Clinical Case of the Month

Intracranial dural arteriovenous fistula causing a myelopathy

I Perakash^{*,1,2,3}, V Punj¹, DT Ota^{1,3}, B Lane⁴, SL Skirboll⁵

¹Spinal Cord Injury Service, Veterans Affairs Health Care System, Palo Alto, California, CA 94305, USA;

²Department of Urology, Stanford University, Palo Alto, California, CA 94305, USA; ³Department of Physical Medicine and Rehabilitation, Stanford University, Palo Alto, California, CA 94305, USA; ⁴Department of Radiology, Stanford University, Palo Alto, California, CA 94305, USA; ⁵Department of Neurosurgery, Stanford University, Palo Alto, California, CA 94305, USA

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Case presentation

Professor Inder Perakash, Dr Vandana Punj and Dr Doug Ota

The patient is a 79-year-old male with a past medical history of atrial fibrillation and diabetes mellitus who presented in March 2000 to a Neurologist with a 6-month history of progressive bilateral lower extremity weakness, urinary retention, and constipation with occasional bowel incontinence. He also noted that his symptoms were worse standing or walking and improved with lying down or while using his stationary bicycle. His initial work-up included a radiograph of the lumbar spine that was unremarkable. Thyroid stimulating hormone level, creatinine phosphokinase level, vitamin B12 level, and erythrocyte sedimentary rate were normal. The patient did not pursue further work up including an MRI at that time. He then presented to the Palo Alto Veterans Affairs Hospital on 15 April, 2000 with a complaint of inability to walk or void after a fall while getting out of bed. On examination, the patient's vital signals were: blood pressure, 146/92; pulse, 104; temperature, 36.2°C and finger stick blood sugar, 214 mg/dl. He was alert and cooperative. Speech was fluent and appropriate. Cranial nerves II to XII were intact. Sensory exam revealed pin prick intact up to T6 and impaired below. On motor examination the patient had 4/5 strength in the upper extremities, 2/5 hip flexors and hip extensors and 3/5 knee flexion, knee extension, ankle dorsiflexion and ankle plantar flexion. Reflexes were 3/4 bilaterally in the upper and lower extremities. Rectal tone was slightly decreased. Prostate examination revealed a hypertrophied prostate without palpable nodules. The patient had 1200 cc of urine in his bladder by catheterization. An emergent MRI of the complete spine was done. At

that time, there was moderate cervical stenosis seen at C3–4 as well as some evidence of a compression fracture in the thoracic spine.

However, there was no evidence of acute cord compression and no initial evidence of cord signal changes. A lumbar puncture was done which showed normal opening pressure, three white blood cells and no red blood cells, with a slightly elevated protein at 75 mg/dl, and a glucose of 88 mg/dl. A repeat thoracic MRI obtained on 24 April 2000 demonstrated signal abnormality of the cervical cord on the scout film. A cervical MRI obtained the next day demonstrated extensive cord edema, with serpiginous enhancing structures on the surface suggestive of a dural fistula (Figure 1). Multiple angiograms revealed an arteriovenous (AV) fistula at the skull base with feeders identified from the right vertebral artery (Figure 2), right external carotid artery, probably the ascending pharyngeal branch (Figure 3) and left external carotid artery, probably the posterior auricular branch (Figure 4). The myriad of arteries feeding the fistula appeared to terminate at the right petrous apex and drained into the right jugular vein. Spinal venous congestion, due to high-pressure flow into the jugular vein, resulted in cervical myelopathy.

Due to the complex anatomy and risk, the patient refused intervention other than medical treatment. Therefore no surgical or vascular interventions were offered. The patient underwent a comprehensive rehabilitation program at the spinal cord unit. At discharge the patient was independent in activities of daily living. He was able to ambulate up to 300 feet using a front wheel walker and required a manual wheelchair for community mobility. The patient's bowel program was digital stimulation every other day and his bladder was managed with a Foley catheter.

Figure 1 T2 weighted MRI of the cervical spine showing abnormal cord signal from C2–T5, and serpiginous enhancing structures on the surface

Questions and responses

- (1) What was the initial differential diagnosis and how did this change during his hospitalization?

Doug T Ota, MD

The repeat MRI obtained approximately one and a half weeks after admission demonstrated significant changes. There were extensive signal abnormalities extending from C2–T5, which followed the gray matter. The cord was not atrophic in appearance. The changes on the MRI were thought to be consistent with infarct or transverse myelitis. Although anterior spinal artery syndrome was considered, the extensiveness of involvement was beyond the distribution of the anterior spinal artery. However a dural AV

Figure 2 Carotid angiograms showing the AV fistula at right skull base with flow from the right vertebral artery

Figure 3 AV fistula with feeders from the right external carotide artery (ECA) (ascending pharyngeal branch)

fistula would be consistent with the patient's symptoms and MRI findings. Therefore angiography of the aortic arch, abdominal aorta, intercostals, subclavian and right vertebral artery was performed which

demonstrated a dural fistula at the skull base and serpiginous enhancement on the surface of the cord.

Figure 4 AV fistula with feeders from the left external carotid artery (ECA) (posterior auricular branch)

Figure 5 Post contrast T1 weighted MRI of the cervical spine showing dilated spinal veins on the surface of the cord (arrows)

The feeders to the fistula could not be clearly identified. With the changes demonstrated by angiography and a follow-up MRI that showed dilated veins (Figure 5) and serpiginous structures on the edematous cord, the differential diagnosis became consistent with a myelopathy secondary to a fistula. Also at this time, during an evaluation with occupational therapy, the patient reported that he would become dizzy and hearing a ‘whooshing’ sound in his right ear when lying on his right side. A second angiography was performed to investigate the vertebral and carotid vessels, and demonstrated a fistula involving feeders identified from the right vertebral artery, right external carotid artery (probably the ascending pharyngeal branch), and left external carotid artery (probably the posterior auricular branch) to terminate at the right petrous apex and drained into the right jugular vein. The diagnosis of an intracranial AV fistula, draining through anastomotic connections and producing a myelopathy of the cervical and thoracic spinal cord was thus confirmed.

(2) How do you account for the improvement in the patient’s status?

Doug T Ota, MD

Case reports and retrospective reviews have demonstrated a reversal of the pathophysiology of a myelopathy due to AV fistulas^{1–3} with subsequent stabilization or improvement of the functional status.^{4,5} However, in this case the patient was not keen for any interventions that would interrupt the dural fistula, including surgical resection or embolization. On admission, the patient’s manual motor testing was 2/5 hip flexion and hip extension; 3/5 knee flexion, knee extension, ankle dorsiflexion and ankle plantarflexion bilaterally. Functionally he required moderate assistance for supine to sit; maximum assistance for sit to stand; and was dependent with ambulation. On discharge his manual motor testing was 3/5 hip flexion and knee extension and 4/5 knee flexion bilaterally. Functionally he required minimum assistance for supine to sit; contact guard assistance for sit to stand; moderate assistance for floor to chair, and stand by assistance for ambulating 50–300 feet using a front wheel walker. The patient’s neurological status stabilized without further decline, despite the lack of intervention to interrupt the fistula. The patient’s management for the fistula was limited to medical management. This included strict control of his type II diabetes that had been diagnosed prior to admission but had not been followed for over a year. His blood pressure was also closely controlled, although by discharge, he required no pharmacological management to maintain blood pressure within normal limits. The control of blood pressure is a factor, as change in systemic arterial pressure has direct transmission to the spinal cord without the normal dampening effect of the venous plexus. His heart rate remained controlled with digoxin.

The patient participated in a full rehabilitation program that included physical therapy, which began with pre-gait training and eventually progressed to gait activities through range of motion, stretching and strengthening exercises. He had occupational therapy to address self-care activities including feeding, bathing, dressing, toileting, transfers and wheelchair propulsion for community level mobility. Another important component was intervention and treatment by the psychosocial team to assist with coping mechanisms, as depression and decreased motivation were barriers to progress in his rehabilitation program. Although the patient did not have a definitive correction of the pathology that produced the myelopathy, close medical management to correct factors that may contribute to his condition combined with a comprehensive rehabilitation program produced an outcome of functional gains by discharge.

- (3) How does this patient's fistula compare and differ from a typical dural fistula?

Doug T Ota, MD

The typical arteriovenous fistula is located within the dural sleeve of the nerve root. Frequently, the location is in the thoracic or lumbar nerve root. The vessels involved are the intradural radicular artery and vein. The shunting is single rather than a mass of angiomatous tissue, which is consistent with a fistula rather than an AVM. The pathophysiology proposed is venous hypertension with subsequent increase in congestion of the coronal plexus that drains the cord parenchyma.^{6–8} The increased pressure within the parenchyma produces ischemia by decreasing arteriole perfusion. In this particular case, the vascular anomaly was located at a distance away from the cord in an intracranial location. There were multiple feeder vessels involved with the fistula, and the venous outflow involved vessels that were not directly associated with the spinal cord venous drainage. However, through a complex series of anastomotic venous channels the anterior spinal vein does connect to the petrosal vein. The resultant increase in venous hypertension ultimately reaches the cord parenchyma via the valve-less anastomotic channels and produces a myelopathy similar to the typical dural AV fistula. In both cases the patient is middle-aged or older. Symptoms include bowel and bladder dysfunction, sensory changes, and weakness, especially in the lower extremities. Early identification and treatment is important as intervention in either case can often prevent further neurological decline or reverse the myelopathic changes.

- (4) What is the etiology, symptomatology and the natural history of AV fistulas?

Barton Lane, MD

Etiology of dural arteriovenous fistulas (in the brain or spinal cord) is usually not clear. They may arise spontaneously in older adults (40–60 years of age),

often without antecedent history: but predisposing factors may include trauma some years earlier with symptoms developing later in life. Other factors could include prior surgery, or prior thrombophlebitis involving the venous sinuses (perhaps undiagnosed). Most often, no etiology is discovered.

Symptoms at presentation may include: tinnitus, bruits, cranial nerve palsies, neurological deterioration, dementia and (if in the spinal cord), progressive paraplegia.

Natural history: If not treated, these fistulas worsen over time. There are rare cases of spontaneous cure.

- (5) What therapeutic options are available for the management of dural AV fistulas?

Stephen L. Skirboll, MD

The therapeutic options for dural AV fistulas include conservative therapy, compressive maneuvers in the neck, transarterial or transvenous embolization, surgical sinus exposure and intraoperative embolization, direct surgical resection of the lesion, and a combination of the above. The decision to treat these lesions depends on both the clinical profile and the angiographic characteristics. For example, a lesion that appears both clinically and angiographically 'benign' may not need any treatment. In contrast, those patients who present with hemorrhage and/or neurological deficit, as well as with angiographically determined risk factors (for example, cortical and dilated venous structures) require definitive therapy. In this patient, the dural AV fistula did cause a neurological deficit, however, he refused to undergo invasive treatment. If this patient had reconsidered, I would discuss this with our interventional neuroradiology team to determine if this could be adequately treated by an endovascular approach. If this was not possible or only partially treated, direct surgical treatment could have been considered, which would include isolation of the fistula, complete excision of the fistula, or disconnection of arterialized leptomeningeal venous drainage from the site of the fistula. Such surgery can be greatly facilitated by preoperative embolization. One should also mention that radiosurgery may be a reasonable option, though our experience with this as a definitive treatment is relatively new. This treatment has the potential to obliterate the dural AV fistula, albeit in a slow process over 1 or 2 years. This could be combined with arterial embolization (after the radiosurgery) in certain cases to possibly reduce some of the symptoms in a more immediate fashion.

Discussion

Professor Inder Perakash, Vandana Punj, MD and Doug T Ota, MD

Spinal arteriovenous malformations and fistulas have been documented and categorized as causes of

myelopathies. Anson and Spetzler classified spinal cord vascular malformations into four categories in 1992.⁹ Their classification categorized arteriovenous malformations into dural lesions or intradural lesions with intramedullary and extramedullary locations. The myelopathy is produced as a result of increased blood flow from the high-pressure arterial system through the fistula into the lower pressure venous system. Due to the valve-less nature of the spinal venous plexus, backward flow can occur, producing venous congestion with resultant hemorrhage or ischemia. Most commonly the congestion is produced from connections between a spinal radicular artery and a spinal medullary vein (Type I dural AV Malformation). Other authors have described myelopathy due to intracranial dural AV fistulas from branches of the carotid arteries draining into spinal arteries.^{10,11} In this particular case venous congestion of the spinal medullary veins was produced from a more distant fistula involving feeders identified from the right vertebral artery, right external carotid artery, and left external carotid artery terminating at the right petrous apex and draining into the right jugular vein. Although the location of this patient's vascular anomaly was intracranial and extradural, clinically the patient presented in a pattern that is similar to a type I dural AVM. Similar to our case, patients with type I AVM's are usually over 40 years of age with progressive symptoms over a period of months including progressive lower extremity weakness, urinary retention and constipation with occasional bowel incontinence. Our case report illustrates the complex and varied nature of arteriovenous malformations involving the spinal and cranial vasculature that can lead to a myelopathy.

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