Primary cerebral lymphoma: the “ghost tumor”

Case report

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A case of primary cerebral lymphoma is presented. The particular biological features of these tumors may pose a special problem. In this case, the lesion was diagnosed on computerized tomography (CT) but was not found at surgery. The disappearance of the lesion was associated with a long period of corticosteroid therapy between CT diagnosis and the operation.

KEY WORDS • cerebral lymphoma • corticosteroid therapy • microglioma • reticulum cell sarcoma

During the last decade, the terms “malignant lymphoma” and “lymphoreticular tumor” have been introduced in place of “histiocytic sarcoma,” “reticulum cell sarcoma,” or “microglioma.” Whatever the preferred nomenclature, these rare tumors comprise only 0.3% to 1.5% of all intracranial neoplasms, but have increased in frequency during the last few years.

The progressive clinical course of primary lymphoma of the brain cannot be differentiated on clinical grounds alone from the more common malignant gliomas, but some case reports have documented the remission of this tumor without radiation therapy. We describe a case of primary cerebral lymphoma which temporarily showed total remission after corticosteroid therapy.

Case Report

This 59-year-old man was admitted as an emergency to this department on September 10, 1982, because of progressive generalized headache, left hemiparesis, urinary incontinence, and drowsiness.

Examination. Neurological examination revealed an obtunded patient, with left homonymous hemianopia and hemiparesis. Computerized tomography (CT) showed a mass with an irregular densitometric pattern in the right ventricular atrium, notable surrounding edema, and marked enhancement after contrast administration (Fig. 1). Dexamethasone was given intramuscularly, at a dose of 4 mg/6 hr, and 72 hours later a great improvement in the clinical picture was observed. For this reason the operative procedure was postponed, and the patient was maintained on corticosteroid therapy.

Operation. On October 5, 1982, a craniotomy was performed, but no tumor mass was encountered within the right lateral ventricle, nor in its lateral wall. Multiple biopsies were taken in the region, but histological examination only showed nervous tissue with inflammatory cells, suggesting a brain infarction zone.

Postoperative Course. After surgery, the neurological situation was unchanged, but on the 10th postoperative day the patient suffered a pulmonary embolism secondary to a silent deep venous thrombosis of the left leg. He was referred to the intensive care unit where therapy with anticoagulant drugs was established. Dexamethasone, which had been stopped on the 7th postoperative day, was started again at a dose of 4 mg/6 hr intravenously. On November 5, 1982, the dose of corticosteroids was diminished, and a CT scan on the same day showed a very small lesion in the right ventricular atrium region, which enhanced after contrast administration (Fig. 2). On November 15, two days after corticosteroids had been stopped, a new CT scan showed complete disappearance of the lesion (Fig. 3). Over the following days, changes in the behavior of the patient were observed, showing progressive disorientation and irritability. Lumbar puncture was performed and cerebrospinal fluid (CSF) analysis was normal. Tumor cytology in the CSF was also negative.
Temporary disappearance of primary cerebral lymphoma

FIG. 1. Computerized tomography scans on September 10, 1982. *Left:* Plain scan showing a mass in the right ventricular atrium. *Right:* The mass shows marked enhancement after contrast administration.

FIG. 2. Computerized tomography scans on November 5, 1982, after surgical exploration. *Left:* Plain scan showing a very small lesion in the right ventricular atrium region. *Right:* The mass is enhanced after contrast administration.

FIG. 3. Computerized tomography scans, plain (*left*) and after contrast administration (*right*), taken on November 15, 1982. There is complete disappearance of the lesion.

FIG. 4. Computerized tomography scans, plain (*left*) and after contrast administration (*right*), taken on January 25, 1983. A lesion is again visible in the right ventricular atrium.

FIG. 5. Histological picture of the tumor. H & E, × 70.

On January 25, 1983, a CT scan again showed a mass in the right ventricular atrium (Fig. 4). The neurological picture deteriorated rapidly, with worsening of the left hemiparesis. Dementia and aggressiveness developed. The diagnosis of primary cerebral lymphoma which remitted with steroid therapy was then suspected, and intravenous treatment with dexamethasone, at a dose of 4 mg/6 hr, was started again. Electrophoresis of plasma proteins was performed but no discrete bands or abnormal proteins were found. The department of internal medicine evaluated the patient, and reasonably ruled out any sign of extracerebral lymphoma. On March 16, 1983, CT showed no changes in the lesion, but treatment with dexamethasone was continued. Performance of a stereotaxis procedure for biopsy or a new surgical exploration was proposed, but the patient died.

Postmortem Examination. The craniotomy was reopened and a firm white mass, with some soft fleshy areas, was found in the wall of the right lateral ventricle, partially filling the ventricular cavity at the level of the atrium. Pathological study showed a malignant cerebral lymphoma (Fig. 5).

Discussion

In our case, remission of a primary cerebral lymphoma occurred consistently in conjunction with the administration of corticosteroids, as has happened in at least five other previously reported cases.5,8,10,11 It has
been stated before that corticosteroids may favorably alter the biological activity of tumor tissue in some cases of cerebral lymphoma, predisposing to clinical remission and disappearance of the tumor on CT scanning, although the response varies depending upon the immunological competence of the host. Another possible mechanism is a direct cytotoxic effect of the corticosteroids on the lymphoreticular cell, but it is very difficult to explain why this effect is transitory and not consistent. In conclusion, the temporary disappearance with corticosteroid therapy of a cerebral lymphoma is a possibility to be kept in mind when a diagnosed tumor is not found at surgery, and the patient has been treated with corticosteroid therapy during an interval between radiological diagnosis and the operation.

References


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