

Clinical Reasoning: A 59-year-old woman with acute paraplegia



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SECTION 1

Case presentation. A 59-year-old woman with a history of hypertension developed acute bilateral flaccid leg weakness while watching television. She had shifted her weight while sitting on the couch and suddenly felt a sharp pain in her lower back and right leg. When she stood up to walk she noticed that her legs were numb; over the course of 1 hour she became unable to move her legs. She could not urinate voluntarily, and had dribbling incontinence. She was brought to the emergency department for evaluation.

Questions for consideration:

1. Where does acute paraplegia localize and what is the differential diagnosis?
2. What features of the history help make certain entities more or less likely?

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SECTION 2

When evaluating a patient with acute bilateral leg weakness, the concern for a possible neurosurgical emergency requires prompt diagnosis and treatment. The etiology can be considered as one addresses the possible locations for the lesion. An anatomic approach begins with the most central causes. A parasagittal lesion in the brain, whether compressive or vascular, may cause predominant lower extremity dysfunction by involving the motor cortex bilaterally. A pontine lesion may cause leg weakness through involvement of select corticospinal fibers. A spinal cord lesion at any level may cause isolated lower extremity weakness and sensory loss, since the lamination pattern of the ascending and descending tracts places the leg fibers peripherally where they are vulnerable to external compressive lesions throughout their course. Another important location to consider is the cauda equina, which may be damaged by extrinsic compression or by inflammatory and infiltrative processes. Acute demyelinating radiculopathy and neuropathy, such as Guillain-Barré syndrome, must be considered. Peripheral causes of weakness include disorders of muscle or neuromuscular junction. Finally, psychogenic disorders such as conversion reaction must be considered, but only as a diagnosis of exclusion.

The constellation of bilateral leg weakness, radiating back pain, and overflow urinary incontinence suggests a lesion within the lower spinal cord or cauda equina. The presence of numbness makes a process involving muscle or neuromuscular junction untenable. The time course hints toward the possible etiology. The hyperacute onset of her symptoms is concerning for a compressive or vascular etiology. An inflammatory process of the cord would typically have a more subacute, escalating presentation.

In this patient, examination revealed complete paralysis of the legs proximally and distally, with the exception of 2/5 left toe strength. Tone was diminished in the legs symmetrically, and normal in the arms. Sensation to light touch, vibration, pinprick, and temperature was absent below the umbilicus on the right and decreased on the left. Joint position sense at the toes was impaired bilaterally. Deep tendon reflexes were absent in the lower extremities and 2+ in the upper extremities. Plantar responses were extensor bilaterally. She had decreased rectal tone. The remainder of the general and neurologic examination was unremarkable.

Questions for consideration:

1. How does the examination modify the differential, and help guide the workup?
2. What testing would you obtain at this point?

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Figure 1 T2-weighted thoracic spine MRI



T2-weighted thoracic spine MRI, revealing central cord T2 hyperintensity from T4 to T8 (black arrow) and suggesting prominent flow voids within and on the surface of the cord (white arrow).

SECTION 3

The pattern of severe lower extremity weakness, sensory loss, hyporeflexia, and extensor plantar responses is most consistent with acute myelopathy. The likely location of the lesion is the lower thoracic cord based on the sensory level at the umbilicus. Diminished sensation of temperature, vibration, and joint position implies involvement of both the anterolateral system and the posterior columns. Since these sensory tracts receive different arterial blood supplies, the lesion extends beyond one arterial distribution, which makes an isolated anterior spinal artery infarction less likely. By this reasoning, the most likely etiology is extrinsic spinal cord compression or intramedullary hemorrhage.

Since spinal cord compression may be treatable with prompt neurosurgical intervention, the first step in the evaluation of this patient is an emergent thoracic spine MRI (figure 1).

Although the MRI was technically limited due to motion degradation, it revealed central cord T2 hyperintensity from T4 to T8 and suggested prominent flow voids within and on the surface of the cord. There was no significant cord expansion or evidence of an extrinsic compressive process. In addition, there was no intraparenchymal pathologic enhancement with gadolinium contrast.

Laboratory evaluations revealed an elevated white blood cell count of 16.8 cells/dL with 89% neutrophils. Erythrocyte sedimentation rate was 19 mm/hour. The remainder of her blood cell counts, chemistries, and coagulation panels were within normal limits.

Question for consideration:

1. How do the MRI findings change the differential diagnosis and guide the diagnostic evaluation?

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SECTION 4

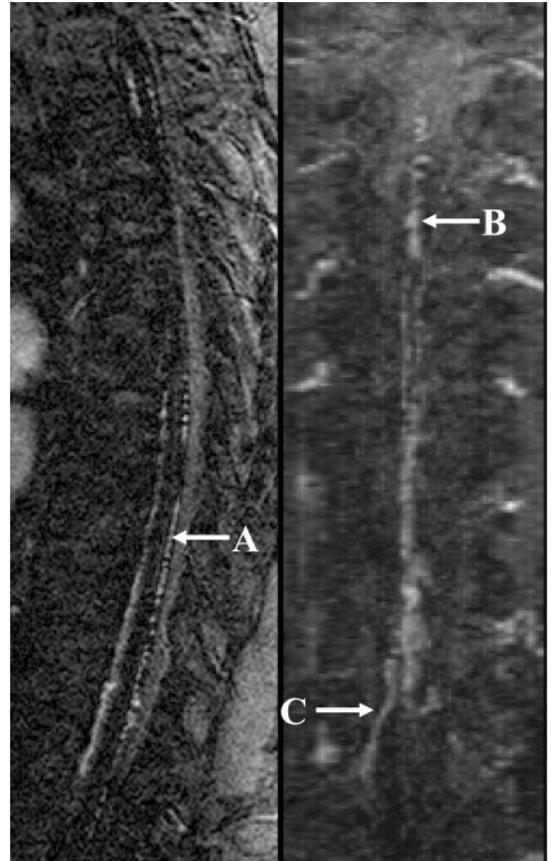
Central cord T2 hyperintensity is a nonspecific finding of intrinsic spinal cord damage that may be secondary to ischemia, inflammation, edema, hemorrhage, or traumatic injury. The absence of gadolinium enhancement implies integrity of the blood–CNS barrier and makes an acute inflammatory lesion less likely.

Diffusion-weighted imaging of the spine did not demonstrate an area of restricted diffusion. However, the sensitivity and specificity of diffusion-weighted imaging in detecting spinal cord infarction are unclear. The results have a relatively poor signal-to-noise ratio, with vulnerability to artifact from movement, breathing, and CSF flow.¹

The prominent flow voids over the surface of the spinal cord raise the possibility of a vascular malformation, and require additional evaluation. Dynamic gadolinium-enhanced MR angiography may localize an arteriovenous shunt when conventional MRI fails to do so.² These images can be a useful adjunct in guiding the selective intercostal artery angiography needed to confirm the diagnosis of a spinal vascular malformation but often may not adequately differentiate between feeding arteries and draining veins. Evaluation by catheter angiography, which precisely identifies the arterial supply and venous drainage of a malformation and also allows assessment of potential treatment options, is therefore necessary in cases where a spinal arteriovenous shunt lesion is suspected.

Gadolinium-enhanced spinal MR angiogram confirmed the presence of prominent intradural, extramedullary veins, and suggested an abnormal arterial-venous connection within the right T11/12 intervertebral foramen, most consistent with a spinal-dural arteriovenous fistula (figure 2). MR

Figure 2 Gadolinium-enhanced MR spinal angiography



Gadolinium-enhanced MR spinal angiography demonstrated dilated extramedullary vessels (A) and venous congestion (B), and also suggested an abnormal arterial-venous connection within the right T11/12 neural foramen (C).

angiography of the abdominal and thoracic aorta did not reveal a dissection.

Questions for consideration:

1. What would you suggest next for this patient?
2. What is the diagnosis and prognosis?

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SECTION 5

Selective angiograms of the right and left segmental arteries from T3 through L3 were obtained. The left L2 arteriogram demonstrated an intramedullary AVM at the T12-L1 level with arterial feeders arising from the hairpin loop of the radiculomedullary artery of Adamkiewicz (figure 3; see video). The shared origin of the arterial feeders and the anterior spinal artery from the artery of Adamkiewicz was confirmed on multiple oblique views. In addition, multiple dilated venous channels were seen extending to the lower thoracic levels. Venous drainage was into the inferior vena cava at the right L1 level. No dural arteriovenous fistula was identified. The remainder of the segmental arteries appeared normal. No arterial or venous aneurysms were seen.

These findings are consistent with a spinal intramedullary AVM, type 2 (glomus).

Treatment options were believed to be limited. The patient was not treated with arterial embolization or surgical intervention because the risk for either procedure was considered too significant. Embolization was not performed because the caliber of the arterial feeder was too small for direct selective embolization of the feeding vessels. Because the L2 radicular artery was shown to feed the anterior spinal artery via the radiculomedullary artery of Adamkiewicz, emboliza-

tion of this branch would produce further cord infarction. Surgery was not performed because the lesion was inaccessible in its intramedullary location.

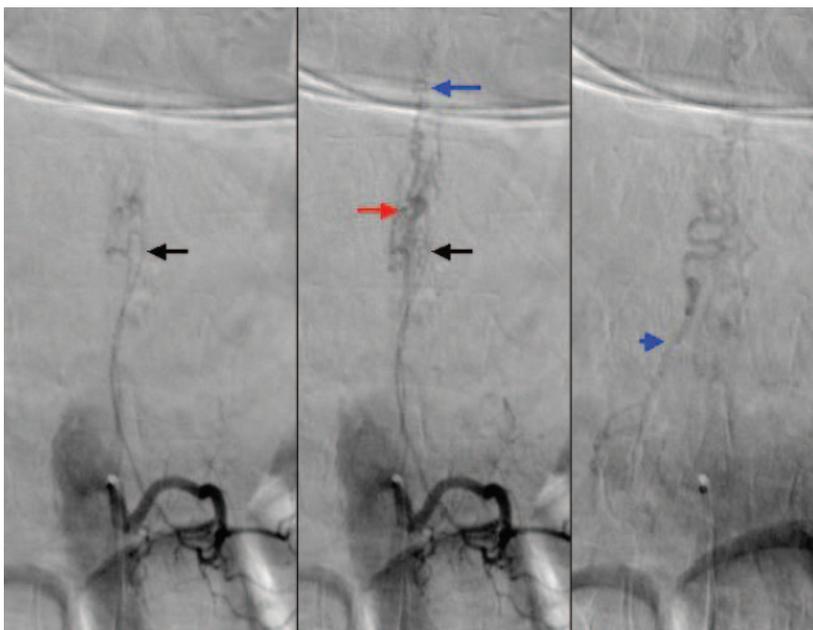
The patient remained stable, and after 1 week she was discharged for further rehabilitation. At 2-month follow-up, she had made minimal recovery in her leg strength.

DISCUSSION The blood supply to the spinal cord is provided by the single anterior and dual posterior spinal arteries (figure 4). From the anterior and posterior spinal arteries arise small sulcal and penetrating intramedullary arteries. The caliber of the anterior spinal artery narrows in the thoracic cord, resulting in greatly diminished descending blood flow.³ Blood flow to lower portions of the spinal cord arises from radiculomedullary arteries that reconstitute the anterior spinal artery and radiculopial arteries that reconstitute the posterior spinal arteries.³ Radiculomedullary and radiculopial arteries originate from radicular arteries. Thirty-one pairs of radicular arteries pass through the intervertebral foramina to supply each spinal nerve and the dura. They originate from large segmental arteries, which include the ascending cervical, deep cervical, vertebral, intercostal, lumbar, and sacral arteries. Only 6 to 10 radicular arteries give rise to radiculomedullary branches, but the exact number and anatomic location is quite variable.⁴ Of the radiculomedullary arteries supplying the lumbar cord, the largest is named the artery of Adamkiewicz.⁴

The venous anatomy of the spinal cord includes intramedullary veins that collect into the anterior and posterior superficial veins, which drain into radicular veins. Anterior and posterior radicular veins drain into the epidural (or internal vertebral) venous plexus. The venous plexus drains into thoracic, abdominal, and intercostals veins.⁴ Of note, the venous drainage of the spinal cord does not contain valves, and under pathophysiologic conditions flow may become retrograde.

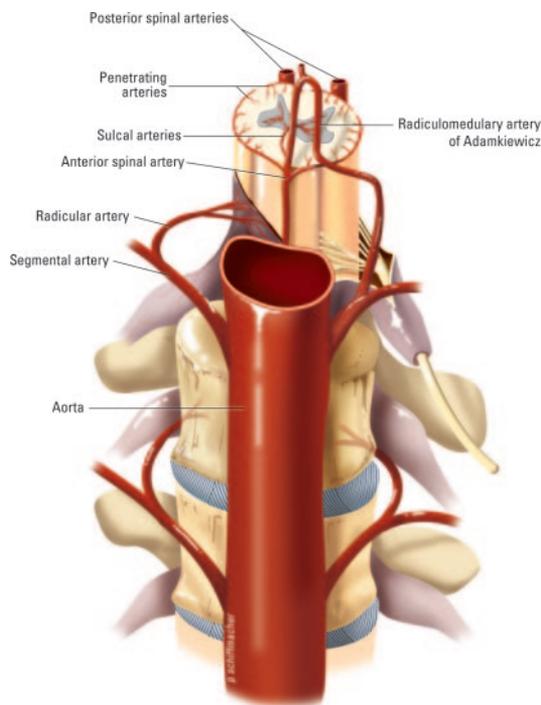
There are several types of spinal cord vascular malformations, each defined by its anatomic characteristics (table). These include the dural arteriovenous fistula (type 1 spinal dural AV fistula), intramedullary arteriovenous malformations (glomus or type 2 spinal cord AVM, and juvenile or type 3 spinal cord AVM), and direct perimedullary fistulas (type 4 spinal cord AV fistula).⁵ Other spinal cord vascular malformations, the discussion of which is beyond the scope of this report, include cavernomas, telangiectasias, venous angiomas, epi-

Figure 3 Selective microcatheter left L2 arteriogram demonstrates an intramedullary arteriovenous malformation (AVM) (red arrow) at the T12-L1 level with arterial feeders arising from the artery of Adamkiewicz (black arrow)



Multiple dilated venous channels extend up to the lower thoracic level (blue arrow) and drain the AVM (blue arrowhead) into the inferior vena cava. See video.

Figure 4 Arterial supply of the spinal cord



Note that the blood supply to the anterior spinal artery is reconstituted by the artery of Adamkiewicz. Courtesy Paul Schiffmacher.

dural AVMs, paravertebral vascular malformations, vertebral hemangiomas, and complex syndromic vascular malformations, including Cobb

metameric angiomatosis and Osler-Weber-Rendu disseminated angiodysplasia.⁵

Glomus (type 2) spinal AVMs are intramedullary lesions, which contain intervening cord parenchyma within the abnormal tangled vessels.⁵ In the glomus AVM, the vascular nidus is compact.⁵ The arterial supply is through an enlarged radiculomedullary artery that also supplies the cord via anterior or posterior spinal arteries. There is high flow through the lesion, as demonstrated by angiographic transit times.⁶ Drainage occurs by shunted anterograde flow through engorged medullary spinal veins and the epidural venous plexus. These lesions can occur anywhere in the cord.⁶ They typically affect younger patients, and are thought to result from defective vascular embryogenesis.

The pathophysiology of acute decompensation in intramedullary AVMs is likely related to intraparenchymal or subarachnoid hemorrhage, venous hypertension, or local ischemia. High-pressure turbulent blood flow predisposes to local arterial and venous aneurysms, which are vulnerable to rupture and cause hemorrhage.⁷ Alternatively, if there is sufficient shunting of blood through the high-flow AVM away from the cord parenchyma, cord ischemia may result.⁶ More recently, venous congestion has been emphasized as an additional inciting factor in the pathophysiology of some spinal intramedullary AVMs.⁸ In-

Table	Characteristics of the different types of pathologic spinal arteriovenous connections		
AVM type	Type 1: Dural arteriovenous fistula	Types 2 and 3: Intramedullary spinal AVMs	Type 4: Perimedullary arteriovenous fistula
Feeding vessels	Dural branch of radicular artery	Radiculomedullary artery	Single or multiple radiculomedullary arteries
Arteriovenous shunt	Within dural sleeve	Type 2: Compact intramedullary shunt Type 3: Diffuse intramedullary tangle	Direct connection on surface of cord, no vascular nidus
Draining vessels	Retrograde flow through medullary vein, causing engorgement of valveless intraparenchymal radial veins and epidural venous plexus	Anterograde flow through engorged medullary spinal veins and epidural plexus	Single or multiple medullary veins
Flow dynamics	Slow flow	Fast, turbulent flow	High flow
Location	Lower thoracic, lumbar spine	No predilection	No predilection
Etiology	Acquired, possibly due to thrombosed epidural plexus	Congenital	Congenital
Typical population	Elderly men	Young adults	Young adults
Treatment	Embolization or surgical disconnection	Type 2: Occasionally, embolization or surgery	Surgical treatment of smaller lesions and embolization of larger lesions
		Type 3: Rarely treatable	

AVM = arteriovenous malformation.

deed, our patient had evidence of extensive MRI cord signal abnormality and diffuse angiographic venous engorgement suggesting chronic venous hypertension. Although these potential pathophysiologic mechanisms have each received support in the literature, definitive evidence for any particular mechanism is lacking. In most cases there is likely a combination of underlying pathophysiologic mechanisms.

Type 2 intramedullary AVMs can occasionally be treated with surgical disconnection or resection, but the risk of poor outcome may be substantial, depending on the location and extent of the lesion.^{6,9} In some cases, embolization of an intramedullary AVM can be successful, depending on the anatomic characteristics of the lesion and the results of test occlusion of the parent vessel.¹⁰

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