Rapidly progressive dementia caused by nonenhancing primary lymphoma of the central nervous system.

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Rapidly Progressive Dementia Caused by Nonenhancing Primary Lymphoma of the Central Nervous System

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Summary: A 76-year-old woman had rapidly progressive dementia over 4 months. Proton density– and T2-weighted MR images of the head showed increased signal in the periventricular and subcortical white matter of both cerebral hemispheres and the brain stem. Enhancement was not seen after administration of contrast material. Because of the rapid progression of the dementia and increasing signal abnormalities within the cerebral white matter, the patient underwent a craniotomy with biopsies of the right frontal lobe. Pathologic specimens were positive for malignant large-cell lymphoma, B-cell phenotype. The MR appearance is atypical for primary lymphoma of the central nervous system.

Index terms: Dementia; Lymphoma

An elderly woman with rapidly progressive dementia without immunocompromise is described. Magnetic resonance (MR) images showed an unusual appearance of a primary lymphoma of the central nervous system (CNS) that did not enhance in the regions of diffuse white matter signal abnormality after administration of contrast material.

Case Report

A 76-year-old woman came to our institution for evaluation of a rapidly progressive encephalopathy manifested by lethargy, confusion, disorientation, and dysphasia. She had been in good health before her neurologic deterioration and was without evidence of immunosuppression. A contrast-enhanced T2-weighted MR image obtained elsewhere showed extensive signal abnormalities in the deep and subcortical white matter of both cerebral hemispheres without enhancement. A lumbar puncture revealed an increased protein level, low glucose level, and lymphocytosis. Cerebrospinal fluid cultures, results of cytologic examination, and tests for Lyme disease and human immunodeficiency virus were negative. An electroencephalogram showed generalized, nonspecific slowing that was slightly worse on the left side. A cerebral angiogram showed no evidence of vasculitis.

Repeat contrast-enhanced MR imaging at our institution (Fig 1) revealed a significant increase in signal abnormality on T2-weighted images in the white matter of both cerebral hemispheres compared with the outside examination obtained 1 month earlier. Increased signal was also noted in both thalami. Neither mass effect nor enhancement was apparent on a postcontrast study. Results of an open right frontal lobe biopsy demonstrated malignant large-cell lymphoma, B-cell phenotype. The patient was treated with steroids only. Chemotherapy and radiation therapy were offered, but their chance for success was thought to be very low, and the family declined. The patient was transferred back to a local nursing home and died approximately 1 month after dismissal. An autopsy was not performed.

Discussion

The cause of this patient's rapidly progressive encephalopathy eluded diagnosis despite multiple clinical, laboratory, and imaging studies. The MR findings in this case are unusual for primary CNS lymphoma and are more consistent with toxic or metabolic abnormalities; inflammatory, ischemic, or infectious processes (such as progressive multifocal leukoencephalopathy); or a progressive demyelinating disorder. Lack of enhancement on MR images has often resulted in delay of diagnosis of this disease (1).

Primary CNS lymphoma accounts for 1% to 2% of all CNS neoplasms (2). The prevalence of this tumor is increasing in immunocompromised patients (2). The vast majority of cases are non-Hodgkin B-cell lymphomas histologically (3). This neoplasm can occur at any age, but it is most common in the sixth and seventh decades (2). Patients present with headaches, seizures, focal and diffuse neurologic deficits, and encephalopathy (2). Usual imaging features of primary CNS lymphoma

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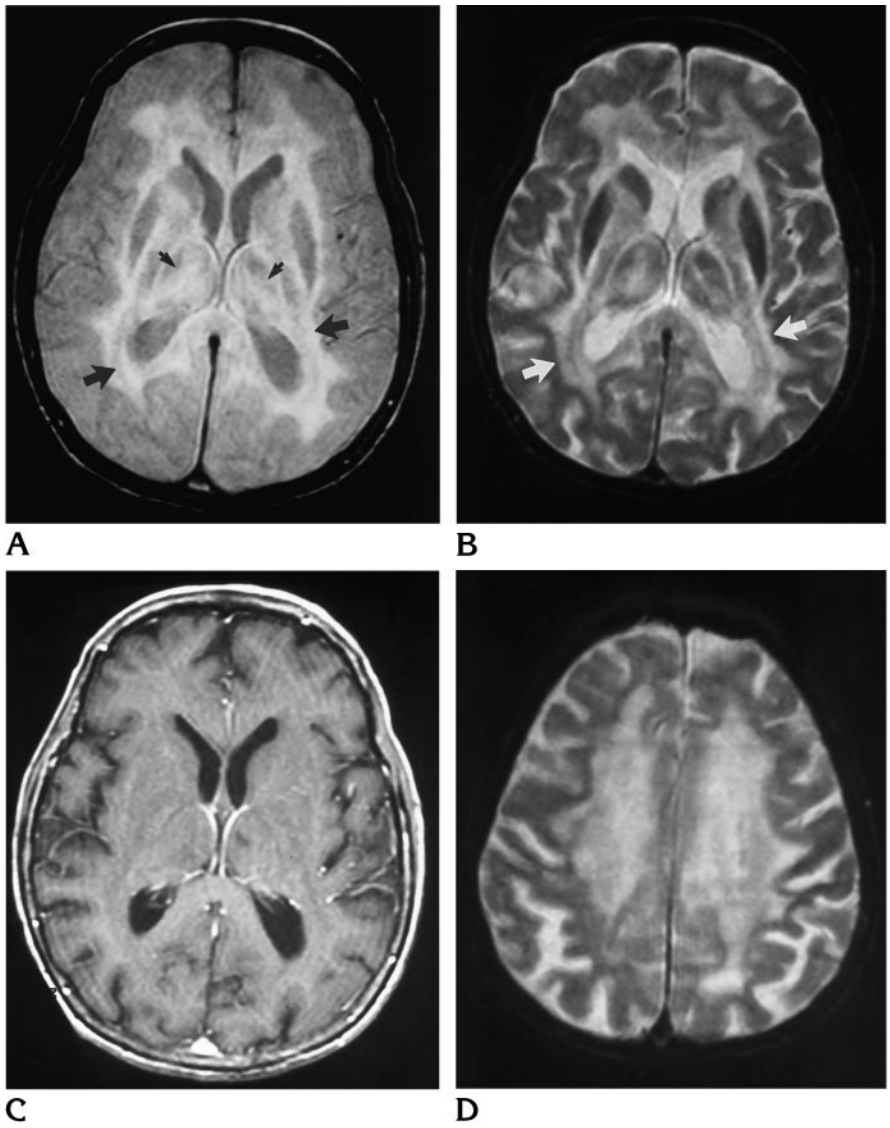

Fig 1. Axial proton density–weighted (2200/30 [repetition time/echo time]) (*A*), T2-weighted (2200/80) (*B*), and contrast-enhanced T1-weighted (480/26) (*C*) MR images at the ventricular level show widespread increased T2 signal in the white matter (*large arrows*) and thalami (*small arrows*) bilaterally. No appreciable enhancement or mass effect was noted after administration of contrast material. T2-weighted image (*D*) at level of centrum semiovale shows high signal without mass effect.

in the immunocompetent population are isointense to mildly hyperintense signal on T2-weighted sequences, often involving the basal ganglia, periventricular white matter, and corpus callosum. Lesions may be single or multiple, and they often show homogeneous and significant enhancement on postcontrast studies (2). In immunocompromised patients, this neoplasm may appear more heterogeneous on imaging studies, including regions of ring enhancement (necrosis) and hemorrhage (2). Unenhanced computed tomography scans often show lymphomas to be hyperdense with strong enhancement after intravenous administration of contrast material (4).

The findings of progressive, diffuse white matter signal abnormality without significant enhancement or mass effect in this patient suggest that her lymphoma was diffusely infiltrative and had not resulted in significant disruption of the blood-brain barrier. Her rapid cognitive decline and the desire to identify any potentially treatable cause of this unknown process led to the biopsy. Awareness that CNS lymphoma can rarely present as a nonenhancing infiltrative lesion may shorten the interval between onset of clinical symptoms and diagnosis in patients with this unusual form of CNS neoplasm.

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