Primary diffuse large B-cell lymphoma of the dura mater and cranial vault

Case report and review of the literature

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Primary high-grade lymphoma of the dura mater and cranial vault has rarely been reported. The authors treated a 61-year-old man who presented with a slow-growing scalp mass that involved the cranial vertex. Magnetic resonance imaging revealed an oval mass of the dural type with peripheral edema in the bilateral parietal region, with attachment to the cranial vault and extension to the subgaleal space. After subtotal resection, pathological examination yielded a diagnosis of malignant large B-cell lymphoma. Twenty-three months postoperatively, after undergoing radiation therapy and chemotherapy, the patient is neurologically intact and without systemic dissemination of the malignancy. This is a case of primary malignant B-cell lymphoma of the dura mater with extensive involvement of the skull, which is a very rare event. Imaging-based diagnosis and combined therapy consisting of surgery, radiation therapy, and chemotherapy for the disease are discussed, and the literature on extraaxial malignant lymphomas is extensively reviewed.

KEY WORDS • B-cell lymphoma • extraaxial lymphoma • meninges • cranial vault

DI FFUSE large cell, mixed and immunoblastic lymphomas of B-cell origin can be considered together as aggressive lymphomas. Approximately 30% of cases originate in extranodal sites, particularly the gastrointestinal tract and the Waldeyer ring, but also in bone, skin, sinuses, eyes, ocular adnexa, gonads, the CNS, thyroid, and lungs. In unselected series of cases the incidence of primary CNS lymphomas in brain tumors varies from 0.3 to 1%, and the entity constitutes approximately 1% of all lymphomas. Within the CNS, these lymphomas have a predilection for the midline or para-ventricular structures. Primary extraaxial involvement of large cell type B lymphoma is infrequent, with the worldwide literature containing only a few cases. The precise anatomical location of the neoplasm reported here, without systemic manifestation, makes this case unique in its type.

Case Report

History and Examination. This 61-year-old man was admitted with generalized headaches, which had developed over a period of 3 months. His medical history was unremarkable. The patient’s physical examination demonstrated no focal neurological abnormality other than a palpable biparietal mass involving the scalp. Admission CT scanning revealed a high-density extradural mass in the superior sagittal sinus invading the vertex and subgaleal space that was homogeneously enhanced after contrast administration. In addition, CT scans with bone windows revealed involvement of both cranial tables without osteolysis. No cerebral edema was in evidence. On MR imaging, the mass measured 13 × 8 × 2 cm. It was slightly hypointense on T1-weighted and hyperintense to gray matter on T2-weighted images. Tumor enhancement after administration of gadolinium was homogeneous. Associated swelling of the pericranium and subcutaneous tissue involvement were observed (Fig. 1). Downward displacement of a patent superior sagittal sinus was present. Based on the location of the lesion, precontrast signal intensities, and an additional lesion outside the cranium, a metastatic tumor rather than a typical meningioma was suspected.

Operation. The large excrescent mass was resected through a right parietal craniotomy with contralateral extension. The tumor was gray, firm, relatively avascular, and densely attached to the dura mater from which it arose. The adjacent bone was severely infiltrated in both cranial tables, with continuous infiltration of the galea and subcutaneous fatty tissue. Our diagnosis (based on evaluation of frozen sections) was poorly differentiated malignant neoplasm that we strongly suspected was a lymphoma.

Postoperative Course. The patient’s postoperative course was uneventful and he recovered completely. Results of a radiographic skeletal survey were negative. Follow-up CT
scanning of the chest, abdomen, and pelvis demonstrated no evidence of pathological entities. Assessment of a bone marrow aspirate yielded normal results.

Pathological Findings. On microscopic examination, the neoplasm was seen to be composed of atypical, moderately large, mitotically active, occasionally cleaved, pleomorphic lymphoid cells. The neoplastic cells were infiltrating and destroying osseous tissue. Tumor immunophenotyping was done using flow cytometric analysis (CD5, CD10, CD19, and CD20) and by histochemical evaluation of the lymphoid cells. These exhibited strong immunoreactivity both to leukocyte common antigen and to B-cell marker (L26). The nuclear staining proliferation index with Ki 67 (MIB-1 marker) exceeded 90% of tumor cells. No staining was noted for chromogranin, synaptophysin, or the epithelial markers AE1 and AE3. The final neuropathological report confirmed the diagnosis of a high-grade meningeal diffuse large B-cell lymphoma with extensive involvement of a marginal zone (Fig. 2).

Adjuvant Therapy and Outcome. After the pathological findings were reviewed, the patient was treated with CHOP chemotherapy every 3 weeks for six cycles. After that, fractionated whole-brain radiation therapy was delivered in 18 fractions of 180 cGy each. After treatment, the patient remained free of neurological symptoms. Head CT scans and MR images obtained 4 months after surgery and adjuvant therapy revealed no residual tumor. After a follow-up duration of 23 months, the patient is neurologically intact and has no systemic dissemination of the malignancy.

Discussion

The medical literature offers a wide variety of reported cases of extraaxial primary CNS lymphomas. Neverthe-
Primary B-cell dural lymphoma

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs), Sex</th>
<th>Location of Lesion</th>
<th>Cranial Vault Involvement</th>
<th>Symptoms</th>
<th>Postop Treatment</th>
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<tbody>
<tr>
<td>Hollas et al., 1985</td>
<td>60, F (Case 1)</td>
<td>lt frontal</td>
<td>yes</td>
<td>seizures, scalp mass</td>
<td>RT</td>
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<td>Parekh et al., 1993</td>
<td>65, F</td>
<td>lt parietal</td>
<td>yes</td>
<td>scalp lump</td>
<td>RT</td>
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<tr>
<td>Sato et al., 1993</td>
<td>65, M</td>
<td>rt parietal</td>
<td>yes</td>
<td>scalp mass, hemiparesis</td>
<td>RT &amp; chemo</td>
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<tr>
<td>Landys et al., 1995</td>
<td>62, M</td>
<td>frontoparietal</td>
<td>yes</td>
<td>headaches</td>
<td>chemo</td>
</tr>
<tr>
<td>Paige &amp; Bernstein, 1995</td>
<td>51, M</td>
<td>bilat occipital</td>
<td>yes</td>
<td>scalp mass, headaches</td>
<td>chemo &amp; RT</td>
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<tr>
<td>Curty et al., 1997</td>
<td>19, M</td>
<td>rt parietal</td>
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<td>scalp lump, headaches</td>
<td>chemo</td>
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<tr>
<td>Freudenstein et al., 2000</td>
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<td>no</td>
<td>headaches, seizures</td>
<td>RT &amp; chemo</td>
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<td>77, M</td>
<td>lt orbitofrontal</td>
<td>yes</td>
<td>ocular proptosis</td>
<td>RT</td>
</tr>
</tbody>
</table>

* Chemo = chemotherapy; RT = radiation therapy.

Conclusions

Although it is extremely uncommon, primary malignant lymphoma should be considered in the differential diagnosis of scalp masses. This is especially true when a mass involves both dura mater and cranial bone, the patient has a painless clinical presentation, and the aforesaid imaging characteristics are observed. Nonetheless, the number of occurrences of this entity reported in the literature is too small to allow consideration of final conclusions.

References

13. Hsiang JN, Ng HK, Poon WS: Atypical mononclonal plasma cell...

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