Vascular disorders of the spinal cord
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Introduction

This article includes discussion of vascular disorders of the spinal cord, spinal cord arteriovenous malformations, spinal cord hemorrhagic syndromes, spinal cord infarction, and transient cord ischemia. The foregoing terms may include synonyms, similar disorders, variations in usage, and abbreviations.

Overview

The author provides updated information regarding newly described etiologies associated with spinal cord ischemia.

Historical note and terminology

Jackson reported the first nontraumatic spinal epidural hematoma in 1869. Adamkiewicz first described the blood supply of the spinal cord in 1882. In the same year, Bastian theorized that occlusion of spinal vessels was responsible for softening of the spinal cord. Singer in 1902 reported 2 cases of vascular myelopathy with pathological changes. Dejerine first described intermittent claudication of the spinal cord in 1906. In 1909, Spiller reported the first case of anterior spinal artery syndrome. The etiology in this particular case was syphilitic vasculitis. In 1912, Marie and Foix described the syndrome of “trophomalacie anterieure,” ischemic lesions in the anterior horns due to small-vessel disease. In 1926, Foix and Alajouanine described a clinicopathological correlation between subacute myelopathy and spinal arteriovenous malformations in 2 cases. In 1968, Newton and Adams presented the first reports of angiographic embolization of spinal vascular malformations (Silver and Buxton 1974; Fieschi et al 1985; Mattle et al 1987; Muraszco and Oldfield 1990; Sliwa and Maclean 1992; Hodes et al 1994).

Clinical manifestations

Presentation and course

The clinical manifestations will be described independently for the different vascular syndromes of the spinal cord, which are grouped into spinal cord infarction or transient ischemia syndromes, spinal cord hemorrhagic syndromes, and spinal vascular malformations.

Spinal cord infarction or transient ischemia syndromes. The clinical presentation varies depending on the vascular territory involved. The following vascular syndromes have been identified: anterior spinal artery syndrome, posterior spinal arteries syndrome, transverse infarction of the spinal cord, central cord infarction, venous infarction, transient spinal ischemia, and lacunar cord infarction.

Anterior spinal artery syndrome. This syndrome is classically heralded by pain. Pain can be diffuse, radicular, or girdle-like in distribution and is described as lancinating or dull. The symptoms usually occur suddenly and may progress over minutes to hours. The pain in upper cervical cord infarctions can mimic a cluster headache attack with a Horner syndrome (de la Sayette et al 1999). Traditionally, patients present with an acute myelopathy involving the anterior two thirds of the cord, with paralysis, loss of bowel and bladder control, and loss of temperature and pain sensation below the lesion. Vibratory and position senses are intact due to preservation of the posterior columns (Sandson and Friedman 1989; Toole 1990; Sliwa and Maclean 1992; Bogousslavsky and Caplan 1995).

Partial forms of the anterior spinal artery syndrome are common. Infarction can be limited to the gray matter of the anterior horn because of its greater susceptibility to ischemia. Patients show acute flaccid motor paralysis and normal sphincter and sensory functions (Bogousslavsky and Caplan 1995). Monoplegia is rare, but has been described (Nelson and Ho 2016).

Sometimes anterior spinal artery infarction presents as a partial Brown-Sequard syndrome (incomplete hemicord syndrome due to the preservation of the posterior columns). Vascular explanations for this phenomenon include duplication of the anterior spinal artery as well as unilateral sulcomissural artery involvement (Bogousslavsky and Caplan 1995).
Anterior spinal artery infarction at the caudal end of the brainstem can occur. In such cases, the lesion tends to be unilateral, causing a hemiparesis with contralateral hemianesthesia for all modalities. Conus medullaris infarction causes a cauda equina syndrome, with sphincter paralysis, perianal and perineal sensory loss, and variable motor and sensory deficits in lower extremities (Sliwa and Maclean 1992; Bogousslavsky and Caplan 1995). Man-in-the-barrel syndrome has also been reported following anterior cervical cord infarction (Berg et al 1998).

Posterior spinal arteries syndrome. These lesions are rare and often difficult to recognize, probably due to the extensive collateral circulation in that region. Patients usually have pain along the spine and paresthesias or dysesthesias in the legs. Vibration and position sense are absent below the level of the lesion. Segmental anesthesia for all modalities is present. Usually the infarcted area extends anteriorly, with a variable degree of paralysis and sphincter dysfunction. Whenever this happens, the weakness is not as severe as in the anterior spinal artery cases (Sandson and Friedman 1989; Toole 1990; Sliwa and Maclean 1992; Bogousslavsky and Caplan 1995).

Transverse infarction of the spinal cord. The syndrome is characterized clinically by a cord section, with flaccid paralysis and total loss of sensation below the lesion, as well as sphincter dysfunction. The motor paralysis is often preceded by acute pain of spinal or radicular character, corresponding to the level of cord infarction. Pain usually subsides rapidly. Tendon and abdominal reflexes are lost. Autonomic disturbances may occur (Bogousslavsky and Caplan 1995).

Central cord infarction. Ischemia in the arterial border zone between anterior spinal artery and posterior spinal arteries may result in this type of clinical infarct. The picture, however, is believed to be clinically indistinguishable from the anterior spinal artery syndrome (Sliwa and Maclean 1992; Bogousslavsky and Caplan 1995).

Venous infarction. The clinical presentations of this type of infarction vary, although certain similarity to the arterial infarcts is found. Presentation is often of a subacute transverse myelopathy, with associated pain in the back, abdomen, or legs. There is some degree of motor and sensory loss, but without a well-defined pattern. The level of sensory loss may not be as easily delineated as in arterial infarcts (Sliwa and Maclean 1992; Bogousslavsky and Caplan 1995).

Transient spinal ischemia. Transient ischemia of the cervical cord may cause "drop attacks." The clinical picture in the lumbar region ("spinal cord claudication") may be of weakness of the lower extremities precipitated by effort and relieved by rest. The pedal pulses should remain palpable. Episodes are typically painless. When the lesion is in the abdominal aorta, subsequent ischemia of the gluteal muscles can induce pain in the hips (Leriche syndrome) (Toole 1990).

Lacunar cord infarction. Lacunar infarcts sometime present as subacute progressive involvement of the anterior horns. Lacunar cord infarction is also referred to as "vascular myelopathy in old age." The usual findings are wasting and weakness of the small muscles of the hand and scattered pyramidal signs, mimicking motor neuron disease (Fieschi et al 1985).

Intramedullary hemorrhage (hematomyelia). Nontraumatic hematomyelia can occur spontaneously, secondary to a vasculitis (Yeo et al 2013), or follow the Valsalva maneuver. The most common presentation is excruciating pain in the back of sudden onset, often with radicular radiation. The pain is followed by immediate loss of muscle tone below the lesion and sphincter dysfunction. As the blood accumulates, it displaces and compresses the ascending and descending tracts. Usually the more laterally placed portions of the spinothalamic tracts are preserved. In these cases sensation is preserved in the sacral dermatomes, although the sensation in the lumbar and thoracic regions may be lost ("sacral-sparing anesthesia") (Toole 1990).

Spinal epidural or subdural hematomas. Epidural hematomas are approximately 4 times more common than subdural hematomas in the spinal region. They can be preceded by a lumbar puncture when it is performed in a patient who is taking anticoagulants. Sometimes they occur after straining or Valsalva maneuvers. In patients with coagulation defects, a hematoma can appear spontaneously. The usual presentation is intense local pain in the neck or back, with some radicular radiation. The pain is followed, usually hours later, by a variable degree of weakness and sensory and sphincter dysfunction, usually symmetric (Mattle et al 1987; Bogousslavsky and Caplan 1995).

Spinal subarachnoid hemorrhage. Clinically, spinal subarachnoid hemorrhage is usually manifested by excruciating pain in the back, with radicular irradiation. Opisthotonos followed by variable weakness of the extremities can occur.
Meningeal signs may be present (Toole 1990).

**Dural fistulas.** Patients with dural fistulas usually have sensory and motor deficits that progress gradually over months or years. The legs are affected more frequently than the arms. Patients often complain of symptom exacerbation with postural or physical activities (Muraszco and Oldfield 1990; McCormick 1993; Bogousslavsky and Caplan 1995).

**Intradural fistulas.** Persons with intradural fistulas frequently have an acute hemorrhagic presentation. Transient ischemic cord dysfunction has been described. The arms are usually affected (cervicothoracic location). Bruits can sometimes be heard over the spine at the level of the shunting (Muraszco and Oldfield 1990; McCormick 1993; Bogousslavsky and Caplan 1995).

**Cavernous malformations.** These lesions present as an acute hemorrhage or as a subacute progressive or recurrent myelopathy (McCormick 1993).

**Prognosis and complications**

The prognosis of spinal cord infarction has been addressed in small series. Motor function shows progressive recovery in most of the patients, with the deficit being maximum at the onset of the stroke. The degree of recovery is variable. Complete recovery is rare and some degree of disability is common (de la Barrera et al 2001). A series of 27 patients showed that only 13 (48%) had significant gait impairment at discharge (Novy et al 2006). Adequate specialized spinal cord care may improve prognosis, particularly in patients with pressure sores and urinary complications (Young and Woolsey 1995). Rehabilitation has shown to improve functional outcomes (New and McFarlane 2012). The functional prognosis of spinal cord infarctions tends to be worse than cerebral infarctions (Naess and Romi 2011), but gradual improvement is not uncommon (Robertson et al 2012).

Pain is a disabling long-term complication in many patients (90%), independent of motor function recovery. The frequency and intensity of pain after spinal cord infarction seem higher than with other myelopathies and may have an impact on the quality of life (Young and Woolsey 1995).

Complications of spinal cord infarctions are similar to other forms of myelopathy and include: urinary tract infections or sepsis; pressure sores; autonomic dysreflexia (eg, episodes of uncontrolled hypertension); urinary (Sakakibara et al 2008), bowel, and sexual dysfunction; renal dysfunction; chronic pain; deep venous thrombosis or pulmonary embolism; respiratory dysfunction; and psychological or psychiatric manifestations (Young and Woolsey 1995).

The prognosis of spinal epidural and subdural hematomas depends mainly on the preoperative neurologic status. Incomplete lesions have a better prognosis than a complete sensorimotor lesion. The rate of functionally relevant recovery in the latter is probably less than 50%. The rapidity of onset of symptoms is associated with a poor prognosis, probably because a rapid course is secondary to massive bleeding. Delays in performing laminectomy also are associated with poor prognoses (Mattle et al 1987). Lumbar hematomas have better prognosis than cervical or thoracic hematomas (Mattle et al 1987).

Complications of epidural or subdural hematomas include rebleeding, surgical side effects of laminectomy, and the same complications of an acute myelitis as stated above.

The prognosis of dural arteriovenous malformations depends on the type. The most common, the dural fistula, has a typical progressive course; 20% to 50% of patients are severely disabled 6 months after the onset of symptoms. The natural history of intradural arteriovenous malformations is uncertain, not only because they are less common, but also because of their tendency to present as an acute hemorrhage (McCormick and Stein 1990; McCormick 1993).

The prognosis after surgical treatment of a dural malformation is directly related to the previous neurologic status. The better the preoperative neurologic exam, the better the outcome.

The prognosis of surgical treatment for intradural arteriovenous malformations depends on surgical accessibility and previous neurologic status. In general, 80% to 90% of patients have good or excellent results when total excision can be achieved. Results are usually not as good as in the dural type. The most favorable outcome is achieved with cervical lesions, lumbar being next. When embolization treatment is done, recanalization of the nidus is a common complication (Muraszco and Oldfield 1990; McCormick 1993).
**Clinical vignette**

A 57-year-old man with a history of smoking, coronary atherosclerotic disease, chronic obstructive pulmonary disease, and renal stones underwent emergency Dacron graft replacement of a dissecting aortic aneurysm between the renal and distal common iliac arteries. After the surgery the patient was noted to have flaccid paraplegia with absent reflexes and a pin-prick sensory level below the navel. His vibration sense was intact at the knees and ankles. MRI of the thoracic cord demonstrated an extensive cord infarction.

**Biological basis**

**Etiology and pathogenesis**

Spinal cord infarcts are the result of a number of arterial diseases including atherosclerosis, dissection of the aorta, embolism, or vasculitis. Vascular malformations can produce mass effect, ischemia, or hemorrhage. Spinal cord hemorrhages may result in secondary cord ischemia or mass effect.

**Segmental vessels.** There are approximately 31 pairs of segmental vessels. They arise from different sources (in craniocaudal order): vertebral arteries, ascending cervical arteries, deep cervical arteries, superior intercostal artery, aorta, ilioiliac artery, and lateral sacral arteries (Sliwa and Maclean 1992).

Segmental arteries divide into anterior and posterior rami. The posterior rami enter the intervertebral foramen and constitute either the anterior or posterior (or both) radicular arteries. Radicular arteries are also known as medullary or radiculomedullary arteries (Sliwa and Maclean 1992).

Anterior radicular (medullary) arteries are bigger than their posterior counterparts. They supply the anterior spinal artery, which acts as an anastomotic channel between them. Their number varies from 2 to 17 (average 10). They are distributed unevenly throughout the cord. The cervical-upper thoracic region is highly vascularized, whereas the midthoracic area (T4 to T8) is poorly perfused because it is supplied by a single small radicular artery known as the “arteria radicularis magna” or artery of Adamkiewicz (usually T5 to T7 level, but variable) (Sliwa and Maclean 1992).

Posterior radicular (medullary) arteries are smaller in size. They supply the posterior spinal arteries, which act as anastomotic channels between them. Their number is between 10 and 23. They tend to be of smaller diameter in the lumbosacral region (Sliwa and Maclean 1992).

**Longitudinal vessels.** There are 3 longitudinal vessels: 1 anterior spinal artery and 2 posterior spinal arteries. The anterior spinal artery is an anastomotic channel that runs anteriorly in the midline. The anterior spinal artery constitutes the caudal continuation of the 2 vertebral arteries, at the level of the foramen magnum. It extends throughout the length of the cord in the anterior median fissure. The posterior spinal arteries run in the posterior lateral aspect of the cord. They usually originate from the vertebral arteries but occasionally come from the posterior inferior cerebellar arteries (Toole 1990; Sliwa and Maclean 1992).

**Vasa coronae**. These surface vessels originate from the anterior spinal artery and posterior spinal arteries, and form a vascular ring around the cord (Sliwa and Maclean 1992).

**Penetrating vessels.** They originate from the anterior spinal artery, posterior spinal arteries, and vasa coronae and perfuse the central portions of the cord. They mostly supply the peripheral anterior white matter of the cord (Sliwa and Maclean 1992).

**Central (sulcal) arteries.** They are branches of the anterior spinal artery. They enter the cord through the anterior median fissure and give longitudinal branches. There are more than 200 sulcal arteries, which are unevenly distributed throughout the cord (lumbar>cervical>thoracic). They mainly perfuse anterior horns, deep gray matter of posterior horn, and central white matter (Sliwa and Maclean 1992).

**Posterior median septum arteries.** These are branches of the posterior spinal arteries that supply the dorsal funiculi and the central gray matter. The rest of the posterior one third of the cord is perfused by other penetrating branches of posterior spinal arteries (Sliwa and Maclean 1992).

**Extrinsic veins.** There are 1 or 2 anterior spinal veins and usually a single posterior spinal vein. In addition,
posterolateral and anterolateral spinal veins form discontinuous channels. The medullary veins show little correlation with the arteries. There are some 6 to 25 anterior veins. There is a great anterior medullary vein, usually between T11 and L3. Posterior medullary veins average 20 in number.

Extrathecally, an internal venous plexus lies in the epidural space, and an external plexus drains the vertebral bodies (Sliwa and Maclean 1992).

**Intrinsic veins.** There are 2 anterior central veins (draining central white matter and medial anterior horn), 2 posterior central veins (median septum white matter and gray commissure), and an intrinsic venous drainage for the rest of the cord (Sliwa and Maclean 1992).

**Vascular physiology of the spinal cord.** There is no definite explanation as to why spinal cord infarcts are so rare compared with cerebral infarctions. Both the presence of rich anastomotic channels (vasa coronae) and the existence of extraspinal anastomoses are speculated to play a role. Differences in tissue vulnerability to ischemia have also been implicated. Although blood perfusion in the brain is in the range of 50 mL/min per 100 g, animal studies suggest a value of 10 to 20 mL/min per 100 g, depending on the cord segment. The gray matter is more highly perfused than the white (5-fold difference) and is also more vulnerable to ischemia (Satran 1988; Weinstein 1993).

Like the brain, the spinal cord vasculature shows the phenomenon of autoregulation, with a similar autoregulatory range of 60 to 120 mm Hg. It has been hypothesized that variations of the regulatory curve may occur in individual vessels or areas, accounting for the apparent regional susceptibility to ischemia in hypoperfusional states (Hickey et al 1986; Singh et al 1994).

**Spinal cord border zones.** Longitudinally speaking, the lower thoracic and lumbar cord (T6 to L3) is considered to be the area most vulnerable to ischemia. The last radicular feeding vessel, the artery of Adamkiewicz, supplies this region. This vessel makes an acute angle of entry in the cord, and is considered to be an end artery, with no functional back-up anastomosis (Singh et al 1994).

In a transverse section, the anterior spinal artery territory (anterior two thirds) is believed to be more susceptible to ischemia than the posterior one third of the cord. The explanation may be the existence of more efficient functional anastomoses at the level of the posterior spinal arteries region (Sliwa and Maclean 1992).

The gray matter is considered to be more susceptible to ischemia than the white matter, probably due to the differences in perfusion requirements between the 2 tissues (Dawson and Potts 1991).

**Etiology of spinal cord infarcts.** Spinal infarctions have been described associated with or secondary to many different diseases. Because data are obtained from small personal series, and often single case reports, it is difficult to determine the true frequency of each etiology. Therefore, the order of the following etiologies does not correlate with their particular incidence. Patients with spinal cord infarction tend to be younger than those with cerebral infarction, more commonly female and less commonly hypertensive (Naess and Romi 2011).

Etiologically, infarctions are divided into noniatrogenic, iatrogenic, venous infarcts, and spinal transient ischemic attack (See Table 1).

**Table 1. Etiology of Spinal Cord Infarctions**

**Noniatrogenic etiologies**

- Diabetic arteriopathy (Toole 1990)
- Atherosclerosis (Sandson and Friedman 1989)
  - Associated renal transplants (Bruno and Adams 1988)
- Inflammatory or infectious
- Syphilis (Bogousslavsky and Caplan 1995)
- Tuberculosis (Bogousslavsky and Caplan 1995)
- Arachnoiditis (Bogousslavsky and Caplan 1995)
- Herpes zoster (Sandson and Friedman 1989)
- HIV/varicella-zoster virus (Kenyon et al 1996)
- Pregnancy/varicella zoster virus reactivation (McNamara and Allworth 2016)
- Cocciidiomycosis (Wrobel and Rothrock 1992)
- Schistosomiasis (Liblau et al 1991)
- Cryptococcal (Bogousslavsky and Caplan 1995)
- Mucormycosis (von Pohle 1996)
- Sarcoidosis (Toole 1990)

• Vasculitic
  - Lupus (Sandson and Friedman 1989)
  - Giant cell arteritis (Gibb et al 1985)
  - Panarteritis nodosa (Sliwa and Maclean 1992)
  - Postinfectious/vaccination? (Sandson and Friedman 1989)
  - Granulomatous angiitis (Giovanini et al 1994)

• Non-inflammatory arteriopathies dissection (eg, congenital afibrinogenemia) (Laufs et al 2004)
  - Cervical spondylosis (anterior spinal artery compression) (Hughes and Brownell 1964)
  - Continuous hyperextension (Surfer myelopathy) (Shuster and Franchetto 2011)
  - Intervertebral foraminal disease (Ram et al 2004)
  - Lumbar artery compression by the diaphragmatic crus (Rogopoulos et al 2000)

• Aortic disease
  - Dissecting aortic aneurysm (Sandson and Friedman 1989)
  - Dissecting radicular artery aneurysms (Sato and Roccatagliata 2012)
  - Aortic occlusion/thrombosis (Fujigaki et al 1992)
  - Trauma (Hughes 1964)
  - Takayasu disease (Nair et al 1985)
  - Marfan syndrome (Wityk et al 2002)

• Embolic
  - Nucleus pulposus embolism (Hughes 1971)
  - Caisson disease (decompression sickness) (Sandson and Friedman 1989)
  - Atheromatous (Sliwa and Maclean 1992)
  - Paradoxical embolization (Mori et al 1993)
  - Atrial myxoma (Sandson and Friedman 1989)
  - Fibroelastoma (Pello and Ashkenazi 2011)
  - Endocarditis (Sandson and Friedman 1989)

• Hypoperfusional states (Singh et al 1994)
  - Cardiac arrest (Cheshire et al 1996)
  - Cardiac tamponade (Lin et al 2002)

• Disc protrusion (Bogousslavsky and Caplan 1995)
• Vertebral artery lesions (Toole 1990)
  - Cervical hyperextension injuries
  - Fractures with spine dislocation
  - Vertebral artery dissection (Bergqvist et al 1997)

• Tumor (Toole 1990)
  - Radicular artery compression

• Recreational drugs
  - Heroin (Malik and Woolsey 1991)
  - Cocaine (Di Lazzaro et al 1997)
  - Traumatic needle stick (Joseph 2004)
• Hypercoagulable states
  - Antiphospholipid syndrome (Hasegawa et al 1993)
  - Protein S deficiency (Ramelli 2001)
  - Congenital afibrinogenemia (Bas et al 2009)
• Anemia (Cheshire et al 1996)
• Sickle cell anemia (Rothman and Nelson 1990)
• Moyamoya disease (arterial fibrosis) (Kiriakov and Odinokova 1989)
• Malignant histiocytosis (infiltrative) (Falguera et al 1987)
• Malignancy (cholangiocarcinoma) (Thar et al 2015)
• Intravascular malignant lymphomatosis (IML) (Liu et al 2009)
• CADASIL (Hutchinson et al 1995)
• Cryptogenic (Monteiro et al 1992)

Iatrogenic causes
• Aortic surgery (risk increases if previous colectomy) (Salam et al 1993)
  - Aortic coarctation
  - Aortic aneurysms
• Gastrectomy (Cheshire et al 1996)
• Surgery for mesenteric vascular occlusion (Bbairaktari et al 2007)
• Thoracolumbar sympathectomy (Hughes and MacIntyre 1963)
• Coronary artery bypass surgery (Rossi et al 2008)
• Retropertitoneal tumor resection (Linz et al 1997)
• Pneumonectomy (radicular artery ligation) (Dawson and Potts 1991)
• Scoliosis surgery (radicular artery ligation) (Dawson and Potts 1991)
• Thoracoplasty (radicular artery ligation) (Dawson and Potts 1991)
• Intra-aortic balloon pump (Singh et al 1983)
• Hepatic transplantation (arterial reconstruction) (Goss et al 1997)
• Renal artery embolization
• Pineal region tumor resection (positional?) (Nitta et al 1997)
• Posterior fossa surgery (Martinez-Lage et al 2009)
• Resection thoracic dumbbell neuroblastomas (Boglino et al 1999)
• Anterior cervical fusion (Baba et al 1993)

Drugs
  - Neoarsphenamine (Sliwa and Maclean 1992)
  - Epidural/spinal anesthesia (vasoconstriction?) (Sliwa and Maclean 1992)
  - Intrathecal phenol (Hughes 1970)
  - Zolmitriptan (Vijayan and Peacock 2000)
  - Carmustine and cisplatin (Wang et al 2000)
  - Nitroglycerin (sudden hypotension) (Huang et al 2005)
  - Sildenafil (Walden and Castillo 2012)
• Dural fistula embolization/surgery (if common vessels) (Muraszco and Oldfield 1990)
• Transposition of the great arteries (Lemke et al 1996)
• Embolization of bronchial artery (Cheng et al 1996)
• Endovascular occlusion spinal dural arteriovenous malformation (Mascalchi et al 1998)
• Epidural catheter placement (hyperlordosis) (Amoiridis et al 1996)
• Central line placement (hemothorax) (Williams et al 2003)
• Cerebral angiography (Bejjani et al 1998)
• Peripheral angiography (Bozkurt et al 2003)
• Coronary angiography (Aramburu et al 2000)
• Coronary angioplasty (Vatankulu et al 2010)
• Transcatheter arterial chemoembolization of hepatocellular carcinoma (Park et al 2012)
• Endovascular ultrasound celiac plexus neurolysis (Fujii et al 2012)

Venous infarction
• Sepsis (Sliwa and Maclean 1992)
• Embolic (Sliwa and Maclean 1992)
- Fibrocartilaginous (Roa et al 1982)
- Caisson disease (Sliwa and Maclean 1992)
- Hypercoagulable states (Sandson and Friedman 1989)
  - Associated to malignancies (Sliwa and Maclean 1992)
  - Acute pancreatitis (Wei et al 1997)
- Intramedullary tumor (Sandson and Friedman 1989)
- Venous thrombosis (Roa et al 1982)
- Meningitis (Mathew et al 1993)
- Endoscopic sclerotherapy for esophageal varices (Heller et al 1996)

**Spinal transient ischemic attack**

- Paget disease (flow diversion to bone) (Porrini et al 1987)
- Dural arteriovenous malformation (Bogousslavsky and Caplan 1995)
- Atherosclerosis (Sandson and Friedman 1989)

**Epidemiology**

**Etiology of nontraumatic hemorrhages.** Epidural and subdural hematomas can occur spontaneously in patients receiving anticoagulants or in those who have a coagulopathy or blood dyscrasia. Sometimes they occur after straining or coughing. They have also been reported after lumbar puncture in the setting of coagulation dysfunction (Bogousslavsky and Caplan 1995).

Other risk factors include alcoholism and the use of antirheumatic drugs. Portal hypertension also can predispose patients to spinal bleeding (Mattie et al 1987).

The source of the epidural bleeding is of arterial origin. Vascular malformations are less frequent causes of epidural and subdural hematomas. Vertebral hemangiomas are also an unusual source of bleeding (Mattie et al 1987; Toole 1990).

Hematomyelia can be the result of intraparenchymal arteriovenous malformations, bleeding diatheses, hemorrhages into spinal tumors (especially ependymomas), cavernous angiomas, and inflammatory myelitis (McCormick and Stein 1990; Bogousslavsky and Caplan 1995). It has been described in association with abdominal aortic coarctation (Iwata et al 1997).

Nontraumatic spinal subarachnoid hemorrhages are usually due to arteriovenous malformations or coagulation defects. Other causes include rupture of an inflamed vessel, aneurysmal rupture, and endometriosis (Lombardo et al 1968; Toole 1990; Bogousslavsky and Caplan 1995; Chen et al 2001).

**Dural arteriovenous fistulas (type I arteriovenous malformation).** They represent a low-flow shunt in the dura covering either the proximal portion of a nerve root or the adjacent spinal dura (or both). They are supplied by a dural artery, which is a branch of the intervertebral segment of a spinal artery. The fistula empties in the coronal venous plexus of the cord, producing venous congestion of the cord (the intrathecal venous system is valveless). Venous congestion is the physiopathological mechanism in these patients, although spontaneous thrombosis has also been reported. This syndrome, known as Foix-Alajouanine syndrome, typically occurs in older males and is characterized by a slowly progressive myelopathy (Muraszko and Oldfield 1990; Renowden and Molyneux 1993).

**Intradural arteriovenous malformations.** These lesions are usually located partially or totally within the pia of the spinal cord. Two types are differentiated: glomus type (type II arteriovenous malformation), which is usually a packed mass of blood vessels located in the anterior cord and is the most common type, and juvenile type (type III arteriovenous malformation), in which an elongated mass usually is supplied by several medullary arteries. They have huge flow, and vertebral and paraspinal involvement. A third type, a direct arteriovenous fistula (type IV arteriovenous malformation) (anterior spinal artery–anterior spinal vein) that lies in the cord parenchyma or surface, is rare (Hayakawa et al 1980; Muraszko and Oldfield 1990; McCormick 1993; Renowden and Molyneux 1993).

These fistulas have a rapid flow, accounting for the bruit that is often present. The usual presentation is an acute hemorrhage. Other symptoms are transient cord ischemia, due to a steal phenomena, and a local mass effect, exerted through venous varices and arterial aneurysms. Cord arteriovenous malformations have been described associated
with Kartagener syndrome (triad of situs inversus totalis, sinusitis, and bronchiectasis) (Hayakawa et al 1980; Muraszco and Oldfield 1990; McCormick 1993; Renowden and Molyneux 1993).

**Cavernous malformations.** These lesions are being reported more frequently as MRI techniques improve. They are capillary vessels arranged in a sinusoidal network. They can cause hemorrhage, local mass effect, and neurotoxic effect of hemosiderin (McCormick 1993), and they are often associated with cryptic venous malformations found during surgery (Vishteh et al 1997). They may be associated with previous radiation (Maraire et al 1999).

**Spinal cord infarctions.** The frequency of ischemic myelopathy is unknown. It is a relatively rare disease, which is frequently described in small series or even case reports. In the large retrospective series of general spinal cord injury units, El-Tolarei and Fuller attributed to ischemia 1% of 2500 patients (Dawson and Potts 1991); Mazaira-Alvarez and colleagues found a 0.6% incidence of ischemia in 1100 patients (Mazaira-Alvarez et al 1982).

Reviews of autopsy records show variable figures. In a series of 300 autopsies, Mannen found ischemic myelopathies in 25 cases (8.3%). Blackwood only found 5 cases (0.1%) in a series of 3737 autopsies (Sandson and Friedman 1989). Sandson and Friedman found that spinal strokes represented 1.2% of the admissions to their stroke service in a 4-year period (Sandson and Friedman 1989).

Vascular surgery data show that spinal cord infarctions occur in 4% to 30% of persons who undergo elective thoracoabdominal aneurysm repair, and it may be as high as 40% in the repair of acute aortic dissection. The complication rate after aortic coarctation surgery has been estimated around 1% to 5% (Mazaira-Alvarez et al 1982; Salam et al 1993; Grabenwober et al 1994; Wisselink et al 1994). However, infrarenal abdominal operations may have a better prognosis, as shown by a review of 1112 patients in which only 2 cases of spinal cord infarction occurred (Dimakakos et al 1996).

Spinal venous infarctions are probably rarer than the arterial infarctions, although the exact frequency is unknown (Sliwa and Maclean 1992).

The incidence of spinal vascular malformations is probably low, although the dural type of fistula is not uncommonly found in older patients (McCormick 1993; Bogousslavsky and Caplan 1995).

Nontraumatic spinal and epidural hematomas are considered to be rare diseases. A series from Mattle and colleagues showed 10 cases seen in a period of 10 years (Mattle et al 1987).

**Prevention**

No data are available regarding primary and secondary prevention for spinal cord infarctions in the general population other than treating any underlying disease that could represent a potential causative condition. There are data regarding prevention of iatrogenic spinal cord infarctions that occur during abdominal surgery. There are suggestions about the importance of preventing hypotensive episodes during surgery (Singh et al 1994).

Hypothermia is a protective mechanism for preventing ischemia during surgery in both experimental and clinical studies (Grabenwober et al 1994; Wisselink et al 1994).

A higher rate of spinal infarction is associated with prolonged aortic clamping times as well as poor surgical techniques. A history of previous colectomy also increases the chance of ischemic infarction. The latter is due to ligation of the hypogastric arteries during the preceding colectomy, which eliminates a critical collateral source of flow, particularly in individuals where the great radicular artery has a high origin (Salam et al 1993). Preoperative detection of the artery of Adamkiewicz by MRA may predict patients at lower risk for cord infarct after thoracoabdominal aortic aneurysm surgery (Hyodoh et al 2005).

The use of somatosensory evoked potentials during surgery is a common practice to minimize the ischemic injury in the spinal cord. The technique of stimulating at the level of the spinal cord by an epidural needle has proved to be more reliable than the stimulation of the peripheral nerves in the lower extremities. The latter presents frequent false positives due to lower-limb ischemia. The epidural stimulated evoked potentials seem to provide a reliable way of continuously monitoring for cord ischemia (Drenger et al 1992). Epidural cooling is a technique that has been proposed to reduce the rate of spinal cord infarction during thoracoabdominal aneurysm repair (Cambria et al 2000). Operative CSF drainage and naloxone administration are used to prevent postoperative paraplegia (Tefera et al 2000).
The use of cytoprotective agents, such as dextrorphan, has been shown to be beneficial in minimizing ischemic cord damage. The use of high doses of barbiturates also has been advocated. Experimental studies have also shown protective effects from the combination of deferoxamine and allopurinol to prevent cord ischemia in the swine model. Intrathecal magnesium sulfate can protect the thoracic cord in dogs (Weinstein 1993; Qayumi et al 1994; Rokkas et al 1994; Simpson et al 1994). Curcumin, a diarylheptanoid has showed neuroprotection in a rabbit spinal cord injury model (Liu et al 2013).

Mannitol and calcium channel blockers have been used to minimize reperfusion injury after the clamping (Weinstein 1993).

Venous infarctions may be prevented by avoiding the known precipitating factors, such as hypercoagulable states or sepsis.

Epidural, subdural, and cord hemorrhages may be prevented by correcting coagulation dysfunction and monitoring the hematological parameters in anticoagulated patients. Avoiding high-risk maneuvers in those cases, such as lumbar punctures, may lower the incidence. Avoiding high-risk combinations, particularly nonsteroidal anti-inflammatory drugs in alcoholic patients with a coagulopathy from liver disease, has been advocated (Mattle et al 1987).

Surgery or embolization in selected cases may be beneficial in preventing complications from arteriovenous malformations (Muraszco and Oldfield 1990).

**Differential diagnosis**

Spinal cord infarctions, as well as hematomyelia, need to be delineated from other causes of acute myelitis. The picture needs to be differentiated from an acute cord compression, such as a herniated disc, tumor, or abscess. Other causes of acute transverse myelitis include infectious (bacterial, viral, Lyme, larva migrans), demyelinating myelopathies (multiple sclerosis or Devic disease, postinfectious), metabolic (arsenic, sulfas, heavy metals), trauma, or postradiation. Acute polynuropathies, such as acute inflammatory demyelinating polyneuropathy, also may be confused with infarction of the spinal cord (Brass and Stys 1991). The pain associated with cervical cord infarctions can mimic angina pectoris (Cheshire 2000).

Besides the conditions mentioned above, spinal epidural and subdural hematomas need to be differentiated from other forms of epidural collections, such as abscesses, metastases, or granulomatous disease. Occasionally patients with previous cardiac history were thought to have an acute myocardial infarction (Mattle et al 1987).

The differential diagnosis for arteriovenous malformations is the same as for the acute hemorrhagic syndromes described above. If the presentation is that of intermittent cord ischemia, differential diagnosis should be made with vascular claudication of the lower extremities and Paget disease of the bone (Porrini et al 1987; Toole 1990; Bogousslavsky and Caplan 1995).

Arteriovenous malformations (especially dural) need to be differentiated from other forms of subacute progressive myelopathy, such as demyelinating disease, metabolic or toxic myelopathies, spinal cord tumors, granulomatous disease, infections, and subacute extrinsic cord compressions by abscesses, tumors, or discs. Cervical spondylosis, syringomyelia, and motor neuron disease need to be included in the differential as well (Muraszco and Oldfield 1990).

**Diagnostic workup**

The evaluation for an acute spinal cord infarction includes neuroimaging studies to exclude the presence of an acute extrinsic spinal cord compression or an intraspinal mass. An MRI of the spinal cord should provide the needed information. (embed="pagecomponents/media_embed" entry_id="8920")

Series regarding the use of MRI report a high yield. Approximately 93% of T2-weighted images detect the ischemic lesion at the time of the onset of symptoms. T1-weighted images are less reliable because 70% appear isointense at onset and only 18% are hypointense (Fortuna et al 1995). Diffusion-weighted MRI may be useful for detection of early ischemic cord lesions (Weidauer et al 2002).

MRI with gadolinium may be useful in following patients because of the characteristic "sequential changes" that infarcts show with time (nonenhancement in acute stages, enhancement at "medium age" of 1 to 4 weeks, and then a
progressive tendency to nonenhancement). This pattern helps differentiate these lesions from spinal cord tumors (Hirono et al 1992; Pelser and Van Gijn 1993; Fortuna et al 1995; Tseng et al 1995). Signs of vertebral body infarction on MRI are a useful radiological marker of spinal cord infarction because these 2 conditions are often associated (Faig et al 1998).

Admitting laboratory tests should include complete chemistry and cell blood count, sedimentation rate, and VDRL. If infectious or demyelinating disease is suspected, lumbar puncture may be indicated. If acute inflammatory demyelinating polyneuropathy cannot be excluded, emergent nerve conduction studies may be useful. Other forms of myelitis should be excluded by the appropriate specific tests, if clinically indicated.

The workup for hemorrhagic syndromes should include complete coagulation studies and liver function tests. MRI is now the first test to be ordered when an arteriovenous malformation is present. Intradural arteriovenous malformations may be seen as flow void and a low-intensity signal in T1-weighted images. However, the most common type of arteriovenous malformation in the spine, the dural fistulas, may be missed by MRI. In cases of dural fistulas, cord swelling is sometimes evident in both T1 and T2 images. T2 hypointensity involving the cord periphery may be a sign suggestive of spinal dural arteriovenous fistula (Hurst and Grossman 2000). Contrast-enhanced MR angiography of the spinal vessels provides improved characterization of the spinal vasculature and may be useful in screening for spinal dural fistulae (Bowen and Pattany 2000). Myelography is a sensitive method for detecting spinal arteriovenous malformations (Muraszco and Oldfield 1990).

Once the lesion has been visualized by MRI or myelogram, an arteriogram is necessary to determine the origin of the feeding vessels in those patients in whom surgical intervention may be considered (Muraszco and Oldfield 1990).

Management

There is no specific approved treatment for acute spinal cord infarctions. The relative rarity of the disorder impedes organization of clinical trials. Intravenous rtPA has been used anecdotally, but the safety and efficacy is not known (Etgen and Hocherl 2016). Treating the causative or underlying condition may be important in the acute setting. The use of anticoagulation, antiplatelets, high-dose steroids, cord revascularization, opiate antagonists, and sympathectomy has been advocated, but their role in the acute setting is unknown. Although neuroprotective agents are beneficial in the experimental setting, their clinical role remains uncertain. Other neuroprotective maneuvers used during aortic surgery may eventually have a role in the acute spontaneous infarction. The availability of an adequate facility for treating patients with spinal cord disease also may have a tremendous impact on outcome thanks to adequately managing specific medical complications and initiating appropriate rehabilitation (Toole 1990; Dawson and Potts 1991).

Case reports have shown a possible beneficial effect of intra-arterial (Adamkiewicz) injection of dexamethasone and urokinase (Baba et al 1993). Spinal epidural and subdural hematomas should be decompressed with laminectomy procedures. Occasionally, fresh frozen plasma needs to be given to correct underlying coagulopathies. The above described general measures for managing a myelopathy should be applied in this case also (Mattie et al 1987).

Spinal dural arteriovenous malformations can either be treated by surgery (usually 80% to 90% good results after incision) or embolization. The latter has a high recanalization rate (Muraszco and Oldfield 1990; McCormick 1993).
history of these lesions (McCormick 1993).

Special considerations

Pregnancy

Pregnancy may predispose individuals to venous infarctions, but only isolated case reports can be found in the literature (Roa et al 1982).

Anesthesia

Spinal infarcts secondary to spinal anesthesia have been reported. It is not known if the cause is the anesthetic itself or the vasoconstrictor that is concomitantly administered (Hughes 1970; Sliwa and Maclean 1992).

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References especially recommended by the author or editor for general reading.

### ICD and OMIM codes

**ICD codes**

**ICD-9:**
- Vascular myelopathies: 336.1

**ICD-10:**
- Vascular myelopathies: G95.1

### Profile

**Age range of presentation**

- 0-01 month
- 01-23 months
- 02-05 years
- 06-12 years
- 13-18 years
- 19-44 years
- 45-64 years
- 65+ years

**Sex preponderance**

male=female

**Family history**

family history may be obtained

**Heredity**

none

**Population groups selectively affected**

none selectively affected
Occupation groups selectively affected

none selectively affected

**Differential diagnosis list**

other causes of acute myelitis
acute cord compression
herniated disc
tumor
abscess
bacterial infections
viral infections
Lyme disease
larva migrans
demyelinating myelopathies
multiple sclerosis
Devic disease
postinfectious causes
metabolic causes (arsenic, sulfas, heavy metals)
trauma
postradiation
acute polyneuropathies
acute inflammatory demyelinating polyneuropathy
angina pectoris
other forms of epidural collections
abscesses
metastases
granulomatous disease
acute myocardial infarction
vascular claudication of the lower extremities
Paget disease of the bone
subacute progressive myelopathy
demyelinating disease
metabolic or toxic myelopathies
spinal cord tumors
granulomatous disease
subacute extrinsic cord compression (by abscesses, tumors, or discs)
cervical spondylosis
syringomyelia
motor neuron disease

**Other topics to consider**

Cavernous malformations
Cerebral arteriopathies
Drug-induced cerebrovascular disease
Neurologic complications of diseases of the aorta

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