50th ANNUAL DIAGNOSTIC SLIDE SESSION 2009

CASE 2009-6

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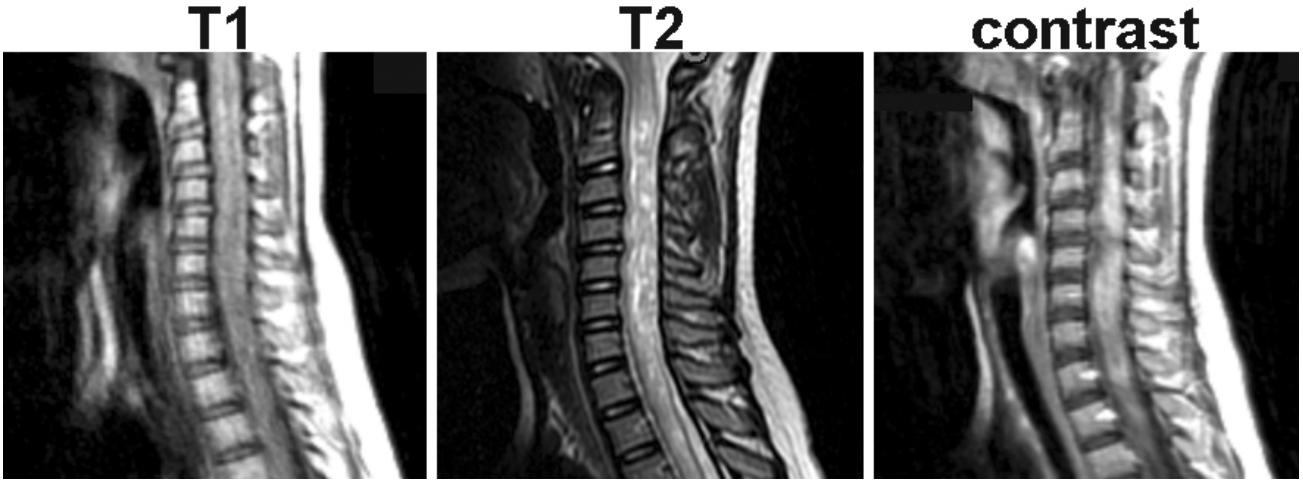
Clinical History: The patient was a 10-year-old female with a 1-year history of intermittent pain when she turned her neck and persistent rightward head tilt. More recently, she developed progressive right-hand weakness with paresthesias and increasing difficulty with ambulation secondary to progressive leg weakness.
Physical examination was significant for profound right-hand weakness and sensory impairment, and bilateral lower extremity weakness and proprioceptive impairment.
MRI 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.



Material Submitted:

1.) 1 H&E-stained section of the spinal lesion
2.) Virtual slides of the intraoperative smear preparation and permanent sections (http://image.upmc.edu:8080/NeuroPathology/Murdoch/Slide.8/view.apml?cwidth=1003&cheight=598&chost=image.upmc.edu:8080&csis=0)

Points for Discussion:

1.) Additional special stains and studies
2.) Diagnosis
3.) Pathogenesis

References:

1.) Wingerchuk DM, Hogancamp WF, O'Brien PC, Weinshenker BG. The clinical course of neuromyelitis optica (Devic's syndrome). *Neurology* 1999 Sep 22; 53(5): 1107-14.
2.) Lucchinetti CF, Mandler RN, McGavern D*, et al.* A role for humoral mechanisms in the pathogenesis of Devic's neuromyelitis optica. *Brain* 2002 Jul; 125(Pt 7): 1450-61.
3.) Banwell B, Tenembaum S, Lennon VA, et al. Neuromyelitis optica-IgG in childhood inflammatory demyelinating CNS disorders. Neurology 2008 Jan 29; 70(5): 344-52.