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ID IMAGES

Primary Intramedullary Spinal Cord Lymphoma Presenting as a Cervical Ring–Enhancing Lesion in an AIDS Patient

Julia Bini Viotti,[1] Susanne Doblecki,[1] Corneliu C. Luca,[2] Nicholas Mackrides,[3] Francisco Vega,[3] and Maria L. Alcaide[1]

[1]Division of Infectious Diseases, Department of Medicine, [2]Department of Neurology, and [3]Hematopathology Division, Department of Pathology and Laboratory Medicine, University of Miami Miller School of Medicine, Miami, Florida

Primary intramedullary spinal cord lymphoma (PISCL) is rare and constitutes only 1% of central nervous system lymphomas. We report a case of PISCL in a 37-year-old man with advanced AIDS. To our knowledge, only 4 cases of PISCL in the setting of HIV/AIDS have been reported in the literature. Despite treatment, prognosis remains dismal.

Keywords. AIDS; AIDS- related opportunistic infection; HIV; intramedullary spinal cord neoplasms; lymphoma.

CASE

A 37-year-old African American man with untreated HIV infection diagnosed 1 year prior presented after 1 week of neck pain and progressive left-sided weakness and numbness. He was born and raised in Miami, Florida, and denied history of traveling outside the state of Florida or outside the United States. Physical examination revealed left ptosis, hemiparesis, and impaired sensation to light touch, pinprick, and temperature in his left hemibody. Complete blood count showed a white blood cell count (WBC) of 2.100/mcl (reference range between 4.000 and 10.500/mcl), with 11.8% lymphocytes (reference range between 16% and 43%). Serum chemistry was within reference range. CD4 T-cell count was 17 cells/mm^3, and HIV-1 RNA plasma viral load was 147 338 copies/mL. No abnormalities were observed in the brain magnetic resonance image (MRI). Cervical MRI demonstrated a focal isointense T1 and low T2 signal central spinal cord lesion extending from level C2-C3 with associated inhomogenous enhancement (Figures 1 and 2). Cerebrospinal fluid (CSF) analysis showed a cell count of 9 white blood cells/μL, a protein level of 138 mg/dl, and a glucose level of 51 mg/dL. CSF cultures were negative for bacteria, fungi, and viruses. CSF cytology and flow cytometry failed to show any malignant cells or immunophenotypic evidence of lymphoma. CSF venereal disease research laboratory (VDRL), cryptococcus antigen, and polymerase chain reaction (herpes simplex virus 1 and 2, cytomegalovirus [CMV], varicella zoster virus [VZV], john cunningham virus, *Toxoplasma gondii*, and tuberculosis complex) were also negative. The patient underwent C2-C3 laminectomy and intramedullary tumor biopsy.

A diagnosis of primary high-grade B-cell lymphoma was made by histopathology (Figure 3). Epstein-Barr virus (EBV) was detected by qualitative PCR from the CSF. There was no visceral tumor involvement. Antiretroviral treatment was initiated with excellent virological response, but CD4+ cell count remained <50 cells/mm^3. He initially received chemotherapy with methotrexate, high-dose intravenous zivodudine, and rituximab followed by cyclophosphamide, vincristine, dexamethasone, and doxorubicin (hyper-CVAD protocol), and, subsequently, radiation therapy. Disease progression continued despite chemoradiation therapy, and he became tetraplegic and ventilator-dependent. The family elected palliative care. Death from sepsis occurred 115 days after the lymphoma diagnosis.

DISCUSSION

The differential diagnosis of central nervous system (CNS) ring-enhancing lesions in patients living with HIV/AIDS is broad and includes toxoplasmosis encephalitis, primary CNS lymphoma, syphilis, cryptococcosis, tuberculosis, and other infectious and noninfectious etiologies. Spinal cord disease associated with HIV/AIDS is less frequently reported. Almost all diffuse or multifocal pathology processes involving the CNS and/or leptomeninges may also affect the spinal cord, but a single ring-enhancing lesion in the spinal cord is less common. In the setting of advanced AIDS and spinal cord disease without concomitant brain lesions, vacuolar myelopathy, human T-lymphotropic virus 1 (HTLV-1), CMV, and VZV myelitis should also be included in the differential diagnosis, but the imaging presentation is typically abnormal signaling or diffuse enhancement, rather than a space occupying lesion.

Intramedullary spinal cord lymphoma (ISCL) may be primary and originate in the spinal cord or accompany other CNS sites sequentially or concurrently. ISCL may also occur in the context of systemic lymphoma. Primary ISCL (PISCL) is rare and constitutes only 1% of CNS lymphomas [1]. Only 4 cases of PISCL in the setting of HIV/AIDS have been reported in the literature [1–4]. Our case is unique due to location of the lesion.

Received 2 March 2018; editorial decision 22 May 2018; accepted 31 May 2018.

Correspondence: B. V. Julia MD, University of Miami Miller School of Medicine, University of Miami Clinical Research Building, 1120 NW 14th Street, Miami, FL 33136 (jujubini@gmail.com).

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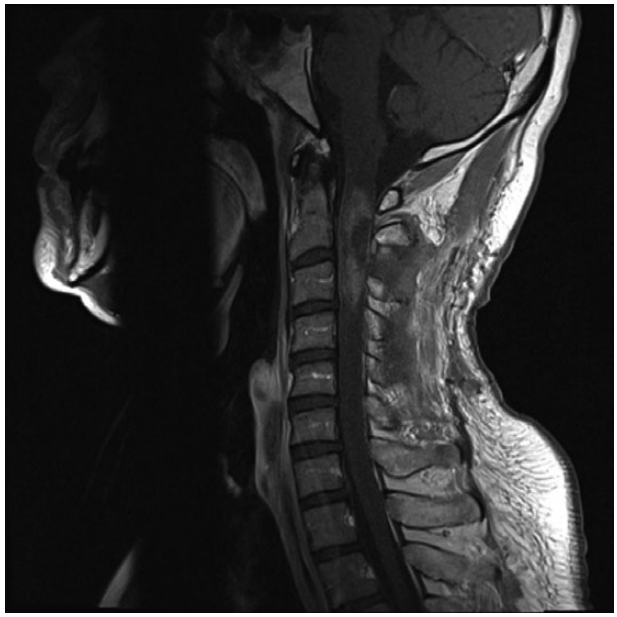

Figure 1. T1 post–gadolinium magnetic resonance sequence demonstrates a large focal intramedulary lesion at the C2-C3 level with nonhomogenous ring enhancement. The lesion measures 1.7 × 1.4 × 4.8 cm.

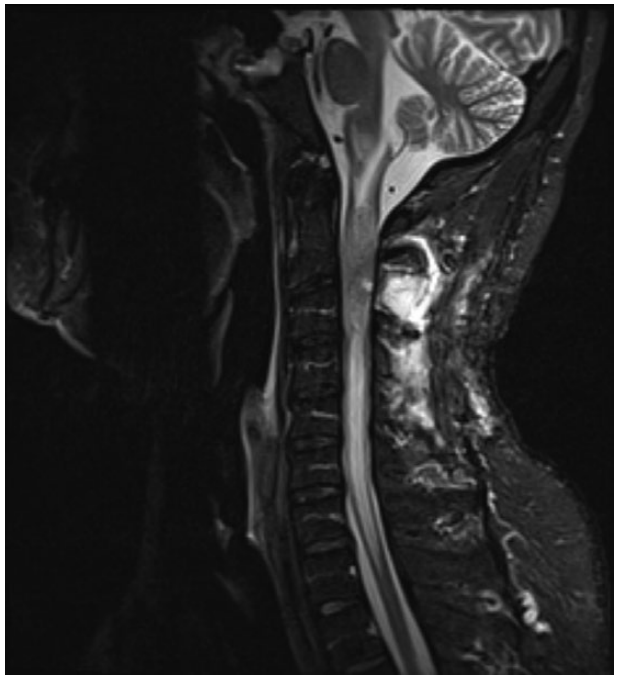

Figure 2. Surrounding T2 signal is noted below and above this lesion; the lesion involves most of the axial dimension of the cord and results in cord expansion.

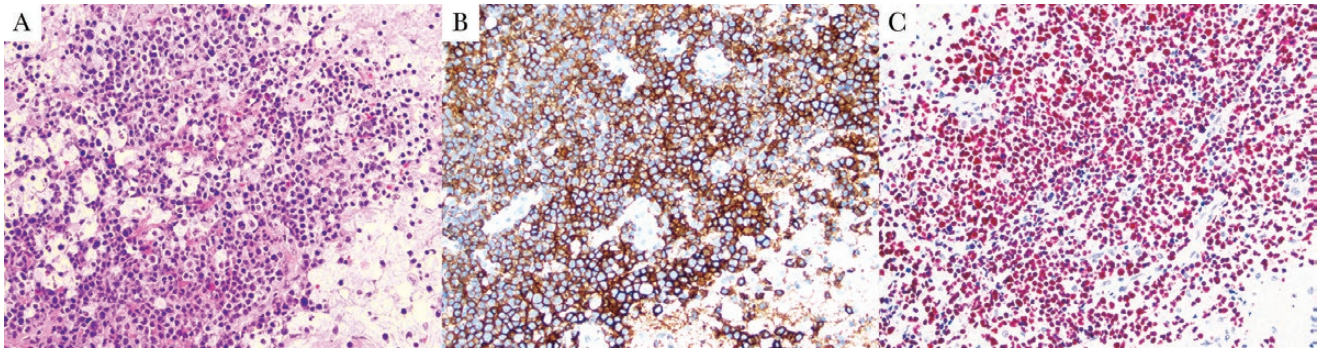

Figure 3. HIV-associated high-grade B-cell lymphoma, Ebstein-Barr virus positive. The biopsy shows diffuse areas with intermediate-sized atypical lymphoid cells associated with necrosis and apoptosis, with focal areas of tingible body macrophages imparting a "starry sky" pattern (A). The lymphoma cells are positive for CD20 (B), supporting the presence of high-grade B-cell lymphoma. Epstein-Barr virus–encoded RNA in situ hybridization is positive (C) (A, B, C, 40×).

Delay to diagnosis is common. CSF cytology is diagnostic in a minority of cases, and although detection of EBV in CSF is suggestive of CNS lymphoma, definitive diagnosis relies on histopathology [1].

High-dose methotrexate-based chemotherapy is the mainstay of induction therapy for primary CNS lymphoma. CNS irradiation and high-dose chemotherapy supported by autologous stem cell transplantation are 2 effective consolidation strategies in patients with a disease responsive to induction chemotherapy. Treatment of EBV with antiviral agents such as zidovudine may be a beneficial approach for the treatment of EBV-related lymphomas in people living with HIV, but it needs further study [5]. Highly active antiretroviral therapy is mandatory for HIV/AIDS patients. Despite treatment, long-term survival is infrequent [1]. The maximum reported survival in the setting of HIV/AIDS was 13 months after diagnosis [2].

Acknowledgments

Financial support. The Miami Center for AIDS Research (CFAR) at the University of Miami Miller School of Medicine (P30AI073961) provided mentorship and assistance with manuscript development.

We thank the Department of Radiology of the Jackson Memorial Hospital in Miami, Florida, for providing the MRI figures. We also thank the Division of Hematopathology of the University of Miami/Sylvester Comprehensive Cancer Center, Florida, for providing the pathology figures.

Potential conflicts of interest. All authors: no conflicts of interest. All authors have submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest. Conflicts that the editors consider relevant to the content of the manuscript have been disclosed.

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