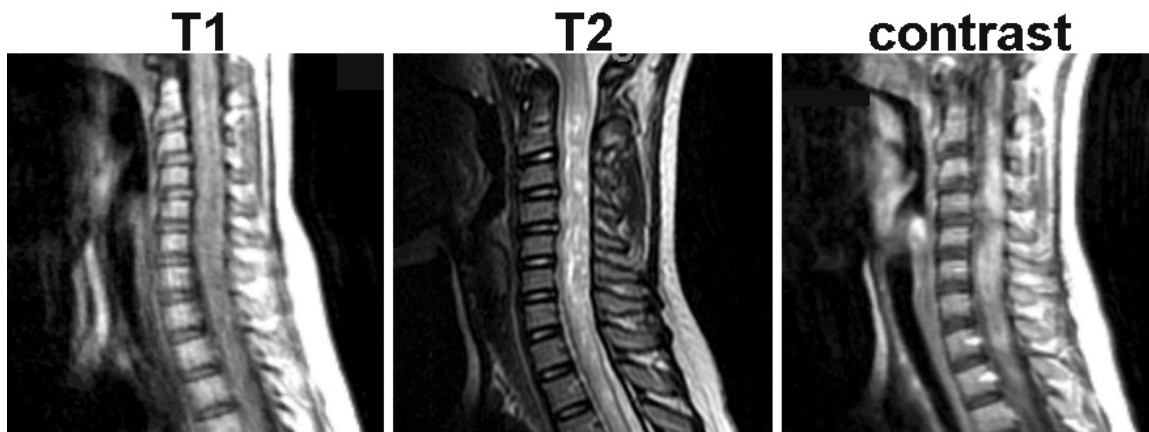


**50th ANNUAL DIAGNOSTIC SLIDE SESSION, 2009
DIAGNOSES AND REFERENCES
MODERATOR: Anthony T. Yachnis, M.D.
EDITOR: Leroy R. Sharer, M.D.**

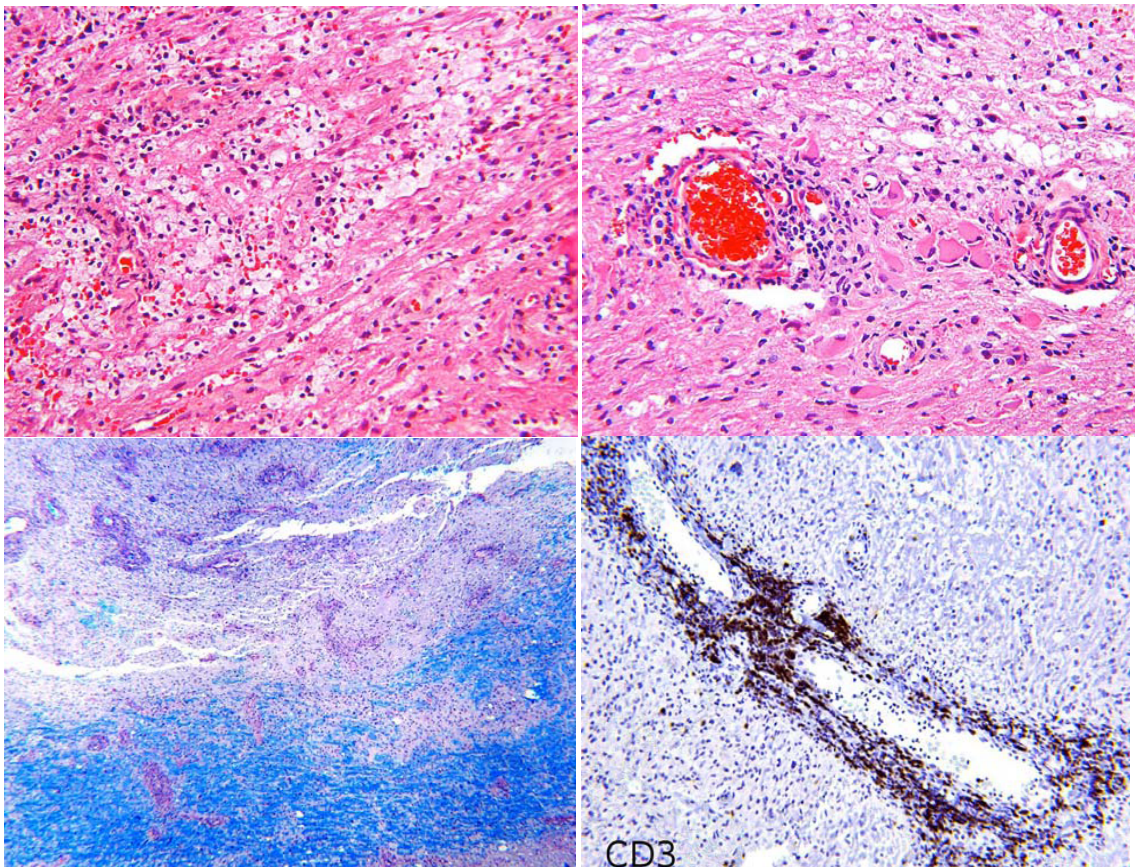
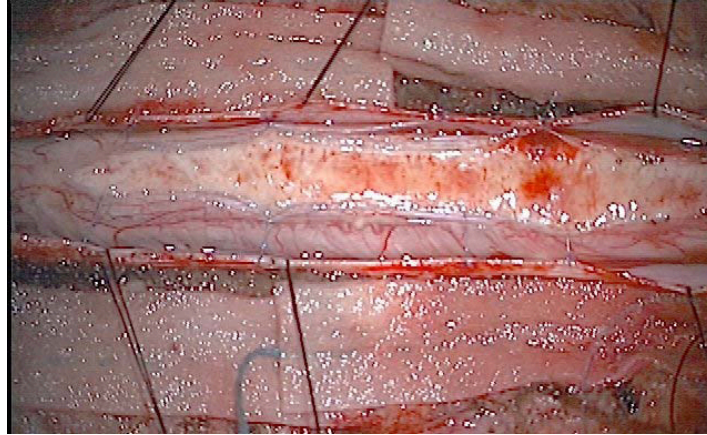
Case 2009-6

Submitted by: Craig Horbinski, Clayton A. Wiley, and Geoffrey H. Murdoch,
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The patient was a 10-year-old African-American female with a 1-year history of intermittent pain when she turned her neck and persistent rightward head tilt. More recently she developed progressive right hand weakness with paresthesias and increasing difficulty with ambulation secondary to progressive leg weakness. Physical examination was significant for profound right hand weakness and sensory impairment, and bilateral lower extremity weakness and proprioceptive impairment. Magnetic resonance imaging showed widening of the cervical spine with increased intramedullary T2 signal and contrast enhancement (see below). The abnormal T2 signal extended from the craniocervical junction to T5, and the abnormal enhancement extended from the craniocervical junction to T3. No abnormalities were seen in the remainder of the brain. Corticosteroid therapy with dexamethasone was instituted, but the patient showed no improvement.



Given the lack of steroid response and the urgent need for decompression, a C1 laminectomy and C2 through T3 osteoplastic laminotomy was performed. Intraoperative ultrasound confirmed the extent of the lesion, which upon exposure was found to be brownish gray and well-demarcated from the surrounding spinal cord, splaying the posterior columns laterally on each side. A distinct plane between the mass and the surrounding cord made gross total resection possible. Postoperatively the patient regained full strength in both upper extremities, with resolution of her symptoms and no evidence of lesion recurrence on an MRI obtained 2 months after surgery.



Diagnosis: Inflammatory pseudotumor in a patient with serum anti-aquaporin-4 antibody (neuromyelitis optica, or Devic's disease)

Comment: Various diagnoses were entertained by the attendees, including ganglioglioma, an infectious process and an inflammatory or lymphomatoid disorder. The lesion on histological examination had patchy loss of myelin on LFB stain, with apparent preservation of some axons, with GFAP-positive gemistocytic cells, macrophages that were positive for CD68, and mainly CD3 positive T cells, with a few CD20 positive B cells. The presenter and his colleagues entertained a diagnosis of "inflammatory demyelinating pseudotumor," until they received a report from Mayo Clinic, that the anti-aquaporin-4 antibody was found in a specimen of the

patient's blood. The patient had been previously worked up for a "bad eye," information that was not known at the time of surgery. Aquaporin-4 is expressed in spinal cord and optic nerve. The prognosis of neuromyelitis optica (NMO) is usually better in children than in adults, and recurrent attacks are more common in females. This girl has had an aggressive course of the disease, and she had relapsed four times since surgery was performed in 2007.

References:

Banwell B, Tenenbaum S, Lennon VA, et al: Neuromyelitis optica-IgG in childhood inflammatory demyelinating CNS disorders. *Neurology* 2008; 70:344-352.

Lucchinetti CF, Mandler RN, McGavern D, et al: A role for humoral mechanisms in the pathogenesis of Devic's neuromyelitis optica. *Brain* 2002; 125:1450-1461.

Wingerchuk DM, Hogancamp WF, O'Brien PC, Weinshenker BG: The clinical course of neuromyelitis optica (Devic's syndrome). *Neurology* 1999; 53:1107-1114.

Note: Dr. Horbinski received the fifth annual O.T. Bailey-Helena Riggs Award for best presentation by a trainee at the Diagnostic Slide Session, selected by a vote of the Charter Members of the Diagnostic Slide Session and presented at the awards ceremony of the American Association of Neuropathologists on June 14, 2009.