Intracranial Rosai-Dorfman Disease With Relapsing Spinal Lesions

A 50-year-old man developed a left lateral occipital tingling sensation, left-sided intermittent diplopia, and numbness over the left lip and cheek. Magnetic resonance imaging (MRI) demonstrated multiple extra-axial dural-based lesions (Fig 1A-1D, yellow arrows). The largest lesions were located at the left frontal convexity and both temporal regions and exhibited homogenous isointense signal intensity to gray matter on T1- and T2- weighted images, subtle hyperintensity on the fluid-attenuated inversion recovery images, and avid contrast uptake. Diffusion-weighted images, apparent diffusion coefficient values, and the exponential diffusion coefficient were all iso-intense to gray matter. Perilesional brain edema (Fig 1B, blue asterisk) and mass effect were also evident. An enhancing dural tail was present (Fig 1D, blue arrow), with no evidence of hyperostosis, calcification, or hemorrhage. Bilateral petroclival lesions that exhibited the same signal behavior were also present, with extension into the prepontine cistern and compression of the corresponding cavernous sinuses and trigeminal nerves at the Meckel’s caves (Figs 1A-1C). The smaller lesions appeared as a plaque-like enhancing focal meningeal thickening with no mass effect, edema, or hyperostosis. They were not discernable on the diffusion-weighted images, apparent diffusion coefficient images, or exponential diffusion coefficient images. Based on the imaging features, multiple meningoitases was the favored pre-operative diagnosis; however, a subsequent excisional biopsy revealed the classical histopathologic hallmarks of Rosai-Dorfman disease (RDD), including emperipolesis, which is the engulfment of lymphocytes by large RDD-like histiocytes (Fig 2A) that exhibited strong immunopositivity for CD68 (Fig 2B) and S-100 protein (Fig 2C). Eighteen months later, the patient had loss of strength in the right foot and difficulties with ambulation. Spinal MRI revealed an L1 homogeneously enhancing anterior subdural lesion compressing the cord (Fig 3B), which could be discerned retrospectively as a subtle area of enhancement at that level on the spine MRI performed 18 months earlier (Fig 3A), suggesting growth from a small precursor. Eight months after resection, a newly developed epidural lesion was noted at the L4 level (Fig 3C).

RDD, also known as sinus histiocytosis with massive lymphadenopathy, is a rare disease that was first described by Destombes in 1965 and recognized as a distinct clinicopathologic entity by Rosai and Dorfman in 1969. RDD in its classic form is a benign histiocytic proliferative disorder that is characterized by massive lymphadenopathy. Microscopically, enlarged lymph node sinuses contain prominent numbers of histiocytes with phagocytosed lymphocytes (emperipolesis; Fig 2). The mean age of onset of nodal disease is 20.6 years, with a male-to-female ratio of 1.4:1. Patients who present with or subsequently develop intracranial involvement, however, become symptomatic at a significantly later mean age (34.9 years), with a strong male predominance. The etiology and pathogenesis of this disease are still unknown. CNS involvement is rare (<5%), with 75% of cases involving the brain and 25% involving the spinal canal. Involvement of the CNS without concurrent nodal involvement is rare. Approximately 50 cases of CNS RDD have been reported in the peer-reviewed literature up to August 2007. Imaging studies typically show an enhancing meningeal-based mass with a variable amount of edema surrounding the lesion. Although a solitary dural-based lesion constitutes the most common clinical presentation, multiple intracranial lesions have also been reported. Locations can include the suprasellar region, cerebral convexity, parasagittal region, cavernous sinus, and petroclival regions. There are only 11 reported cases of spinal RDD. Ten patients had an extramedullary tumor, which was epidural in seven patients and intradural in three patients. Similar to the present case, discovery of symptomatic spinal lesions after resection of the intracranial lesions, has been reported. Based on this collective experience, it is our recommendation that meticulous spinal screening should be performed as a part of the diagnostic work-up for intracranial RDD. No specific therapy is available,
and treatment has included surgery, radiation therapy, and chemotherapy.\textsuperscript{11} Recently, a case of multiple skull-base RDD that was successfully treated with corticosteroid agents was reported, representing the first CNS RDD case to demonstrate definitive resolution of nonsurgically treated intracranial RDD after corticosteroid therapy.\textsuperscript{12} In summary, CNS RDD is a rare benign histiocytic proliferative disorder, with imaging findings typically showing a meningioma-like, dural-based extra-axial mass lesion, which can be solitary or multiple. Definitive diagnosis requires tissue examination, with characteristic histopathologic features including prominent numbers of large RDD-like histiocytes that are immunopositive for both CD68 and S-100 protein, and exhibit emperipolesis. Treatment options include surgery, radiation therapy, and chemotherapy; no specific therapy is currently available. Based on independent reports from multiple institutions, it is our recommendation that meticulous spinal screening should be performed as a part of the initial clinical evaluation of CNS RDD.

Osama Raslan  
Department of Diagnostic Radiology, National Cancer Institute, Cairo University, Cairo, Egypt
A 55-year-old black female with no past medical history came to the emergency department with right-sided chest pain and shortness of breath for the last 8 days. The patient also had anorexia, 5-lb weight loss during 1 week, and cough productive of yellow sputum for the last few days. The patient denied fever, nausea, vomiting, abdominal pain, recent travel, or sick contacts. She did not take any medications and never had a surgery. Her only hospitalization was for a vaginal delivery 30 years ago. The patient denied smoking, alcohol use, or illicit drug use. Her parents had a history of cancer but the patient did not remember the type of cancer or age at the time of their deaths. Vital signs on admission were blood pressure, 110/80 mmHg; regular heart rate, 100 beats/min; respiratory rate, 20 breaths/min; temperature, 98.6°F. Respiratory system examination revealed decreased air entry on the right side of the chest. The rest of the physical examination was unremarkable. Routine laboratory work-up showed WBC count, 8,500/μL; hemoglobin, 10.2 g/dL; platelet count, 506,000/μL; sodium,