Volume 3 (2013) // Issue 1 // e-ISSN 2224-3453

European Association of NeuroOncology Magazine

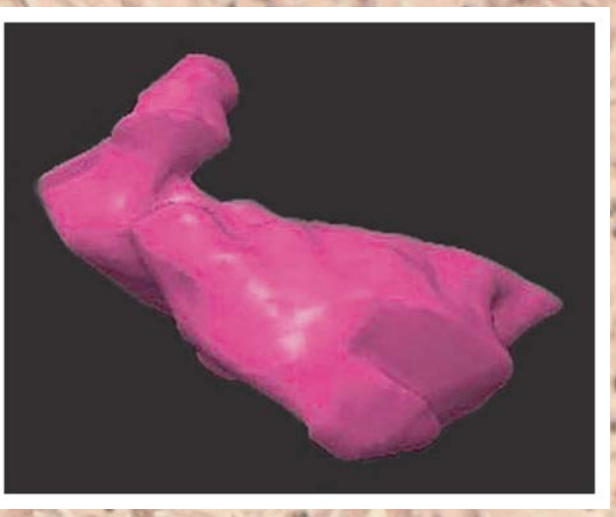
Neurology · Neurosurgery · Medical Oncology · Radiotherapy · Paediatric Neuro-oncology · Neuropathology · Neuroradiology · Neuroimaging · Nursing · Patient Issues

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

Bosa C, Bertero L, Trevisan E

Ruda R

European Association of NeuroOncology Magazine 2013; 3 (1) 27-28



Homepage:

www.kup.at/journals/eano/index.html

Online Database Featuring Author, Key Word and Full-Text Search

THE EUROPEAN ASSOCIATION OF NEUROONCOLOGY

Member of the DOAJ DIRECTORY OF OPEN ACCESS JOURNALS

Krause & Pachernegg GmbH · VERLAG für MEDIZIN und WIRTSCHAFT · 3003 Gablitz, Austria

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

Chiara Bosa, Luca Bertero, Elisa Trevisan, Roberta Rudà

From the Department of Neuro-Oncology, University and Città della Salute e della Scienza, Turin, Italy

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

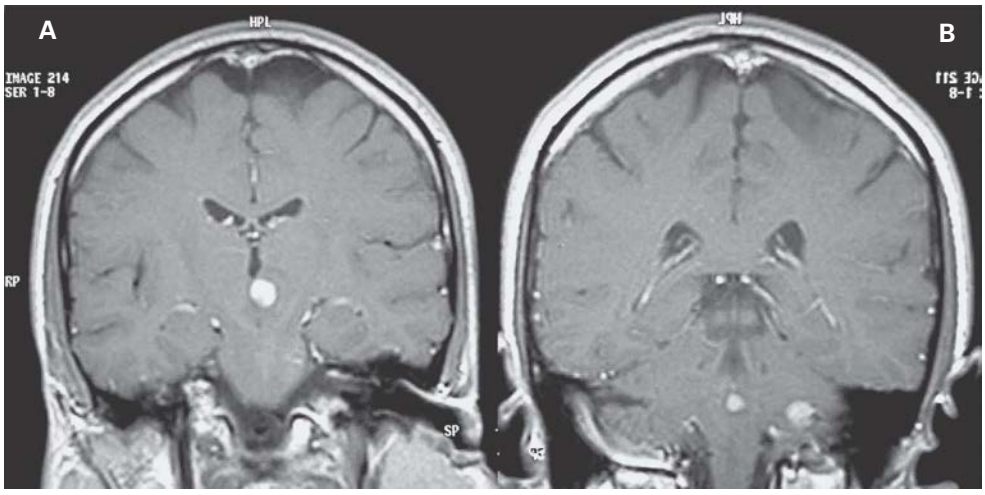


Figure 1: T1-weighted MR images with gadolinium at first relapse in 2007.

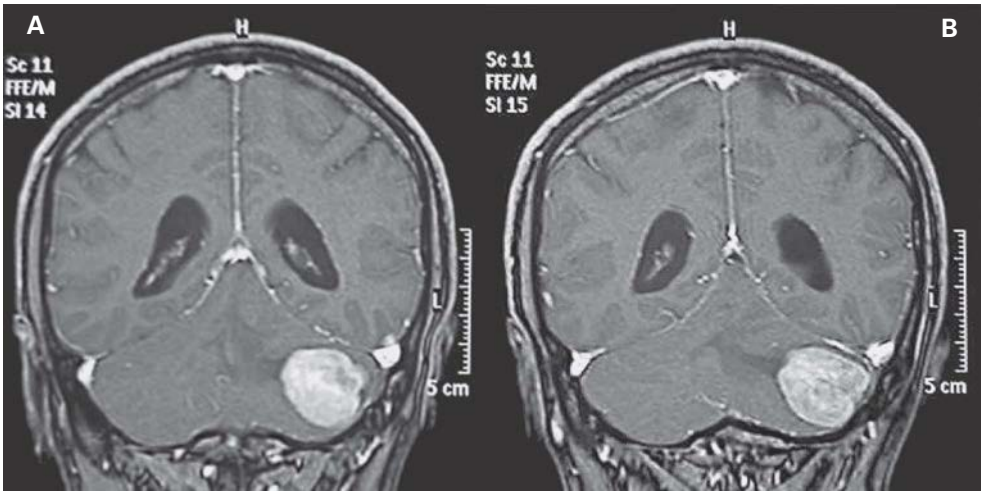


Figure 2: T1-weighted MR images with gadolinium at second relapse in 2009.

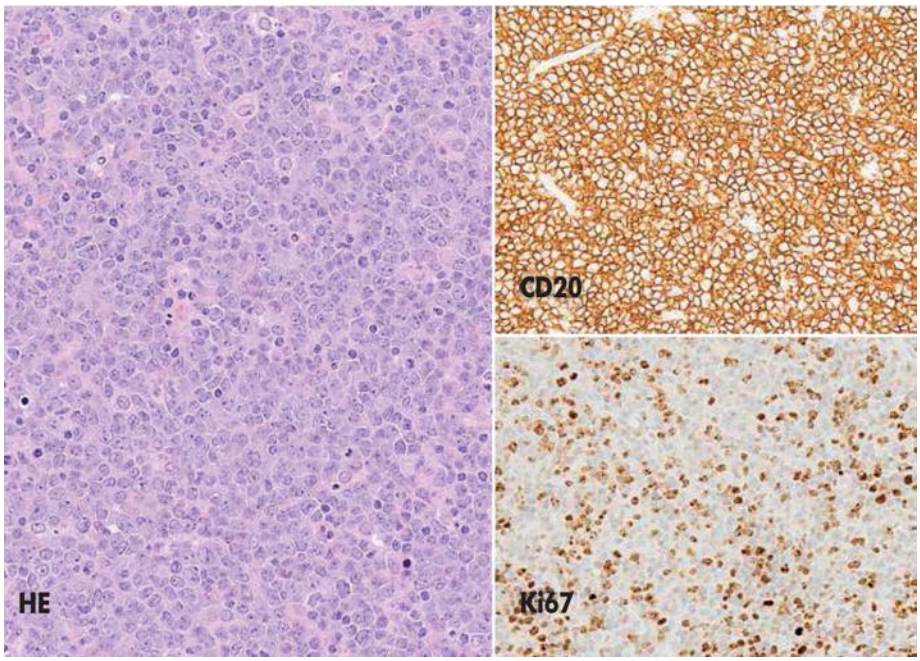


Figure 3. Hematoxylin and eosin staining, CD20 staining, and Ki67 staining.

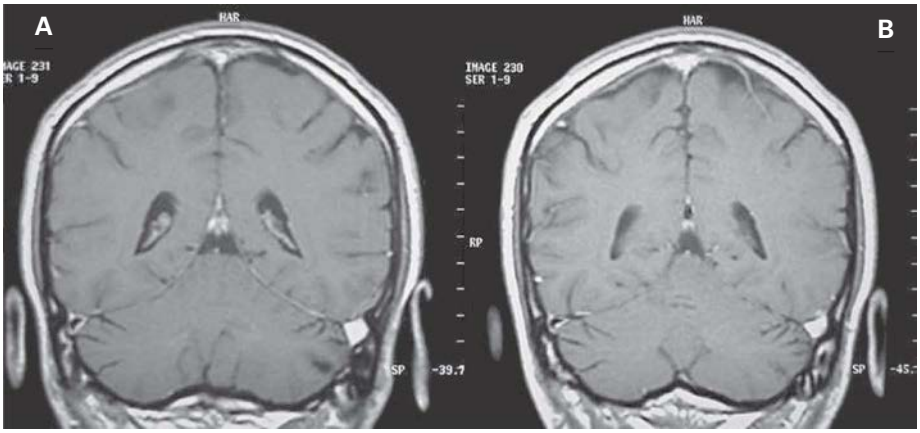


Figure 4: T1-weighted MR images with gadolinium after treatment with high-dose methotrexate and whole-brain radiotherapy.

with steroids and complete clinical and radiological remission was achieved.

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/m^2 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:

Algazi AP, Kadoch C, Rubenstein JL. Biology and treatment of primary central nervous system lymphoma. Neurotherapeutics 2009; 6: 587–97.

Clarke JL, Deangelis LM. Primary central nervous system lymphoma. Handb Clin Neurol 2012; 105: 517–27.

Ferreri AJ. How I treat primary CNS lymphoma. Blood 2011; 118: 510–22.

Gerstner ER, Batchelor TT. Primary central nervous system lymphoma. Arch Neurol 2010; 67: 291–7.

Schäfer N, Glas M, Herrlinger U. Primary CNS lymphoma: a clinician's guide. Expert Rev Neurother 2012; 12: 1197–206.

Correspondence to:
Roberta Rudà, MD
Department of Neuro-Oncology
University and San Giovanni Battista Hospital
Via Cherasco 15, 10126 Torino, Italy
E-mail: rudarob@hotmail.com