

## Spinal Cord Syndromes

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### Abstract

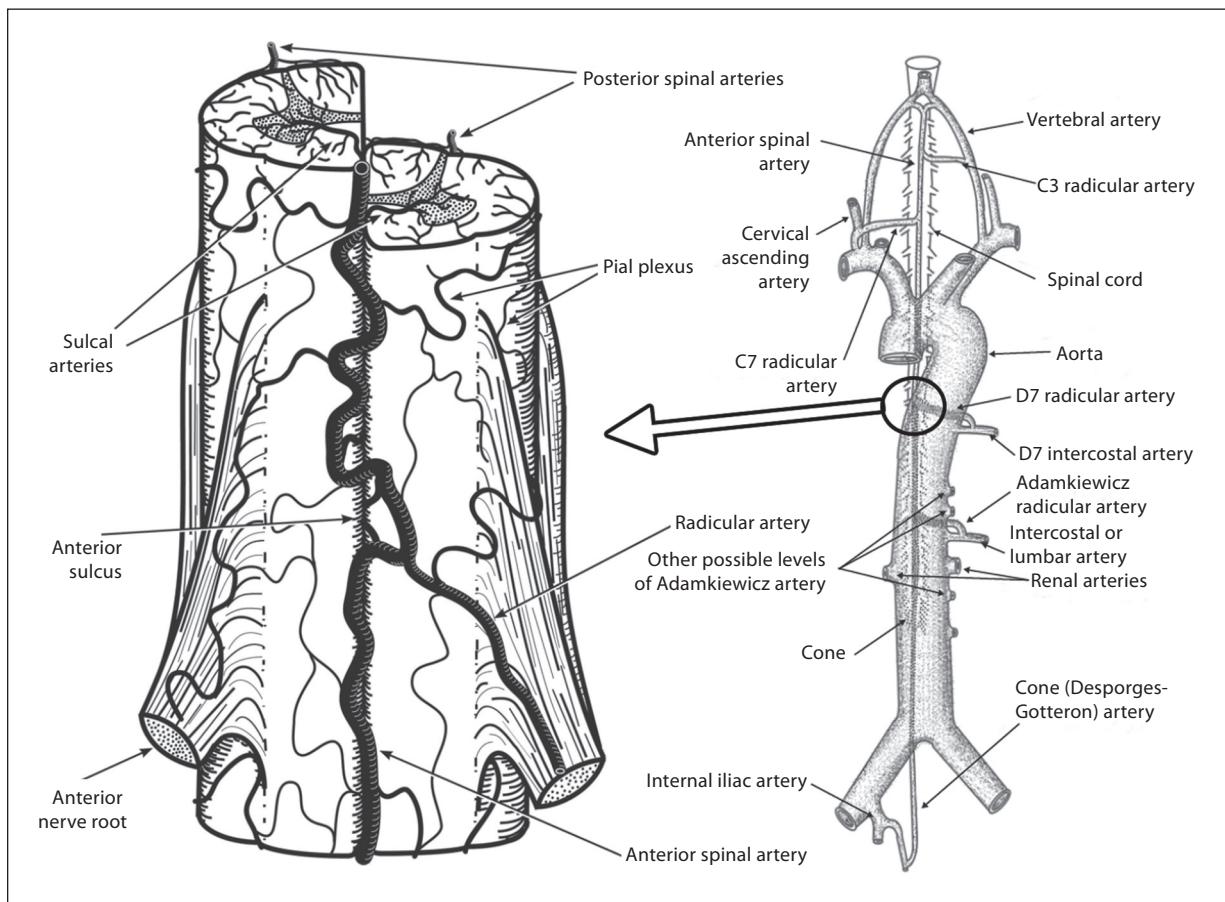
Spinal cord infarction is much rarer than cerebral stroke, but its early recognition is important as it may signify serious aortic conditions. The most frequent type is anterior spinal artery syndrome, presenting with bilateral weakness (usually paraparesis), impairment of spinothalamic sensation and preservation of deep sensation. Depending on its level, it may present with respiratory dysfunction. More rarely, posterior infarcts sparing spinothalamic sensation but involving lemniscal sensation may be encountered. Unilateral, central or transverse infarction may also be seen probably on account of different mechanisms. Other rarer forms of spinal ischemia also include spinal TIAs, venous infarction, fibrocartilaginous embolism and decompression sickness.

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Spinal cord ischemia is much rarer than brain ischemia, it was found to represent only 1% of all strokes in an autopsy series [1]. Aortic diseases are the most frequent cause [2–4]. Diagnosis can be challenging as its clinical manifestations can mimic other myelopathies (mostly inflammatory or infectious) and there is currently no test that is specific and sensitive enough to ascertain the diagnosis. Spinal MR imaging can indeed often detect an acute lesion (67–85% of the cases [3–5]), but diffusion weighted sequences are still too susceptible to artifacts to reliably confirm whether it is ischemic. The spinal vasculature is also impossible to investigate without

using invasive angiographic techniques. However, prompt recognition of typical clinical pictures of spinal cord infarction can be lifesaving as they may point a life-threatening aortic disease (dissection, thrombosis or rupture of an aneurysm) that is potentially treatable if detected early.

Spinal cord infarction can be restricted in an arterial territory or be more widespread according to its pathogenesis (single artery occlusion versus regional or global hypoperfusion). Spinal cord vascular anatomy is important to understand the semiology of spinal cord ischemia. The spinal cord is supplied by 3 spinal arteries running discontinuously along the spinal cord (fig. 1, left). These arteries (one anterior and two posterior) are supplied by several radicular arteries (often arising from direct aortic branches) accompanying nerve roots mostly in cervical and low thoracolumbar regions (fig. 1, right). The spinal arteries are interconnected by a thin but widespread plexus at the surface of the spinal cord. The anterior spinal artery gives unilateral alternating branches (sulcal arteries). The anterior spinal artery may be duplicated in some regions which could explain the occurrence of unilateral anterior infarction. The anterior spinal artery supplies the anterior two-thirds of the spinal cord, the remaining posterior third being supplied by the posterior arteries. We will review here the manifestations of spinal cord infarction and other rarer spinal ischemic conditions.



**Fig. 1.** Anatomy of spinal cord vascularization, aortic branches on the right and spinal cord arteries on the left. Reproduced and modified with permission of *Archives of Neurology* [3].

### Spinal Cord Infarction

The different clinical syndromes are summarized in table 1. Onset of symptoms is acute, usually over a few minutes, but it may be progressive over a few hours. It is often (59–82%) [3, 4] marked by a pain in the back that is usually localized at the level of the actual cord lesion. The pain has also frequently (~80% [3, 5]) a radicular irradiation. Ischemia of local meninges, vertebral body or spinal root has been hypothesized to explain this initial pain. Patients often reported that the symptoms (pain), started during a back movement or a Valsalva maneuver [3].

Anterior spinal artery infarction is by far the commonest (also referred to as anterior spinal artery syndrome). It was initially described by Preobrashenski [6] in the setting of meningovascular syphilis. It is characterized by bilateral motor deficits (mostly paraparesis) and loss of thermalgic sensation with sparing of proprioceptive and vibratory modalities. This dissociated sensory loss is explained by the selective involvement of spinothalamic tracts which spares the posterior columns. On testing, a small suspended sensory loss can be found above the main one. Motor deficits start usually with flaccid tone and will progressively

**Table 1.** Summary of the clinical features of spinal cord infarction. Reproduced with permission of *Archives of Neurology* [3]

Stroke syndrome	Feature
Anterior spinal artery infarct	Bilateral motor deficit with spinothalamic sensory deficit
Anterior unilateral infarct	Hemiparesis with contralateral spinothalamic sensory deficit
Posterior unilateral infarct	Hemiparesis with homolateral lemniscal sensory deficit
Central infarct	Bilateral spinothalamic sensory deficit without motor deficit
Posterior spinal artery infarct	Bilateral motor deficit with lemniscal sensory deficit
Transverse infarct	Bilateral motor deficit with complete sensory deficit

evolve into spasticity with the appearance of corticospinal signs. With cervical cord infarction, the motor deficit can present at times with bibrachial paresis sparing the lower limbs [3]. A focal amyotrophy may develop in the myotomes at the level of the infarct due to the involvement of the anterior horns (for example, hand intrinsic muscles in low cervical infarcts). Sphincter dysfunction is common, usually with initial urinary retention evolving into a spasmodic bladder. Patients often require bladder catheterization acutely and urinary infection is a frequent early complication. Rectal tone is usually diminished acutely, but involuntary defecation can be seen later. In high cervical infarcts, respiratory failure is common and may require intubation and ventilation. Respiratory failure can be dissociated affecting only involuntary breathing with spared voluntary function (Ondine curse) [7]. The respiratory failure should not be considered as irreversible as, in our experience, even unilateral incomplete recovery can be sufficient to allow the patient to breathe spontaneously. Long term outcome mostly depends on the severity of the initial motor deficits. The initial back pain usually subsides over a few days, but the patients are at risk of developing delayed neuropathic pain [8] (with a burning character), which can be extremely difficult to treat.

Posterior arteries infarct is much rarer. It shows marked bilateral motor involvement with sparing of the spinothalamic sensation but impairment of vibratory and proprioceptive sense. When motor

deficits are moderate, incoordination through involvement of spinocerebellar tracts can be seen. Lhermitte's sign is often present as a marker of the involvement of the posterior columns. In subtle cases, testing of the discrimination of the direction of touch (the patient being asked in which direction a touching stimulus is displaced on his skin) can be a sensitive test. As in anterior spinal infarcts, sphincter dysfunction is almost invariably present.

Unilateral infarcts, whether anterior or posterior, are usually less severe and present as an incomplete Brown-Séquard syndrome sparing either lemniscal or spinothalamic sensation. Unilateral infarcts have a better recovery prognosis.

Less frequently, spinal cord infarction can occur in the central region (pencil-shaped), this region may represent a watershed area. Central infarcts present with syringomyelia-like sensory symptoms, as suspended loss of thermo-algic sensation (involvement of crossing spinothalamic fibers), and also some corticospinal signs but without prominent motor deficits. Transverse infarction may be found usually after prolonged hypoperfusion.

### Other Ischemic Spinal Cord Syndromes

Transient spinal cord ischemic episodes (spinal TIAs) are rare, but they can herald the definitive infarct [2, 5]. They usually present with a fluctuating paraparesis over a few minutes to one hour, sometimes in clusters. Exceptionally, these episodes can take the form of spinal cord claudication

as originally described by Déjerine [9]. During those events that are triggered by exercise, the patient experiences lower limb weakness with corticospinal signs which both disappear with rest.

Spinal cord venous thrombosis is extremely rare. Recent cases are described after interventional procedures (mostly esophageal varices ligation or sclerotherapy [10]). It presents with a progressive painful myelopathy not restricted to a particular vascular territory. Some authors wondered if absence of valves in the epidural spinal venous plexus might favor reflux of thrombi in the spinal venous circulation.

Fibrocartilaginous embolism is a rare condition presenting with a progressive painful myelopathy often following unusually intense exercise or minor trauma. Deficits appear after a delay following the onset of the pain. In the majority of the cases reported, the outcome was very poor with death within a few days. This is however potentially a bias as this diagnosis can only be ascertained with pathology [11]. In these cases, nucleus

pulposus fragments are found in spinal vessels. The mechanism of this embolism is unclear.

Decompression sickness often presents with a myelopathy whose mechanism probably includes the formation of gas bubbles in spinal cord tissue and vessels (mostly veins). This usually happens in diving when the decompression procedure is not followed, but also in aviation. Symptoms usually start during decompression or shortly after with diffuse pain, ascending sensory deficits with motor and sphincter deficits. A clear sensory level may not be found because of the diffuse nature of the condition. Recompression therapy greatly improves the prognosis with up to 80% of full recovery [12]. Patients may sometimes be left with focal lower motor neuron deficits.

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### References

- 1 Sandson TA, Friedman JH: Spinal cord infarction. Report of 8 cases and review of the literature. *Medicine (Baltimore)* 1989;68:282–292.
- 2 Cheshire WP, Santos CC, Massey EW, Howard JF Jr: Spinal cord infarction: etiology and outcome. *Neurology* 1996; 47:321–330.
- 3 Novy J, Carruzzo JF, Maeder P, Bogouslavsky J: Spinal cord ischemia: clinical and imaging patterns, pathogenesis, and outcomes in 27 patients. *Arch Neurol* 2006;63:1113–1120.
- 4 Masson C, Pruvo JP, Meder JF, Cordonnier C, Touze E, De La Sayette V, Giroud M, Mas JL, Leys D: Spinal cord infarction: clinical and magnetic resonance imaging findings and short-term outcome. *J Neurol Neurosurg Psychiatry* 2004;75:1431–1435.
- 5 Kumral E, Polat F, Gulluoglu H, Uzunkopruc C, Tuncel R, Alpaydin S: Spinal ischaemic stroke: clinical and radiological findings and short-term outcome. *Eur J Neurol* 2011;18:232–239.
- 6 Preobrashenski PA: Syphilitic paraplegia with dissociated disturbance of sensation. *J Nevropat i Psikhiat* 1904;4: 394–433.
- 7 Manconi M, Mondini S, Fabiani A, Rossi P, Ambrosetto P, Cirignotta F: Anterior spinal artery syndrome complicated by the ondine curse. *Arch Neurol* 2003;60: 1787–1790.
- 8 Triggs WJ, Beric A: Sensory abnormalities and dysaesthesias in the anterior spinal artery syndrome. *Brain* 1992;115: 189–198.
- 9 Déjerine J: Intermittent spinal cord claudication (in French). *Presse Med* 1911;19:981–984.
- 10 Debette S, Gauvrit JY, Desseaux G, Nibbio A, Henon H, Pruvo JP, Leys D: Paraplegia after ligation of esophageal varices. *Neurology* 2003;60:879–880.
- 11 Mateen FJ, Monrad PA, Leep Hunderfund AN, Robertson CE, Sorenson EJ: Clinically suspected fibrocartilaginous embolism: clinical characteristics, treatments, and outcomes. *Eur J Neurol* 2011; 18:218–225.
- 12 Blatteau JE, Gempp E, Simon O, Coulange M, Delafosse B, Souday V, Cocharde G, Arvieux, Henckes A, Lafere P, Germonpre P, Lapoussiere, Hugon M, Constantin P, Barthelemy A: Prognostic factors of spinal cord decompression sickness in recreational diving: retrospective and multicentric analysis of 279 cases. *Neurocrit Care* 2011;15:120–127.

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