Pearls & Oy-sters: Clues for spinal dural arteriovenous fistulae

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PEARLS In a patient presenting with progressive myelopathy, a spinal dural arteriovenous fistula (SDAVF) should be among the diagnoses considered.[1] Clinical symptoms may be acutely exacerbated by hydrostatic forces resulting from erect posture,[1] abdominal compression,[2] and the Valsalva maneuver[3] and (as described in our patients) by corticosteroid treatments.[4,5]

MRI findings include abnormally dilated cord vascularity and longitudinally extensive parenchymal signal abnormalities. The parenchymal MRI findings are observed in the lower spinal cord and conus medullaris in 87% of patients and frequently also extend into higher thoracic and cervical regions.[1] The pathogenesis of SDAVF-associated myelopathy (leading to the radiologic parenchymal abnormalities) is cord edema and ischemia caused by arterialized venous hypertension.[6]

OY-STERS The parenchymal cord abnormalities on MRI in patients with myelopathy due to SDAVF taken in isolation may be mistaken for longitudinally extensive transverse myelitis (LETM), a characteristic finding in patients with neuromyelitis optica (NMO) spectrum disorders,[7] or tumor. A thorough clinical history, careful review of spinal MRI for dilated vessels, and spinal angiography in suspicious cases are critical to making the diagnosis of SDAVF.

CASE REPORTS All patients were referred to the Department of Neurology (Multiple Sclerosis and Autoimmune Neurology Division), Mayo Clinic, for management of LETM, and all were ultimately found to have an SDAVF as the cause of myelopathy. All were seronegative for NMO–immunoglobulin G, an antibody marker of NMO, and limited forms of the disorder (relapsing myelitis, usually longitudinally extensive on MRI, and relapsing optic neuritis). Reported worsening in symptoms after corticosteroid treatment provided additional clues.

Patient 1. Patient 1, a 52-year-old man, presented with progressive lower extremity weakness and bladder symptoms. A longitudinally extensive spinal cord lesion from T1 to the conus medullaris was noted on MRI. Evaluation also included normal results for spinal angiography (of the lumbar region only). A diagnosis of LETM was made. The patient experienced acute worsening of leg weakness on both occasions when he received corticosteroids: oral prednisone (he improved to walking with a walker on cessation of steroids) and a course of IV methylprednisolone (he became paraplegic). He then received 9 plasma exchange (PLEX) treatments and improved to walking with a walker again. His symptoms stabilized for 3 months, but he had progressive deterioration thereafter. After 9 months of symptoms, the patient (wheelchair-bound) presented to the Mayo Clinic. Radiologic review revealed abnormal flow voids surrounding the spinal cord and longitudinally extensive parenchymal abnormalities (P1, figure). Angiography (entire spine and brain) demonstrated an SDAVF fed by the left ascending pharyngeal artery and drained via a large intradural spinal vein. Six months after suboccipital craniectomy with obliteration of the dural arteriovenous fistula, the patient had radiologic and sensory neurologic improvements but was still wheelchair-bound.

Patient 2. Patient 2, a 51-year-old woman, who had one episode of optic neuritis 14 years previously, developed a slowly progressive myelopathy over 3 months. The patient had rapid deterioration in walking twice (at 3 and 6 months into her illness) after high-dose methylprednisolone treatment for presumed transverse myelitis. She had a confluent area of cord signal abnormality on MRI from T6 to the conus medullaris. After 18 months of symptoms, the patient presented to Mayo Clinic for evaluation.

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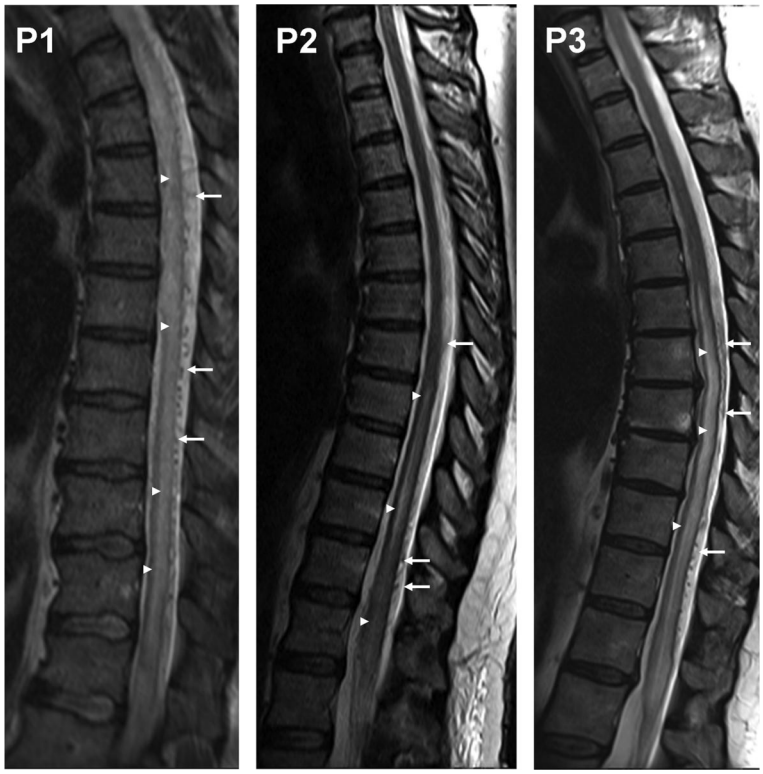
Figure Sagittal T2-weighted images demonstrating abnormal vascular flow voids in the dorsal aspect of the thecal sac suggestive of a dural arteriovenous fistula, in patient (P) 1, 2, and 3 (arrows)



Longitudinally extensive parenchymal cord abnormalities are demonstrated (arrowheads).

She had a flaccid paraplegia on examination, and marked, abnormal vascularity was additionally noted on spinal cord MRI (P2, figure). Angiography revealed a SDAVF arising from the left L3 lumbar artery, disconnection of which provided radiologic improvement only at 6 months.

Patient 3. Patient 3, a 58-year-old woman, had progressive loss of sensation and strength of her lower extremities over 5 months, remaining ambulatory without aid. An outside neurologic evaluation was pertinent for myelopathic findings on examination, and MRI revealed a longitudinally extensive cord lesion from T4 to the conus medullaris. A diagnosis of LETM was made and within 24 hours of receiving the first of 5 doses of intravenous methylprednisolone, the patient discontinued the therapy because of rapid deterioration to needing a cane to walk. Oral prednisone was initiated on 2 occasions over the following month but was discontinued each time within 4 days because of worsening weakness. Further spontaneous improvements ensued before her Mayo Clinic evaluation, 7 months into her illness. MRI of her spine demonstrated a prominent thoracic cord and conus flow voids, in addition to parenchymal changes (P3, figure). Spinal angiography demonstrated an SDAVF at L2, subsequently treated surgically elsewhere; the patient had gradual improvement to walking without a cane.

DISCUSSION These patients, originally considered to have LETM, illustrate key diagnostic features that could have led to early diagnosis of SDAVF: 1) a progressive rather than subacute course; 2) the presence of abnormal, dilated vessels surrounding the cord on standard T2 sagittal MRI with subsequent confirmation of SDAVF with angiography; and 3) acute worsening with corticosteroid therapy. Worsening of paraplegia with corticosteroids has been previously reported in 2 patients with SDAVF.[4,5] This phenomenon may be caused by an increase in shunting across the SDAVF because of the high intravascular volume with rapid saline infusion[5] or perhaps (we speculate) by the hypertensive effects of hypercortisolemia (patients 1 and 3 reported deterioration with oral prednisone). The clinical improvement noted by patient 1 after PLEX may have occurred because of relative volume depletion and hypotension[8] (both may occur with PLEX), thereby reducing venous engorgement and cord edema. Comprehensive spinal (and even cerebral) angiography may be required to find the SDAVF, which may be remote from observed cord parenchymal abnormalities, as demonstrated in the case of patient 1.

DISCLOSURE

Dr. McKeon receives research support from the Guthy-Jackson Charitable Foundation. Dr. Lindell estimates that 10% of his clinical effort is spent performing and interpreting spine MRI and spinal angiography. Dr. Atkinson serves on the editorial board of *Endocrine Practice*. Dr. Weinshenker serves on data safety monitoring boards for Novartis and Biogen Idec, serves on the editorial boards of the *Canadian Journal of Neurological Sciences* and the *Turkish Journal of Neurology*; has received research support from Genzyme Corporation and the Guthy-Jackson Charitable Foundation; and receives license royalties from RSR Ltd. for a patent re aquaporin-4–associated antibodies for diagnosis of neuromyelitis optica. Dr. Piepgras reports no disclosures. Dr. Pittock may accrue revenue for patents re aquaporin-4–associated antibodies for diagnosis of neuromyelitis optica and aquaporin-4 autoantibody as a cancer marker and has received research support from Alexion Pharmaceuticals, Inc. and the Guthy-Jackson Charitable Foundation.

REFERENCES

1. Atkinson JL, Miller GM, Krauss WE, et al. Clinical and radiographic features of dural arteriovenous fistula, a treatable cause of myelopathy. Mayo Clin Proc 2001;76:1120–1130.
2. Madsen JR, Heros RC. Spinal arteriovenous malformations and neurogenic claudication: report of two cases. J Neurosurg 1988;68:793–797.

3. Khurana VG, Perez-Terzic CM, Petersen RC, Krauss WE. Singing paraplegia: a distinctive manifestation of a spinal dural arteriovenous fistula. Neurology 2002;58:1279–1281.
4. Soderlund ME, Benisty S, Gaston A, Djindjian M, Cesaro P, Creange A. Can myelopathies secondary to arterio-venous dural fistulae be aggravated by intravenous corticosteroid therapy? Rev Neurol 2007;163:235–237.
5. Cabrera M, Paradas C, Marquez C, Gonzalez A. Acute paraparesis following intravenous steroid therapy in a case of dural spinal arteriovenous fistula. J Neurol 2008;255:1432–1433.
6. Aminoff MJ, Barnard RO, Logue V. The pathophysiology of spinal vascular malformations. J Neurol Sci 1974;23:255–263.
7. Wingerchuk DM, Lennon VA, Lucchinetti CF, Pittock SJ, Weinshenker BG. The spectrum of neuromyelitis optica. Lancet Neurol 2007;6:805–815.
8. Rodnitzky RL, Goeken JA. Complications of plasma exchange in neurological patients. Arch Neurol 1982;39:350–354.

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