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Primary CNS Lymphoma: An Unusual Case of Prolonged Response to Steroids and Extended Survival (21 Years)

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Case Report

In 1991, a 27-year-old man presented at the emergency department of a country hospital with diplopia, tickling paraesthesia, and a right-sided hemiparesis. He underwent contrast-enhanced brain MRI which documented a lesion with inhomogeneous enhancement in the right paramedian pontine region. Other examinations (CSF analysis with isoelectric focussing, EMG, SSEP) were negative and steroids (dexamethasone 8 mg/d) were then administered with clinical improvement.

The MRI examinations at 1 and 6 months after steroid therapy were stable and the patient remained asymptomatic.

In 2007, 16 years after the first episode, the man was admitted to our hospital with dizziness and diplopia. Contrast-enhanced MRI showed multiple enhancing areas in the right cerebellar hemisphere, the floor of the fourth ventricle, and the mesencephalic-diencephalic region. The original pontine lesion on brain MRI was unchanged and spine MRI was normal (Figures 1A, 1B). The patient was extensively investigated (systemic work-up and CSF analysis), and the spectrum of steroid-responsive non-neoplastic lesions in the CNS (multiple sclerosis, neurosarcoidosis, Behçet disease, and atypical tuberculosis) was ruled out. Primary central nervous system lymphoma (PCNSL) was then suspected. An HIV serum test was negative. The patient refused a biopsy, then was treated...
In April 2009, at the age of 45, the patient presented again at the emergency department with headache, vomiting, and dizziness. We performed contrast-enhanced brain MRI which showed a new, large enhancing lesion in the left cerebellum with distortion of the fourth ventricle but no evidence of hydrocephalus (Figures 2A, 2B).

The lesion was surgically partially resected and the histological diagnosis was that of diffuse large B-cell lymphoma (Figure 3). The examinations performed for the staging of lymphoma (contrast-enhanced spine MRI, CSF analysis, chest-abdomen CT scan, whole-body PET scan, testicular ultrasound, and bone marrow biopsy) ruled out systemic involvement.

The patient started chemotherapy with high-dose methotrexate (3.5 gr/m² body surface every 2 weeks). He completed 6 cycles with good tolerance and nCR (near complete response) on MRI. Whole-brain radiotherapy (WBRT; 36 Gy + boost of 9 Gy) was then performed. At the first brain MRI performed after the end of treatment (December 2009) complete response was observed (Figures 4A, 4B) and subsequent serial MRI examinations showed no recurrence (last MRI performed in January 2012). The patient is now neurologically normal without cognitive deficits and is continuing follow-up with MRI every 6 months.

Comment

Primary CNS lymphoma, when affecting young and immunocompetent subjects and being located in critical areas for biopsy, can present diagnostic problems at onset. Treatment with steroids can lead to a regression of lesions in up to 40% of patients, thus deferring the definite histological diagnosis.

Our case shows some peculiar features: a long disease history (overall, 21 years) with multiple recurrences involving exclusively the brain stem, cerebellum, and thalamus; an unusual and protracted response to steroids alone and complete response to standard chemo- and radiotherapies even if performed very late (18 years after the onset of symptoms).

Further Reading:


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