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Primary dural lymphoma: Complete remission after treatment with radiation therapy

Jeremy C. Jones, Arjun Gupta, Harris Naina
Department of Internal Medicine, University of Texas Southwestern Medical Center, Dallas, TX

Abstract. Central nervous system (CNS) involvement in sarcoidosis is rare and typically occurs in 5-10% of patients. Neurological symptoms in a patient with known sarcoidosis can be attributed to neurosarcoidosis without thorough evaluation. Primary Dural Lymphoma (PDL) is an extremely rare form of non-Hodgkin lymphoma. Although PDL is technically a subtype of primary CNS lymphoma, the two entities vary markedly in their histological grade, clinical course, prognosis and treatment. The most common dural- based lesion found on CNS imaging is meningioma. It shares many imaging, clinical and epidemiologic features of PDL which often leads to misdiagnosis of PDL as meningioma. We present a case where a PDL was diagnosed after CNS symptoms failed to resolve after steroid therapy for presumed neurosarcoidosis. *(Sarcoidosis Vasc Diffuse Lung Dis 2015; 32: 80-82)*

Key words: Primary dural lymphoma, Sarcoidosis, Radiation therapy

Introduction

Primary dural lymphoma (PDL) is a rare subtype of primary central nervous system lymphoma (PCNSL). Unlike traditional PCNSL, which is an aggressive diffuse B-cell lymphoma involving the brain parenchyma, PDL is generally a low-grade, mantle zone lymphoma which follows a more indolent clinical course (1,2).

Case

A 63-year-old African American female with a history of pulmonary sarcoidosis, on chronic low-dose steroid therapy, presented to her primary care physician with complaints of bifrontal, throbbing headache for three months. She continued to have progression in her symptoms with worsening headache and double vision despite treatment with over the counter analgesics. MRI of her brain showed a 4 mm left internal carotid aneurysm, diffuse irregular dural thickening and enhancement along the frontal lobes as well as an enhancing mass in the posterior aspect of the bilateral orbits, contacting the optic nerves (Figure 1). These findings were initially attributed to extrapulmonary complications of sarcoidosis and she was treated with oral steroids with significant symptomatic improvement. She subsequently underwent neurosurgical clipping of her carotid aneurysm and intraoperative dural biopsy of the previously seen thickening; although, preoperative imaging showed the previously seen dural thickening and orbital mass had markedly diminished in size since initial imaging. Pathology from the dural biopsy showed a dense infiltrate of plasmacytoid lymphocytes, plasma cells, and small lymphocytes. Immunohistochemical staining showed cells that were CD20 positive, CD10 and CD5 negative, kappa re-

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Correspondence: Harris V Naina, M.D.
Seay Biomedical Building
2201 Inwood Road
Dallas, Texas 75390-8562
Office Phone: (214) 648-4155
Office Fax: (214) 648-4152
E-mail: harris.naina@utsouthwestern.edu

stricted and with an MIB-1 less than 10%. There were also focally expanded dendritic cell meshworks which stained positive for CD21 (Figure 2). Flow cytometry revealed kappa restricted monoclonal CD5 negative /CD10 negative B-lineage cells. The differential diagnosis included marginal zone lymphoma and lymphoplasmacytic lymphoma. The absence of an associated IgM component in the serum virtually ruled out lympho-plasmacytic lymphoma. Epstein-Barr encoding region (EBER) in situ hybridization was negative as well.

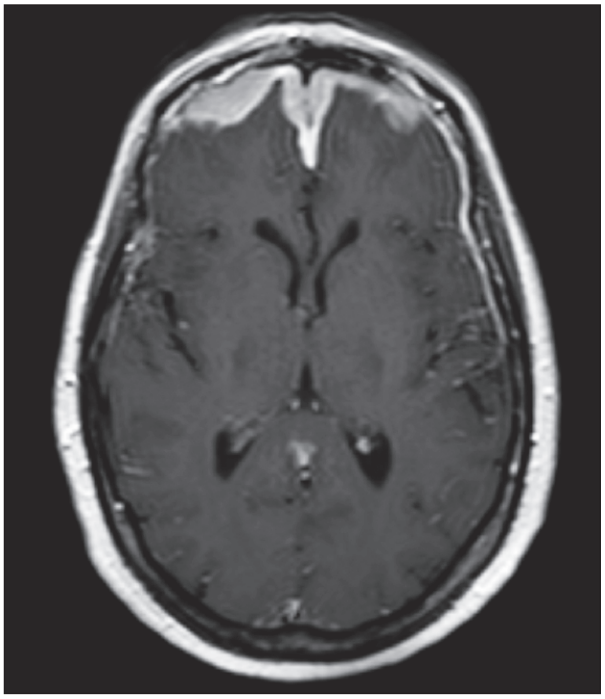

Fig. 1. T_1-weighted post contrast magnetic resonance imaging (MRI) at initial diagnosis showing dural thickening and enhancement along the bilateral frontal lobes.

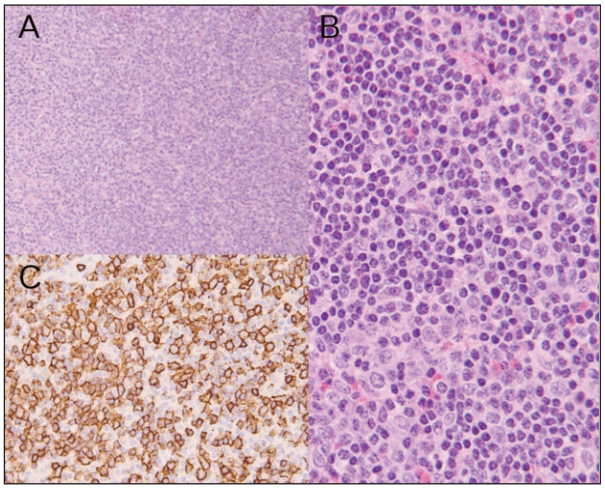

Fig. 2. Microscopic pathology from open dural biopsy. (A) Hematoxylin and eosin (H&E) stain of dural biopsy at 2 x magnification. (B) Hematoxylin and eosin (H&E) stain at 40 x magnification with dense infiltrate of plasmacytoid lymphocytes, plasma cells and small lymphocytes. (C) immunohistochemical staining for CD20.

Bone marrow analysis revealed 30-40% cellular marrow with adequate trilineage hematopoiesis and normal female karyotype with no evidence of any consistent chromosomal rearrangement to suggest an acquired clonal abnormality. CT of the chest, abdomen and pelvis did not show any other lesions.

She received whole brain radiation therapy with a total dose of 3240 cGy delivered over 18 fractions during a 26-day period. She has since been monitored clinically and with brain imaging every 6 months for 32 months following diagnosis. She remained in complete remission both clinically and on imaging studies.

Discussion

This case represents a common presentation of an extremely uncommon disease but it highlights an important point in the differential diagnosis of any dural-based mass found on CNS imaging. By far, the most common primary dural-based mass found on CNS imaging is meningioma with an incidence of approximately 8.2 per 100,000 (3). The overall incidence of primary dural lymphoma is unknown but given the incidence of all primary CNS lymphomas is approximately 0.48 per 100,000; one can assume the incidence of such a rare subtype is orders of magnitude less (4). Both tumors present with iso- or hypointense dural-based masses on T_1-weighted MR images with diffuse contrast enhancement (1). Both are predominately found in middle age females and present with similar clinical signs and symptoms (1,2,5). Taken together, the markedly higher incidence of meningioma, female predominance, similar imaging and clinical presentation all commonly lead to initial misdiagnosis of many PDLs as meningiomas (6-8).

The differential diagnosis for dural based lesions is large and includes neoplastic disease (meningioma, lymphoma, metastasis, pleomorphic xanthoanstrocytoma, hemangiopericytoma), inflamma-

tory conditions (sarcoidosis, Wegener's granulomatosis, Castleman disease), noninflammatory conditions (hematoma, pituitary apoplexy) and infectious etiologies (Tuberculosis, Aspergillosis, Tertiary Syphilis). Presence of known comorbidities (such as sarcoidosis in our patient) may act as a red herring and prevent or delay accurate diagnosis. Infectious etiologies should also be considered in the right clinical setting, especially in an immunocompromised patient.

The overwhelming majority of PDLs are marginal zone lymphomas similar to the mucosa-associated lymphoid tissue (MALT) lymphoma commonly found in gastric mucosa (1,2,5,6,9-14). MALT lymphoma of the gastric mucosa is most commonly associated with *Helicobacter pylori* infection but it has also been linked to other infections (15-18). There are sporadic case reports of MALT lymphoma associated with autoimmune diseases such as scleroderma but to date there are no reports of PDL in a patient with sarcoidosis (13,19,20). Sarcoidosis has previously been associated with multiple lymphoproliferative disorders but more recent literature suggests that this apparent link could have represented selection bias on the part of earlier studies (21). Nevertheless, more data is needed if we are to make firm associations or establish causal links between these diseases. In our patient, it is unknown what role her sarcoidosis or chronic steroid use played in the development of PDL.

Conclusion

PDL is a rare form of PCNSL, generally comprising of marginal zone lymphomas with an indolent clinical course. It may occur in patients with sarcoidosis. Here we present a case of primary dural lymphoma which was successfully treated with radiation therapy alone.

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