Primary Burkitt lymphoma of the brain in an immunocompetent patient

Case report

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Primary central nervous system (CNS) lymphoma is rare and is most often seen in immunodeficient patients. The majority of these tumors are the non-Hodgkin type and are high grade. Primary Burkitt lymphoma of the CNS in immunocompetent individuals has rarely been reported. The authors treated a 49-year-old woman who presented with left-sided weakness that had lasted 1.5 months. Magnetic resonance imaging revealed an oval mass in the left parietal region, with central necrosis and peripheral edema, and no attachment to the leptomeninges or ependyma. Pathological examination yielded a diagnosis of typical Burkitt lymphoma. Six months postoperatively, the patient is ambulatory and has improving neurological signs.

This is a typical case of primary Burkitt lymphoma of the brain in an immunocompetent patient, which is a very rare event. The imaging pattern of the lesion is not typical of brain lymphomas and can result in an incorrect preoperative diagnosis of other brain tumors, such as glioblastoma multiforme. The patient’s treatment and follow-up review are discussed.

KEY WORDS • brain neoplasm • Burkitt lymphoma

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The majority of CNS lymphomas consist of secondary involvement of the brain, spinal cord, or covering membranes by nodal or extranodal (rather than CNS) lymphomas. Primary CNS lymphoma is unusual and is mostly seen in immunodeficient patients, especially those with AIDS; however, well-documented primary lesions have been detected in the brains of immunocompetent patients. These tumors are primarily the non-Hodgkin type, and are high grade and of B-cell lineage, with large cell morphological characteristics, although nearly all other types of malignant lymphoma of the CNS are on record.

Biological markers of Epstein–Barr virus infection have been shown in either tumor cells or sera of immunodeficient and immunocompetent patients with primary CNS lymphoma. In contrast with AIDS-related tumors, CNS lymphomas occur in immunocompetent patients most frequently after their fifth decade of life.

One helpful guide to preoperative diagnosis of primary brain lymphoma is the radiological features of this lesion. It usually produces a solid and homogeneous pattern of contrast enhancement that is quite different from the peripheral ring-shaped enhancement that is typical of GBM. Primary central nervous system (CNS) lymphoma is rare and is most often seen in immunodeficient patients. The majority of these tumors are the non-Hodgkin type and are high grade. Primary Burkitt lymphoma of the CNS in immunocompetent individuals has rarely been reported. The authors treated a 49-year-old woman who presented with left-sided weakness that had lasted 1.5 months. Magnetic resonance imaging revealed an oval mass in the left parietal region, with central necrosis and peripheral edema, and no attachment to the leptomeninges or ependyma. Pathological examination yielded a diagnosis of typical Burkitt lymphoma. Six months postoperatively, the patient is ambulatory and has improving neurological signs.

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CASE REPORT

History and Examination. This 49-year-old woman presented with complaints of an emotional disorder beginning 2 months before and progressive left-sided weakness beginning 1.5 months before admission. On physical examination there was marked weakness on the left side, especially in the upper extremity. Cranial nerve functions and speech were normal. Her history revealed nothing significant except for an iron-deficiency anemia and on-and-off episodes of hypertension, for which she was under the care of a hematologist.

Neuroimaging Studies. Neuroimaging included CT scanning and MR imaging (both sagittal and coronal views) with and without addition of contrast agents.

Operation and Immunohistochemical Studies. Complete resection of tumor tissue was performed, paraffin-embedded blocks were prepared, and all slides were stained with hematoxylin and eosin. The paraffin-embedded blocks were also processed for immunohistochemical studies by using an avidin–biotin kit (DAKO, Glostrup, Denmark) according to the manufacturer’s instructions to identify the following markers: epithelial membrane antigen, cytokeratin, LCA, CD19, human melanoma B-45, glial fibrillary acidic protein, neuron-specific enolase, and synaptophysin.

Results of all laboratory tests, including hematological ones, were within normal limits. Serological tests for the
presence of antibodies to human immunodeficiency virus and Epstein–Barr virus yielded negative results. Different views on MR studies revealed an oval, nonhomogeneous mass in the right parietal lobe, with central necrosis and perilesional edema (Fig. 1). There was no involvement of the ependyma or leptomeninges. In our preoperative diagnosis we had suspected GBM.

Histopathological studies of the tumor material revealed a typical high-grade non-Hodgkin lymphoma of the small, noncleaved, Burkitt type (Fig. 2). There was frequent single cell necrosis; however, diffuse extensive confluent necrosis was absent.

On immunohistochemical evaluation, the tumor cells were found to be positive for LCA, CD19, and epithelial membrane antigen (focal), but all other markers were negative. Considering this cellular immunophenotype, the tumor was deemed a malignant lymphoma (LCA-positive) of B-cell origin (CD19-positive). Negative results on tests for cytokeratin and human melanoma-45 excluded the diagnoses of metastatic carcinoma and malignant melanoma, respectively, which were the main histological differential diagnoses. Also, the absence of neurogenic (neuron-specific enolase) and glial cell (glial fibrillary acidic protein) markers ruled out the possibility of high-grade neurogenic and glial tumors.

**Adjuvant Treatment.** The patient underwent two courses of chemotherapy (at 1-month intervals) with the cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) regimen. She also received 16 cycles of craniospinal radiation, with a total radiation dose of 32 Gy. After that she refused further treatment.

**Posttreatment Course.** One month postoperatively, the patient was ambulatory with the aid of a walker, and 6 months postoperatively her only complaint was mild upper-extremity weakness. Repeated follow-up MR imaging revealed no recurrent lesion (Fig. 3).

**Discussion**

It has been stated that the incidence of primary CNS lymphoma in immunocompetent adults is rising. Although the majority of these lesions are non-Hodgkin types, the Burkitt type of high-grade lymphoma has been sparsely reported. Although the majority of these lesions are non-Hodgkin types, the Burkitt type of high-grade lymphoma has been sparsely reported.8,16,17 A preoperative diagnosis of primary CNS lymphoma can be made with a high degree of certainty if the lesion is hyperdense on CT scanning, is invasive to the surrounding parenchyma, shows homogeneous enhancement, and has broad contact with ependyma and/or leptomeninges.9,11,12 In a large number of cases, Lee, et al.,9 evaluated CT find-
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ings in some brain lymphomas and have correlated these findings with the tumors’ pathological features. These authors have clearly discussed that homogeneous enhancement of primary CNS lymphoma on CT scanning is a common phenomenon, but that peripheral enhancement may be rarely seen, especially in AIDS-related lymphomas. They found central necrosis in the high-grade lymphomas with peripheral enhancement; none of those cases were the Burkitt type.

In suspected cases, cytological studies of the cerebrospinal fluid and/or stereotactic biopsy sampling accompanied by immunocytochemical studies will be helpful and lead to a definitive diagnosis.13 There is little benefit from surgical resection in cases of primary CNS lymphoma, which should be chiefly managed using steroid therapy followed by chemotherapy. Radiation therapy of the CNS is of little value in the management of primary CNS lymphoma, whereas it is of great importance in the treatment of systemic tumors. Our patient was treated initially by total resection, followed by chemotherapy and limited radiation therapy. She tolerated and responded well to this regimen.

Conclusions
Primary Burkitt lymphoma of the brain is a rare tumor. Clinically, the Burkitt lymphoma could not be differentiated from GBM. Histological findings were similar to those of anaplastic astrocytomas, metastatic small-cell malignant tumor, and malignant melanoma. Immunohistochemical studies helped us reach the correct diagnosis in this case. Total resection, chemotherapy, and radiotherapy were used in the management of this patient’s disease.

References

Manuscript received March 26, 2001. Accepted in final form January 18, 2002.

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