Primary midline cranial vault lymphoma simulating a parasagittal meningioma: The role of angiography in preoperative diagnosis

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Abstract
Primary non-Hodgkin's lymphoma (NHL) of the skull with extra- and intracranial extension without systemic or skeletal manifestation in a non-immunocompromised patient is extremely rare. Up to date, only nine such cases have been reported in the literature and in none was the lesion located in the midline. The authors report a unique case of a primary NHL involving the midline of the cranium. The lesion presented as a slowly growing scalp swelling mimicking a parasagittal meningioma. The angiographic findings of mild vascularity in the periphery of the tumor and downward displacement of a patent superior sagittal sinus indicated that the lesion was unlikely to be a meningioma. Neurosurgeons must maintain a broad differential diagnosis in any patient with a scalp mass eroding through the skull and associated neurological symptoms or signs. An intraoperative frozen section is recommended since the identification of a lymphoma is likely to influence the neurosurgeon's decision about the extent of the surgical excision.

Keywords: Non-Hodgkin's lymphoma, malignant lymphoma, scalp swelling, cranial vault, angiography.

1 Introduction
Primary central nervous system (CNS) lymphoma is a non-Hodgkin's lymphoma (NHL), mostly of B-cell origin [4]. It is rare in the non-immunocompromised patients, accounting for 0.85 % to 1.5 % of their intracranial tumors [10]. It may be solitary or multiple with a predilection for the basal ganglia, corpus callosum, thalami, and paraventricular region [14]. Primary involvement of the bone by NHL occurs in 3-4 % of cases [12]. However, in advanced stages of disseminated disease, the bone may be affected in 7-25 % of patients [3]. The most common skeletal locations of NHL are the spine, pelvis, rib, and long tubular bones, especially the lower extremities [5].

Initial presentation of a NHL in the skull vault with extra- and intracranial extension without systemic or skeletal manifestation in a nonimmunocompromised patient is extremely rare. Nine such cases have previously been reported in seven articles in the literature [1, 3, 5, 7, 9, 10, 12] and in none of these patients was the lesion located in the midline. The authors report a unique case in whom the primary NHL was in the midline of the cranium and presented as a slowly growing scalp swelling. The aim of the paper is to increase the awareness of the neurosurgeon of the need to maintain a broad differential diagnosis in any patient with a scalp mass eroding through the skull and associated with neurological symptoms or signs and to discuss the role of angiography in the preoperative distinction between lymphoma and meningioma.

2 Case Report
A 25 year old male Saudi patient presented at the neurosurgical division at King Khalid University Hospital (KKUH) with a one year history of a slowly growing scalp swelling and two months history of headache. On examination, the patient had a firm diffuse, non-tender swelling in the midline of the vertex measuring 5 cm in diameter. He was otherwise alert, and had no neurological deficits apart from bilateral papilloedema. Brain CT scan (Figure 1a, b) showed a large, parasagittal, contrastenhancing tumor in the parietal region. In addition to the large intracranial component, the tumor had eroded
the bone and invaded the subgaleal space producing a scalp swelling. Bilateral external and internal carotid angiography showed a slight vascularity at the periphery of the tumor which was supplied by the external carotid circulation. The superior sagittal sinus was displaced downward but remained patent (Figure 2).

At surgery, the tumor was found to be directly underneath the scalp. The lesion had originated from the skull bone and invaded the pericranium, epidural space, and dura. The inner surface of the dura was inspected and found normal with no evidence of tumor. The lesion was shaved off the dura and the outer surface of the dura was coagulated thoroughly. The diseased bone flap was removed and was replaced by a cranioplasty. Histopathology (Figure 3) revealed diffuse large sheets of highly malignant cells separated by fibroconnective tissue.

The majority of the malignant cells appeared as immunoblasts with focal plasmacytoid features. The immunohistochemical stain for leucocytic common antigen (LCA) was strongly positive while T-cell was focally positive, B-cell, S100 protein, and glial fibrillary acidic protein (GFAP) and cytokeratin were negative. The findings indicated a malignant lymphoma, large T-cell immunoblastic type.

The patient tolerated the operation very well and recovered without neurological deficits. A number of investigations were carried out postoperatively and found normal confirming that there was no evidence of immunosuppression or disseminated disease. These investigations were: HIV screening, abdominal CT scan and ultrasound, chest CT scan, bone scan, gallium whole body scan, bone marrow examination, CSF examination, and immunological assessment which included immunoglobulin and com-

Figure 1a, b. CT scan (I.V. contrast) (coronal view 1a, axial view 1b) show a large, contrast-enhancing, parasagittal lymphoma which erodes the bone and extends intracranially and into the subgaleal space producing a scalp swelling.

Figure 2. Right common carotid angiography shows a slight vascularity at the periphery of the tumor (arrow heads) supplied by the external carotid circulation. The superior sagittal sinus is displaced downward but remains patent.

Figure 3. Tumor biopsy shows a diffuse, large, T-cell malignant lymphoma predominated by immunoblasts (Hematoxylin and eosin X 100).
postoperative brain CT scan showed no evidence of residual tumor (Figure 4). The patient was treated with radiotherapy (5000 cGy in 25 sessions over 5 weeks). At a follow-up of 5 months, he had remained well with no evidence of a tumor recurrence.

3 Discussion

This is a case of a true primary malignant lymphoma of the cranium as the lesion was solitary with no evidence of disease at other sites and no systemic dissemination within 5 months of detection of tumor. Review of this case together with the previously reported nine cases [1, 3, 5, 7, 9, 10, 12] shows that primary NHL of the cranium may affect patients aged 20–73 years (median 57) with no sex dominance. The lesion may be right-sided [1, 3, 5, 9], left-sided [5, 7, 10, 12], or in the midline, as in our case. The most common presentation of this tumor is either as a painless scalp swelling that develops over a period of few days to 2 years [3, 5, 7, 10] or as headache related to the raised intracranial pressure, bone destruction, or meningeal infiltration [1, 7, 9, 10, 12]. Less commonly, patients may present with epilepsy [3, 5], and focal neurological deficits [3, 12] which result from either direct invasion of the cerebral cortex by the tumor [1, 3, 5] or local compression [12].

The differential diagnosis in a patient with a scalp swelling eroding the skull and extending intracranially is among meningioma, lymphoma, metastatic carcinoma, tuberculoma [6] and chronic osteomyelitis. The duration of history, extent of bone destruction, size of the soft tissue mass, and the presence of systemic signs may help to differentiate a lymphoma from a metastatic carcinoma or an inflammatory lesion. The difficulty is in the preoperative differentiation between a primary lymphoma of the midline of the cranium and the much more common tumor, a parasagittal meningioma. On the CT scan, the lymphoma shows a permeative growth pattern and a soft tissue component on both sides of the bone with limited destruction of cortical bone. The tumor is usually non-homogeneously hypodense, lacks calcification, and enhances faintly after contrast injection [3, 5]. A meningioma, on the other hand, stimulates adjacent bony thickening and sclerosis. It is usually isodense on the unenhanced scan, may be partly calcified, and enhances uniformly and brightly [12]. Angiography, which was not discussed in the previously reported cases, may also prove useful in the distinction between a midline lymphoma of the cranium and a parasagittal meningioma. Whereas a meningioma shows a homogenous blush that appears in the late arterial phase and persists into the late venous phase [2] a lymphoma is not usually hypervascular. However, as in our case, focal irregular and poorly demarcated vessel walls may appear in the midarterial phase on the periphery of the tumor [8]. In addition, a lymphoma of the midline of the cranium which originates in the bone, when it extends as a large mass into the epidural space, will displace the superior sagittal sinus downward, and since there is no underlying rigid structure to press against the sinus, it is likely to remain patent even if the tumor assumes a significant size. A large parasagittal meningioma, however, because of its dural origin is likely to invade or compress the superior sagittal sinus against the bone and occlude it.

There is no doubt that a preoperative distinction between a primary lymphoma of the cranium and a parasagittal meningioma is important. A preoperative assessment of the immunological function including HIV screening is essential only if a lymphoma is suspected. In addition, radical surgery is not necessary for a lymphoma because of its radiosensitivity. It is accepted, however, that there may be atypical appearances of meningioma [2] that make the preoperative differentiation between a meningioma and a lymphoma very difficult. It is, therefore, recommended that an intraoperative frozen section be obtained in all tumors to identify very rare tumors such as lymphoma during surgery.

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The lymphoma in the case reported here and in another [10] were classified as large immunoblastic T-cell NHL. It is of interest that the other reported primary NHL of the cranium were classified as diffuse small cleaved cell [1, 5, 7], large non-cleaved cell [3], large immunoblastic B-cell [9, 12], diffuse large cell [5], and diffuse mixed cell [3]. This suggests that the cranium can be the primary location of all of the histopathological subtypes of NHL.

It is generally accepted that primary lymphoma of bone is best treated by local radiotherapy [11]. Even though retrospective series documented improvement in the survival of patients receiving systemic chemotherapy following irradiation, the small sample sizes and the variable agents in these series preclude comment on the routine application of this approach. Our case and six of the previously reported cases [1, 3, 5, 9, 10, 12] were treated by radiotherapy, while only 5 cases [3, 5, 7, 10] received various regimes of chemotherapy.

The data on the outcome of patients with primary NHL of the cranium remains insufficient. Of the 9 reported cases, 3 patients were reported to be alive at a follow-up of 3–72 months [3, 5, 12], while 4 other patients survived a period ranging from 3–20 months [3, 5, 7, 10]. In 2 of the latter group of patients [3, 5], the tumor had invaded the cortex at presentation. The reported dissemination of primary NHL of the cranium after treatment was neuraxial in two patients [3, 10] and skeletal in one [7]. The latter three patients survived a mean of 9 months after diagnosis. Thus leptomeningeal seeding, hematogenous spread of the tumor, and direct invasion of the cortex by the tumor indicate a poor prognosis. In CNS lymphoma [4, 13], the prognosis is also related to the preirradiation Karnofsky score, evidence of multiplicity tumor volume, and the histopathological features of the tumor. It is suggested that small cleaved and non-cleaved cell tumors are associated with longer survival than large cell tumors. Unfortunately at present, there is no sufficient data for us to comment on the importance of the histopathological features for the prognosis of patients with primary NHL of the cranium.

References


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