protective agents in ALS,[5] a logical extension would be to use these drugs in clinical trials. Because most of the currently available COX-2 antagonists exhibit only mild CNS penetrability, it is crucial that these anticipated studies have a way to demonstrate that the administrated COX-2 antagonist actually reaches the CNS and inhibits the enzyme.

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Singing paraplegia: A distinctive manifestation of a spinal dural arteriovenous fistula

Abstract—The unique case of a baritone with a spinal dural arteriovenous fistula (SDAVF) causing recurrent, acute paraplegia during singing is described. This case underscores the presence of impaired venous drainage in these lesions and the high level of clinical suspicion required for their diagnosis in patients with any myelopathy.

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Patients with spinal dural arteriovenous fistulae (SDAVF) typically have slowly progressive paraparesis that may be exacerbated by upright posture, ambulation, pregnancy, and menstruation.[1-5] Because of the relative rarity of these lesions and slow progression of the myelopathy, symptoms may be misinterpreted by patients and physicians for years.[4,6,7] This may lead to delayed diagnosis and treatment with irreversible and avoidable neurologic damage.[2,4,8] In many patients, surgery is curative. We report the case of a baritone with a 10-year history of progressive myelopathy punctuated, in the last 3 years, by acute, recurrent paraplegia while singing. This rare pattern of clinical deterioration, and its association with singing, is a previously undocumented presentation of a SDAVF.

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Case report. A 72-year-old professional baritone was referred for evaluation of a 3-year history of his "legs suddenly giving way" when singing while standing. His symptoms had become so prominent in the 6 months preceding his referral, that to avoid collapsing during his performances, he had resorted to singing while seated. In fact, 10 years before his referral the patient noted the insidious onset of bilateral leg weakness when bending forward, although for the first several years this did not interfere with his activity as a singer. The patient reported no pain and no bowel or bladder dysfunction; however, he reported chronic mild numbness and paresthesia in both feet. His medical history included stable coronary artery disease, benign idiopathic thrombocytopenic purpura, diabetes mellitus, and hypertension. During the 6 months before referral, the patient had undergone several examinations with MRI, including head (normal) and total spine imaging. A lumbosacral spine MRI showed mild vertebral canal narrowing at L4-L5. A thoracic spine examination with MRI showed T2-signal hyperintensity in his mid-to-lower thoracic cord initially interpreted as "nonspecific" and coincidental with mild intervertebral disc herniations from T7-T9. The patient was referred to our institution for further evaluation.

Clinical and radiologic evaluation. At the time of our evaluation, the patient was able to walk independently.

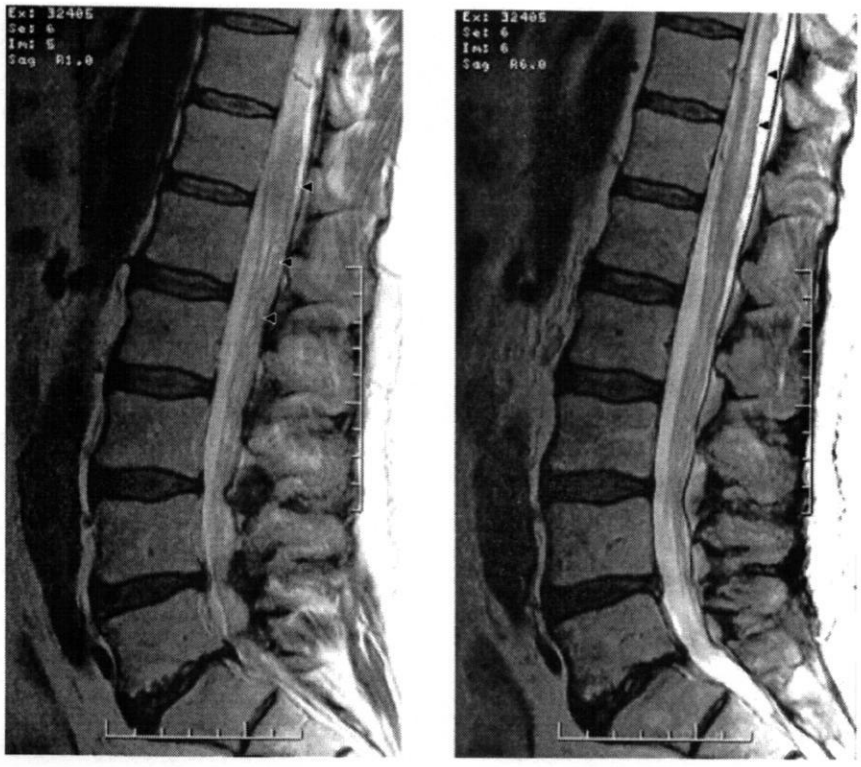

Figure 1. T2-weighted preoperative thoracolumbar MRI. (left) An intrathecal serpentine draining vein (arrowheads) coursing over the posterior aspect of the distal spinal cord. (right) Areas of intramedullary T2-weighted signal hyperintensity (arrowheads). Scale bars = 1 cm/division.

Physical examination revealed only mild symmetric sensorimotor deficits in his legs; the remainder of the neurologic exam was normal. An MRI scan of the thoracolumbar spine showed T2-weighted signal abnormality extending from T7 to the conus medullaris, associated with mild distal thoracic cord enlargement, posterior intrathecal flow voids, and a serpentine intradural vein ascending in the midline posteriorly (figure 1). There was mild heterogeneous enhancement with gadolinium. A spinal angiogram revealed a typical spinal dural arteriovenous fistula immediately inferior to the right L1 pedicle and supplied principally by the dural branch of the right L1 lumbar artery (figure 2). The fistula drained into the conus through a large arterialized vein.

Treatment and outcome. After an L1 laminectomy, the right L1 pedicle was identified and the dura opened in the midline. On the dural sleeve immediately below the right L1 nerve root, we found an arterialized vein entering the subarachnoid space through the dura. We identified the dural nidus, which, together with the proximal arterialized vein, was coagulated. We divided the arterialized vein, thereby obliterating the fistula. The patient left the hospital 2 days after his surgery and was walking independently. Clinical examination at 3 months after surgery showed a remarkable recovery of function: the patient was able to sing while standing throughout almost his entire performance, and his strength was continuing to improve. A repeat thoracolumbar MRI scan performed at this time showed marked reduction in the caliber of the previously swollen distal cord, absence of previously visible intradural flow voids, and decreased intramedullary T2-weighted signal abnormality. At 1 year after surgery, the patient stated that he no longer experienced any unsteadiness related to his previous leg weakness and was now able to stand and sing for as long as necessary.

Discussion. The clinical presentation of a SDAVF causing acute episodes of paraplegia associated with singing, as in the current patient, is unique in our experience. Despite the widely studied clinical features of these lesions, we did not find any similar reports in the literature.

SDAVF are vascular malformations characterized by an abnormal nidus of vessels in the spinal dura fed by dural branches of radicular arteries. They drain into an arterialized vein.[1,3,6-8] In turn, retrograde flow occurs into the coronal venous plexus of the spinal cord. These features were apparent radiologically and intraoperatively in our patient. The clinical findings in these lesions—i.e., a slowly progressive myelopathy[1,3,4,6,7]—are caused by chronic venous hypertension associated with postnidal venous engorgement and intraparenchymal cord edema and ischemia (hence the T2-signal hyperintensity on MRI); the pathologic hallmark is that of a subacute necrotizing myelopathy.[6,7] Interestingly, despite the location of the fistula itself, the MRI T2-signal changes almost always involve the lower cord, often attributed to orthostasis and correlated with exacerbation of symptoms during ambulation.[4] In our patient, the acute paraplegia during singing was most likely caused by acutely increased venous pressure associated with the extremes of respiratory exertion, similar to a Valsalva maneuver.[4,9,10] This exacerbated venous engorgement, cord edema, and cord ischemia. This feature highlights the effect of acutely increased venous hypertension in a lesion whose pathogenesis involves chronically impaired or "marginal" venous drainage.[5]

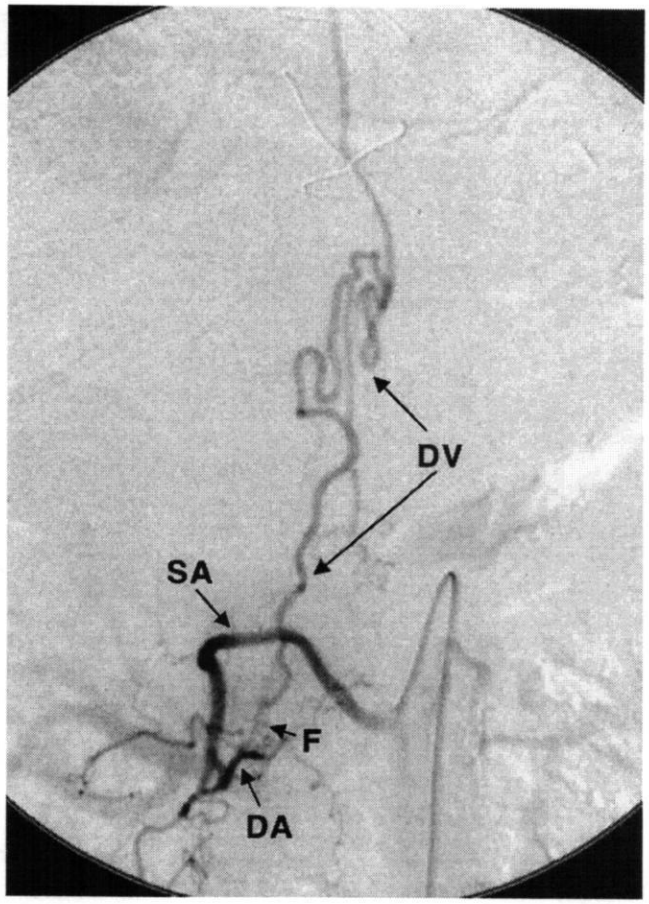

Figure 2. Preoperative spinal angiogram. The right L1 segmental artery (SA) and its distal dural artery (DA) branch. The nidus of the fistula (F), fed by the dural artery, empties into a serpentine draining vein (DV). The fistula was surgically disconnected through cautery and division of the nidus and proximal draining vein.

The diagnosis of a spinal vascular malformation as a cause of myelopathy remains challenging because of the relative rarity of these lesions and their typically insidious presentation and slowly progressive clinical course.[7,8] Furthermore, as in the current patient, symptoms caused by SDAVF often have been inadvertently attributed to intervertebral disc herniation, vertebral canal stenosis, Guillain–Barré syndrome, subacute combined degeneration, transverse myelitis, or a spinal cord neoplasm.[4,6] As a result, treatment is frequently delayed, often for years, leading to avoidable irreversible neurologic deficits.[4,6,7] For this reason, a high level of clinical suspicion for SDAVF is required in patients with any myelopathy, particularly as these lesions are amenable to safe and curative surgical disconnection.[1-4,6,8]

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