoma involving scalp, skull vault, meninges and invading the brain parenchyma and masquerading clinically and radiologically as a meningioma.

Key words: Lymphoma, meningioma, nonHodgkin lymphoma

Introduction

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

Case Report

A 43-year-old female patient presented with progressively increasing swelling over left frontal region of eight months duration. It was occasionally associated with pain. She had no other signs or symptoms. Local examination revealed a 4 x 4 cm, firm, ill-defined, smooth surface, nontender swelling over left frontal paramedian region. There was no local rise of temperature and skin over it was healthy. Her general and systemic examination revealed no other abnormalities. Her blood investigations were normal except mildly raised ESR (14 mm/Hr). Investigations for HIV infection were negative. X-ray chest, ultrasonography of the abdomen, peripheral smear and bone marrow examination were negative for lymphoma. CT scan head showed a lesion with intra and extracranial extension with thickening of bone. The lesion was enhancing with contrast. All these features were in favour of the diagnosis of meningioma [Figure 1]. She underwent bifrontal craniotomy and total excision of the lesion including the involved bone. Tumor was moderately vascular, fleshy in consistency and infiltrating the scalp, bone and brain parenchyma. Histopathology revealed a diffuse, large cell type of lymphoma. There were focal subcutaneous infiltrates, composed of large cells with oval to irregular nuclei and vesicular chromatin. There

Figure 1: Axial CT images showing hyperdense lesion on plain scan (left) left frontal region with extra-cranial extension enhancing with contrast and associated perilesional oedema and mass effect
was no destruction of bone. All these features were suggestive of non Hodgkin lymphoma (Maltoma type) without systemic involvement.

Discussion

NonHodgkin lymphoma represents only 3% to 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% to 2% of patients with lymphoma.[2-5] These lesions are commonly reported in the seventh and eighth decade.[2,6,9] Our patient was HIV negative and presented at young age in comparison to the literature. The clinical symptoms and signs of lymphoma in the skull include a painless scalp lump, headache due to bone destruction or infiltration of meninges with tumor, seizures and focal neurological deficits resulting from the cortical infiltration.[7,9,10] In our case painless subcutaneous scalp lump was not associated with any neurological deficits.[6] 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 with large soft-tissue component bone destruction may not be seen as in present case.[7,9,10] Cerebral computerized tomography (CT) scan will show extra-intracranial extent, bone and dura mater invasion.[8] On plain CT scan and magnetic resonance imaging these lesions are isodense in nature which enhances after contrast administration.[2,6,11] MR imaging 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.[12,11] 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.[2,7] These lesions are effectively treatable by surgery and radiotherapy with a good outcome in most cases.[10,12,13] This can be followed by systemic chemotherapy with cyclophosphamide, vincristine and prednisolone (CVP).[8] Present case was managed by wide excision of tumor and involved bone followed by radiotherapy with good response. 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

A high index of clinical suspicion, awareness of the characteristic CT features and aggressive therapy are required for these unusual cases. Although this condition is rare, the diagnosis must be considered in the differential diagnosis, in any patient with a scalp mass extending through the skull.

References


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