Spinal cord infarction caused by extensive aortic intramural hematoma

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Abstract

We report the case of a 62-year-old Caucasian man, an ex-smoker, who presented to the emergency room complaining of intense lower back pain followed by immediate bilateral loss of inferior limbs motor function. Clinical examination showed complete paralysis and parastesia in both legs, while pain and temperature sensory examination revealed a sensory level at dermatome T6, with normal touch, vibration, and position senses. His blood pressure was 190/100 mmHg. Computed tomography demonstrated dilated thoracic aorta (maximum diameter 44 mm) and abdominal aorta (maximum 58 mm), with extended intramural hematoma (IMH), thus establishing the diagnosis of type A aortic IMH complicated with paraplegia through spinal perfusion deficit. Due to the extension of the lesions, surgical intervention for aortic repair was considered at high risk while cerebrospinal fluid drainage was not recommended by the neurologist. The patient remained stable while hospitalized and was released from the hospital with mild improvement of neurological deficiencies.

Keywords: spinal cord infarction, aortic hematoma, anterior spinal artery syndrome, spinal arteries, paraplegia, aortic dissection.

Introduction

Spinal cord infarction (SCI) is a rare event representing about 2% of all neurovascular vascular diseases [1]. It is caused by acute blood supply interruption in a region of the spinal cord and the neurological presentation is largely defined by the vascular territory involved [2–4]. Although the risk factors for spinal ischemic stroke seem to be similar to those of cerebral stroke, the incidence of the two clinical entities shows significant discrepancy, with the former being a rare event [5]. This may be related to the fact that the spinal cord arteries form an extensive anastomotic network being less prone to atherosclerotic disease compared to the cerebral arteries [6].

Anterior spinal artery syndrome is the most frequent cause of spinal cord stroke. It is characterized by motor function impairment and complete loss of sensation of pain, temperature, and of light touch below the injury level, sometimes with autonomic dysfunction [7].

The involvement of the posterior spinal artery is exceedingly rare. By affecting the posterior columns of the spinal cord, the clinical presentation is with paresthesia and abolition of vibration and proprioception below the level of the ischemia. Sometimes a total transverse SCI could affect both anterior and posterior spinal artery territory. This is very often misdiagnosed as transverse myelitis [8]. Back or neck pain, usually located at the level of the lesion, is often present [9]. Patients with acute onset proprioceptive and walking impairment, especially bladder dysfunction, have a poor prognosis [10].

Most commonly, anterior spinal artery syndrome is reported as a procedure hazard (aortic, spinal, and general surgery) [11, 12], albeit most spinal cord infarcts are spontaneous.

Spontaneous spinal cord infarcts (nonprocedural, non-traumatic) have a definite cause in less than 50% of patients, that is why very often they are misdiagnosed. The mechanism of spontaneous SCI is usually idiopathic associated with aortic pathology, vertebral artery dissection, atherosclerosis. Other rarer reported causes are fibrocartilaginous embolism [13], cardiac embolism [14], decompression sickness from scuba diving [15], coagulopathy, and hematological disorders [16], systemic hypotension, spinal arteriovenous malformations, and vasculitis [17].

The diagnosis is challenging and usually based on anamnesis, clinical exam, and computed tomography (CT) or magnetic resonance imaging (MRI), which are also useful in excluding other conditions. The most precise tool in identifying ischemic lesions of the spinal cord is the MRI [18]. Zalewski et al. proposed three major criteria for SCI diagnosis: clinical exam with the rapid development of severe deficits; neuroimaging, usually MRI of the spine, that exclude compression and specific imaging findings; the analysis of cerebrospinal fluid (CSF), highlighting non-inflammatory findings [19].

Aim

Although SCI can be determined by acute aortic dissection, in extremely rare cases it can be a consequence...
of an intramural hematoma (IMH). In the following paragraphs, we report the case of an aortic IMH which led to SCI.

**Case presentation**

A 62-year-old Caucasian male presented to the emergency room in February 2019 with abrupt onset lower back pain while shoveling snow. Within minutes from the onset of pain, he experienced a bilateral loss of inferior limbs motor function. He had a history of high blood pressure but without optimal medical treatment. He was an ex-smoker and regular alcohol drinker.

Physical examination revealed a preserved mental status and cognition. The heart rate was 65 beats/min, blood pressure was 190/100 mmHg, with no peripheral pulse deficits. At the abdominal examination, a distended bladder was palpable requiring the placement of a urinary catheter. The rectal examination showed a relaxed anal sphincter tone. Neurological examination revealed complete paralysis and parasthesia of inferior limbs, with pain and temperature sensory level at the dermatome T6 level (intersection of the midclavicular line and the horizontal level of the xiphoid process). Additionally, reflexes were absent in the lower limbs, without plantar response. Touch, vibration, and position senses were normal.

The biological tests showed high sensitivity cardiac troponin I (hs-cTnI) of 82.9 ng/L and D-dimer >5 μg/mL. All the other routine blood assets were within normal ranges. The venereal disease research laboratory (VDRL) and human immunodeficiency virus (HIV)-1 and HIV-2 antibodies were negative.

The patient underwent CT examination with intravenous contrast, which revealed dilated thoracic aorta (maximum diameter 44 mm) and abdominal aorta (maximum 58 mm) with extended IMH (Figure 1, A and B; Figure 2). Inward displacement of intimal calcifications was noted and no evidence of intima tear, dissection flap, or penetrating ulcers was present at the time of the CT examination. Also, there was no aortic regurgitation or pericardial effusion on transthoracic echocardiography. MRI excluded acute disc herniation, epidural hematoma and other etiologies that could explain the symptomatology (Figure 3).

**Discussions**

Spinal cord ischemia is a rare condition that leads to severe disability and increased mortality. The most common association is with aortic pathology or surgery although other causes, such as trauma, thromboembolic disease, chronic inflammatory conditions, or tumors were described [20–24].
Spinal cord ischemia presenting with acute paraplegia is extremely rare (1–3% of patients) [25]. We reported the case of a patient with acute onset of paraplegia due to an extensive type A IMH with a shearing effect on the vessels that originated from the aorta.

The spinal cord blood vascularization is represented by the spinal arteries (one anterior and two posterior), with extensive collateral circulation and numerous radicular or segmental medullary arteries. The largest anterior segmental medullary artery and the most important artery that supplies the spinal cord is called the artery of Adamkiewicz [26]. In our patient, the aortic hematoma interrupted the blood flow of the spinal artery and collaterals causing SCI.

Symptoms of SCI depend mainly on the extent and location of the infarction. Based on the injury location, incomplete spinal cord syndromes are classified in the anterior cord, posterior cord, central cord, Brown-Sequard, cauda equina, and conus medullaris syndrome [26]. Anterior cord syndrome is the most frequent and consists of motor function impairment and complete loss of pain, light touch, and temperature sensation below injury level. The preliminary diagnosis is mostly clinical, given that the sensitivity of imaging techniques is relatively low in early stages. However, the MRI is used to exclude other etiologies of anterior spinal artery syndrome, and during follow-up to evaluate the expansion of infarction.

There are three major types of acute aortic syndromes. The most frequent is aortic dissection, followed by IMH and penetrating atherosclerotic ulcer [27]. IMH is defined as bleeding contained within the media, without communication between the hematoma and aortic lumen and represents about 5% to 20% of acute aortic syndromes [27]. It can be caused by rupture of vasa vasorum due to degenerative changes in the media with intact intima, by microscopic intimal tear, by bleeding associated with penetrating atherosclerotic ulcer, and rarely by tumors of the aorta or trauma [28, 29]. Clinical presentation includes acute chest or back pain, while signs of tissue hypoperfusion are much less common than in aortic dissection.

Transesophageal echocardiography, CT examination or MRI are the most useful imaging techniques for acute aortic syndromes diagnosis. Distinguishing IMH between other similar syndromes like aortic aneurysm with intraluminal thrombus or classical dissection with thrombosed false lumen can be difficult, like in our case, but some radiological characteristics guide the diagnosis (Table 1) [30].

| Table 1 – Radiological comparison between aortic hematoma and thrombus |
|---------------------------------------------------------------|
| Intramural hematoma | Intraluminal thrombus |
|---------------------|----------------------|
| Long segment        | Shorter segment      |
| Normal diameter of aorta | Aneurysmal aorta     |
| Smooth interface    | Irregular/lobulated interface |
| Displaced intimal calcification | No displaced intimal calcification |

Treatment of IMH aims to prevent progression to dissection and aortic rupture. The primary goal of medical therapy is to reduce aortic wall stress. β-Blockers are the first line of treatment, commonly associated with vasodilators to obtain optimal blood pressure. Depending on the location, the management can be either conservative or surgical. Indications for surgical treatment depend on the anatomy of the lesion, patient symptomatology, and comorbidities [29]. IMH’s natural history varies from patient to patient, it may extend, progress, regress, or reabsorb. In most patients, a progression to aortic dissection will be seen [31–33]. In Asian patients, IMH has a more benign evolution so aggressive medical therapy and intensive follow-up by imaging showed promising results [29]. After hospital discharge, follow-up should be made by a specialized team every three months in the first year and after that, depending on aortic size, every six to 12 months [29, 32].

Some studies reported a reversal of paraplegia after surgical treatment of aortic dissections [22] but without benefits in treating spinal cord ischemia caused by IMH. An option could be the reimplantation of the artery of Adamkiewicz during surgery, but this artery has a variable origin, being difficult to localize [29]. Also, in the replacement of the thoracic aorta, collateral circulation of the spinal cord will be affected, aggravating the ischemia [33]. In our patient, the multidisciplinary team conveyed for conservative treatment, considering the significant peri-procedural risk.

The prognosis of SCI depends on etiology, severity, and the regions involved. Until presently, no medical treatment was proven beneficial in correcting cord ischemia, although several studies involving corticosteroid therapy or CSF drainage have been reported [34–36]. CSF drainage was not available immediately in our Center and given the long time passed since onset we considered the spinal lesion as irreversible.

The principle of treatment is in the first place reducing spinal cord hypoperfusion, as soon as possible. This can be done by elevation of mean arterial blood pressure (MAP) [34]. Based on the highest level of evidence, MAP goals of 85–90 mmHg for 5–7 days should be considered [37]. Another way of improving spinal cord perfusion is by decreasing CSF pressure, and this is done by CSF drainage [35].

To our knowledge, there have been only six other cases described in the literature besides our case (Table 2) [38–43]. For the first time, in 1996, Ferguson et al. [38] reported a case of IMH with spinal cord ischemia in a 62-year-old hypertensive male. The CT scan revealed a type A IMH. Although with blood pressure control alone neurological deficits disappeared, three weeks later the IMH evolved in a type A aortic dissection. Compared to the reported cases, our patient showed a minimal recovery of neurological function. One year follow-up indicated no progression of the IMH and no signs of aortic dissection.

Conclusions

SCI is a rare condition commonly associated with aortic diseases and procedures. The diagnosis is challenging and usually based on clinical examination and imagistic. The prognosis is strongly related to the etiology, severity, and regions involved. The only treatment proven beneficial in certain cases is increasing MAP in the immediate post-injury period being, and sometimes CSF drainage. SCI is a rare presentation of aortic IMH. Because of atypical clinical presentation, the diagnosis of the acute aortic syndrome could be overlooked or delayed, thus aorta imaging is an important step in early diagnosis.
| Authors (Year) | Age/Gender | Risk factors | Symptoms | Segment involved | Treatment and evolution |
|---------------|------------|--------------|----------|-----------------|-------------------------|
| Ferguson et al. (1996) [38] | 62/M | Hypertension | Sudden onset of intercapsular pain, progressive symmetrical loss of lower limb function. | type B IMH | Neurological deficits gradually resolved with blood pressure control alone, but three weeks later the IMH evolved in a type A aortic dissection. |
| Motoyoshi et al. (2003) [39] | 46/M | Hypertension | Sudden onset of chest pain, progressive symmetrical loss of lower limb sensory. | type A IMH | Cerebral spinal drainage was done for six days with slow disappearance of neurological symptomatology and spontaneous regression of ascending aortic IMH 28 days later. |
| Zhu et al. (2012) [40] | 42/M | Heavy smoking, hypertension | Severe chest pain, motor impairment in lower extremities. | type B IMH | Blood pressure control with neurological recovery quickly, but IMH progressed to aortic dissection, and surgery was performed. |
| Yu et al. (2013) [41] | 42/F | Preeclampsia, hypertension | Sudden onset subternal chest pain radiating to back followed by increasing bilateral lower extremity numbness and weakness. | type B IMH | Evolution was favorable with blood pressure control and CSF drainage. |
| Uillery et al. (2015) [42] | 64/F | Hypertension, atrial fibrillation | Sudden chest and back pain followed by bilateral lower limb paraplegia. | type A IMH | Complete recovery of neurological function with blood pressure elevation and cerebral spinal drainage followed by surgery of the aorta. |
| Tsushima et al. (2019) [43] | 57/M | None | Presented with myocardial infarction, after coronary angiography he acutely developed complete paraplegia, paresis in the lower extremities. | type A IMH | Evolution was unfavorable and he died seven days later. |

CSF: Cerebrospinal fluid; F: Female; IMH: Intramural hematoma; M: Male; SCI: Spinal cord infarction.

Conflict of interests

The authors declare that they have no conflict of interests.

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Table 2 – Clinical features and prognosis of patients with SCI caused by aortic IMH reported in the literature
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