Non-Hodgkin lymphoma of the cranial vault with retrobulbar metastasis mimicking a subacute subdural hematoma

Case report

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Primary skeletal NHL is rare. The authors report a case of a small lymphocytic B-cell lymphoma of the skull occurring in a 53-year-old man who presented with right-hand apraxia. Initial computed tomography and magnetic resonance imaging revealed a hematoma-mimicking lesion in the left frontoparietal subdural area. A frontotemporoparietal craniectomy and biopsy procedure yielded a diagnosis of small lymphocytic B-cell lymphoma, with a metastatic nodule in the retrobulbar area. Three years after undergoing radiation therapy and surgery, the patient has shown neurological improvement without systemic dissemination of the malignancy. The lesion in this case was misdiagnosed as a subdural hematoma, and shows the importance of including lymphoma in the differential diagnosis of subdural mass lesions. (DOI: 10.3171/JNS/2008/108/5/1018)

KEY WORDS • lymphocytic B-cell lymphoma • non-Hodgkin lymphoma • retrobulbar metastasis • subdural hematoma

Primary skeletal NHL is rare, occurring in only 1 in 20 cases of lymphoma; of these rare cases, > 75% originate in the pelvis and limbs.1–3 A primary NHL of the skull is exceedingly rare.1–3 The authors report on a patient found to have NHL of the cranial vault with a retrobulbar metastasis that mimicked a subacute subdural hematoma during preoperative radiological examination.

Case Report

History and Examination. This 53-year-old man without any history of head trauma presented to the emergency department with a 10-day history of right-hand apraxia. He had a history of pulmonary tuberculosis that had been cured after he received antituberculosis medication for 6 months. Laboratory test results (complete blood count, serum chemistry, and urinalysis) were within normal limits, the patient was not immunocompromised, and was HIV negative. The preoperative radiological diagnosis was a left frontoparietal subdural hematoma based on CT scans and MR images (Fig. 1).

Operation and Postoperative Course. We attempted to perform frontal and parietal bur hole trephination, but the liquefied hematoma was not evacuated from the subdural space. Instead, a hypervascular, firm mass lesion was found. A frontotemporoparietal craniectomy was extended from the bur holes. After removal of sponge-like denatured skull vault, a granulated tissuelike tough texture filled the epidural and subdural space, with unusual dural thickening. The subdural space was filled with yellowish, gelatinous tissue. The pathological diagnosis was a small lymphocytic B-cell lymphoma, which was positive for L26 and Bcl-2 and negative for CD3 (Fig. 2). After surgery, the patient’s right-hand apraxia was improved, but recurrent ischemic attacks such as dysphasia and hemiparesis on the right side occurred for 2 weeks. The initial neuroimaging results were reviewed and another left retrobulbar enhancing mass was found on CT and MR imaging, which was later confirmed to be a small lymphocytic B-cell lymphoma using an ultrasonography-guided biopsy procedure. There was no evidence of bone marrow involvement in the NHL on a bone scan. The patient underwent whole-brain radiation therapy at a dose of 3060 cGy and received booster radiation therapy to the skull and left eye of 1980 cGy. After these treatments, there was no evidence of tumor recurrence. The patient currently has mild dysphasia and right-hand clumsiness 3 years after his initial diagnosis.

Discussion

Secondary NHLs often show bone involvement, but primary osseous lymphoma is rare, and accounts for only 3–4% of the patients with lymphoma.3 A true primary malignant lymphoma of the bone is defined as “a solitary mass lesion with no evidence of disease at other sites and no systemic dissemination within 6 months of the detection of tumor.”2 In particular, initial involvement of the calvaria is extremely rare. Lymphomas of the cranial vault usually in-
volve the pericranium, underlying meninges, and subcutaneous tissues. These lesions are effectively treatable using surgery and radiation therapy with a good outcome in most cases.6,8

The signal intensity of a lymphoma on MR images is nonspecific; most reported cases of skull lymphomas were isointense on unenhanced MR images and showed marked enhancement after administration of contrast medium. Magnetic resonance imaging of lymphomas cannot be specific because its appearance can mimic those of metastatic carcinoma, osteomyelitis, or meningioma.7 A high index of clinical suspicion, awareness of the characteristic CT features, and aggressive therapy are therefore required for lymphomas, especially when a scalp mass extends through the skull.9

Small lymphocytic lymphoma and chronic lymphocytic leukemia are morphologically, phenotypically, and genotypically indistinguishable, differing only in the degree of peripheral blood leukocytosis. Chronic lymphocytic leukemia is the most common type of leukemia in adults in the Western world. Small lymphocytic lymphoma constitutes only 4% of NHL cases. Chronic lymphocytic leukemia and small lymphocytic lymphoma are much less common in Asian countries. Most patients present at ages > 50 years (median 60 years old); a male predominance has been noted, with a male/female ratio of 2:1. The course and prognosis of small lymphocytic lymphoma is variable and depends primarily on the clinical stage. Overall median survival is 4–6 years, but patients who present with minimal tumor burden may survive for ≥10 years.3 The prognosis of a patient with a skull lymphoma is uncertain, but either direct cerebral invasion or leptomeningeal seeding should indicate a less favorable prognosis.6,7,15 A primary lymphoma of bone is usually treated with local radiation therapy and/or systemic chemotherapy, and the 5-year survival rate has been reported to be > 60%.10,14 None of the previously reported 16 patients survived longer than 24 months, however, indicating that patients with malignant lymphomas originating from the cranial vault have an extremely poor prognosis.13 Our case showed a good clinical outcome only after surgery and radiotherapy.

Primary NHL of the cranial vault with periorbital metastasis is extremely rare; so far, only 1 other case has been reported in the English literature.7,11 In our case, only neurological deterioration was observed, without a scalp lump or exophthalmus symptoms. This lack of other symptoms led to the misdiagnosis of the lymphoma as a subacute subdural hematoma.

Conclusions
The authors emphasize the rarity of primary cranial vault
lymphoma and its importance in the differential diagnosis of cranial vault mass lesions extending either intra- or extracranially.

References

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