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Case Report

Idiopathic Spinal Cord Infarction Occurring in a Young Adult

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Abstract

Spinal cord infarction is a rare but serious disease that represents a diagnostic and therapeutic challenge for many physicians. It represents about 1% of all neurological vascular emergencies [1]. A multitude of risk factors are identified but the most common cause in young patient remains idiopathic [2].

Here we discuss about 18-year-old young woman who developed progressive tetraplegia after complaining of a sudden and intense back pain. Magnetic resonance imaging exposed an anterior spinal artery infarction from C3 to T1 in a delay of 72hours. Her evolution was marked by many complications such as vasoplegic shock and respiratory failure. After rehabilitation, she still suffers of serious after-effects to this day. However, according to certain prognostic criteria, a potential for functional recovery exists. This potential recovery must be known by physicians and transmitted during the catastrophic diagnosis announcement leading to a little hope for patient.

Keywords: Spinal cord infarction, Idiopathic cause, Magnetic resonance imaging, Prognosis

Introduction

Although a rare entity, Spinal Cord Infarction (SCI) is mostly characterized by a common clinical presentation, the Anterior Spinal Artery (ASA) syndrome. Some exceptional cases of posterior spinal artery syndrome are also described [4]. In ASA infarct, 100% of patients present dissociative anesthesia [2], weakness of upper and/or lower limbs depending on the level of lesion in the cord. A dissociative anesthesia implies a loss of pain and temperature sensation while the proprioception and vibrating sensitivity are spared. Approximatively 60% of cases described neck or back pain and sphincter disorders such as urinary retention or incontinence. Autonomic dysfunction can be observed, ranging from orthostatism to severe hypotension and shock. The duration of symptoms development is variable, estimated from a few minutes to 48hours. The peak of symptoms is reached within 12hours for half of patients, and within 72hours for most of affected people [4]. Rare cases of PSA infarct are characterized by a loss of deep sensitivity: proprioception and vibratory sense below the level of injury [4]. Motor impairment is in the background, weakness is mostly mild and transient. Due to the distribution of vascularization, ASA syndrome concerned both sides while PSA syndrome is more often unilateral.

There are multitudes of etiologies described in literature. The largest Meta-analyze recently published in 2022 [5], highlighted the idiopathic cause in first place (45%), thus confirming previous studies [2,4,6]. Other etiologies involve systemic and chronic conditions (arteriosclerosis, hypertension, diabetes) in 23%, aortic vascular pathologies (aortic dissection, aneurysm, coarctation) in 19,6%, thromboembolic events in 17,7%, compressive causes in 16,9%, trauma in 7%, hypoperfusion in 6%. SCI affects both men and women almost equally. As the non-iatrogenic cause was excluded of the Meta-analyse, it is important to note that aortic surgery is known as the second most frequent etiology in other studies. Currently, only three cases report protein S deficiency.

Case Presentation

An 18-year-old girl consult first her General Practitioner (GP) because of a sudden and intense neck pain irradiating in interscapulary appearing since waking up, without any notion of effort or trauma. A few hours later, she developed descending numbness and weakness of limb. She was then referred immediately by her GP to a neurologist and then to the Intense Care Unit (ICU) because of the extent of paralysis.

Her medical history was blank, and none of the patient's family had a history of cerebrovascular or hematological disorder. Her treatment includes oral contraception. No allergy was reported. At the admission, her vital signs were normal with no fever.

A neurological examination revealed a symmetrical flaccid tetraplegia below C5 associated with are flexia. Tactile and proprioceptive sensitivity were preserved while thermo-algesic sensitivity was absent below C3. She was also suffered from urinary and fecal incontinence. This neurological examination corresponds to a score ASIA-A.

A routine blood test including hematological biochemical and basic coagulation revealed normal results. The electrocardiogram was in sinusal rhythm, the cardiac monitoring shows no arrythmia. Cerebrospinal fluid was also normal: absence of figurative elements, normal proteinorachia and glycorrhachia. Cerebro-cervical magnetic resonance imaging on day 1, 12 hours after the start of symptoms, was negative even the diffusion-weighted image. The second medullar MRI, done on day 3 (Figure 1,2), revealed a hyper intensity on T2-weighted image and decreased apparent diffusion coefficient signal centered on the anterior cords from cervico-bulbar junction to T6, which is compatible with anterior spinal artery infarction. The angiography wasn't contributive to visualize anterior spinal artery, so it was not possible to exclude dissection, vascular malformation aneurysms or ischemic lesion around the ASA. During her stay in ICU, an extensive thrombophilia assessment was carried out by blood sample and made possible to exclude: lupus anticoagulant, homocysteine, antithrombin III deficiency, protein S antigen deficiency, protein C antigen deficiency, anticardiolipin antibodies, anticardiolipin-beta2-glycoprotein I complex antibody, antinuclear antibody, anti-neutrophil cytoplasmic antibody, APC resistance, Bcr-abl et Jak2V617. Only factor VIII was increased in the acute phase but was recontroled normal. A trans-thoracic and trans-esophageal echocardiography have excluded thrombo-emboligenic cardiopathy.

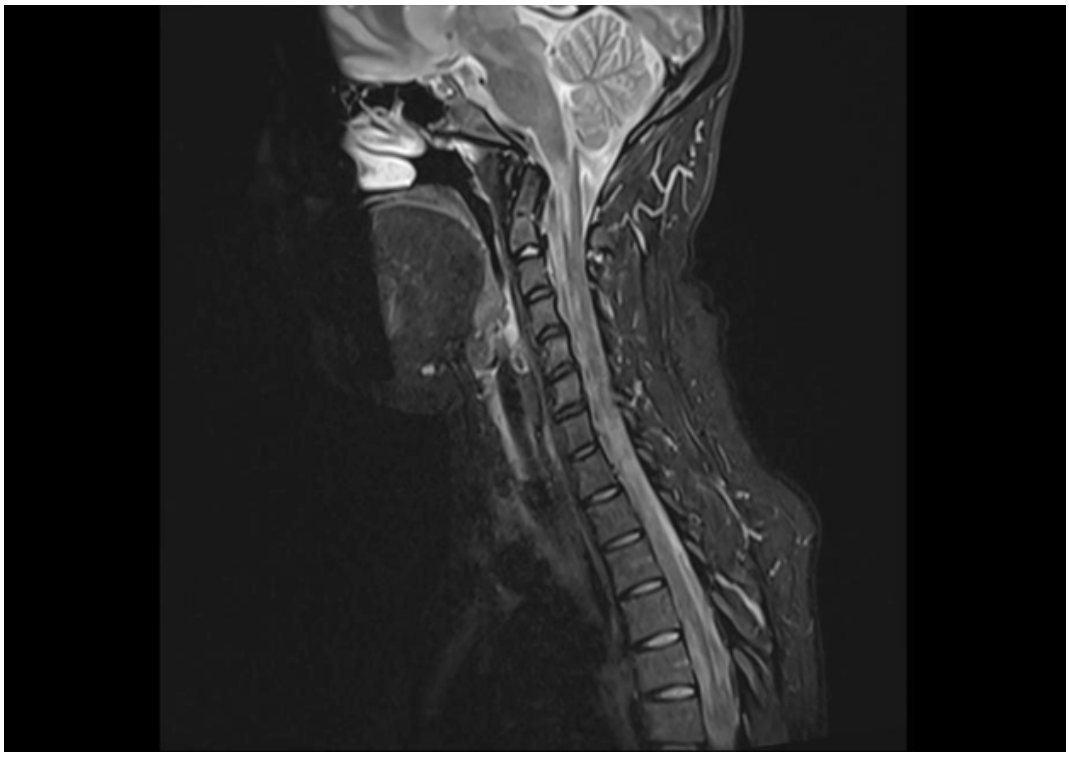

Figure 1: Day 3 MRI imaging displaying a large spinal cord infarction extended from cervical junction to D6.

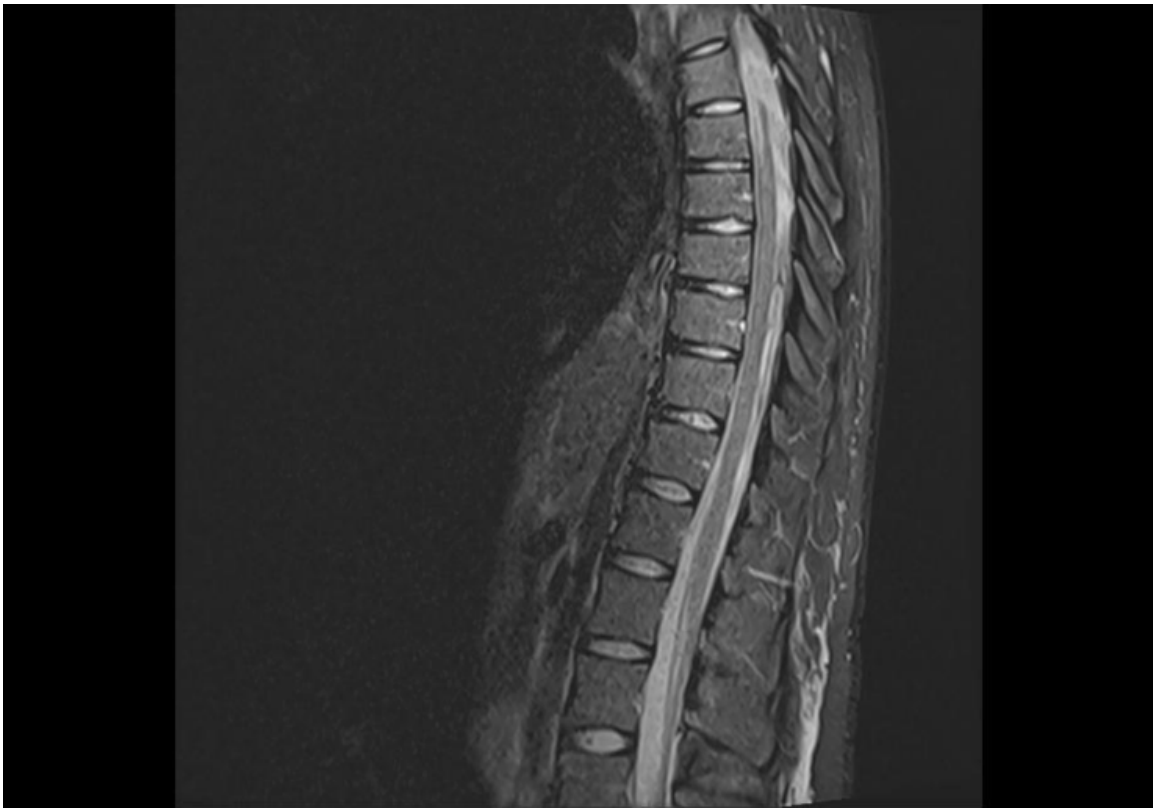

Figure 2: Day 3 MRI imaging showing SCI extending to D6.

Unfractionated heparin and antiplatelet aggregates were initiated immediately, for one month, followed by a prophylactic dose of Low-Molecular-Weight Heparin (LMWH).

Her short-term evolution was complicated by spinal shock on day two treated by infusion of noradrenalin during 24hours, following by constituted orthostatic hypotension. The patient also developed respiratory failure requiring mechanical ventilation, initially by orotracheal intubation and then after by tracheotomy. This can be explained by the high level of the spinal lesion going back to cervico-bulbar junction. Finally, she also needs a long-term bladder catheterization. Due to mechanical ventilation, she developed multiple respiratory infection, which prolonged her stay in ICU. She was finally transferred to a rehabilitation hospital on day 51.

Upon discharge from the hospital, her neurological examination was slightly better. Her upper extremity function improved lightly distally, while only the left lower leg regained a bit of motor function. Manual Muscle Testing revealed (MMT right/left side) biceps brachii 0/0, triceps brachii 0/0, wrist flexion 2/2, wrist extension 2/2, finger flexion 2/2, finger extension 2/2, hip flexion 0/2, knee extension 0/2, ankle flexion 0/2, and toe flexion 0/2. But the most important improvement was the recovery of effective spontaneous breathing.

This clinical evolution can be correlated with the MRI's evolution performed on day 45, which showed regression of the lesion from cervico-bulbar junction to C3 and from T1 to T6, and the appearance of sequelae from C3 to T1.

Discussion

We related a case of cervicothoracic infarction in a young patient with no etiology found. As described in the recent literature, the idiopathic cause is the main one described. More precisely in young patient, SCI is generally caused by vertebral fracture, cardiovascular malformation (aortic aneurism/dissection) or injury, infection myelitis, fibrocartilagenous embolism [10] and thrombotic disorder such as protein S deficiency [7], prothrombin variant [8], primary anti phospholipid syndrome [9]. After the idiopathic etiology, it is important to remember that the second cause of SCI is iatrogenic secondary to aortic surgery or interventional radiology procedure. In our case, all those etiologies were excluded by imagery and study of the coagulation.

Magnetic resonance imaging is recognized as the most appropriate examination to detect spinal cord infarction. But the limitations of MRI imaging must also be taken in account. Ischemic lesion of the anterior spinal cord is characterized on MRI by « pencil-like » hyper intensities on T2 sagittal and « owl eye » or « snake eye » appearance on T2 axial [4]. Diffusion weighted imaging can show some restriction in the early stage, so it is recommended in all cases of ASA syndrome. However, the sensitivity of initial medullar MRI is limited, with 17-45% remaining normal even with DWI in the early stage. The shortest time reported in the literature between the onset of clinical symptoms and abnormalities on DWI is 3hours [11,12], compared to the brain where DWI restriction is visible within 20 to 30minutes. From one study to another, the proportions concerning the location of the infarct vary. In 55 to 65% of cases, the thoracic level is the most affected followed by the cervical level, then by the lumbar level.

Thereby, although MRI is essential in the management of SCI, it should not be used as exclusion criteria. Mainly, MRI rules out other etiology in ASA syndrome such as compressive myelopathy, vascular malformation, myelitis, demyelinating disorders, and tumor [4].

Another critical point about the care of SCI is the lack of consensus concerning the treatment, except the essential remedy to rehabilitation. Naik, et al. studied the different treatment strategies in the acute phase of SCI [5]. Only CSF drainage ET blood pressure increase (mean blood pressure above 90mmHg) improve outcome, while using antiplatelet and anticoagulation therapy tend to improvement outcomes. No significant improvement or even a worsening was observed with steroids [5].

Finally, some large studies are now available to clarify the prognosis in the long-term outcome and identify predictive indicators. Surprisingly, there is chance of recovery in patients with SCI.

The predictive factors are the ASIA score at onset of symptoms, the extent of lesions and their location, while age, sex and co-morbidities are not correlated with outcome. Patients with ASIA-A and B-score had poorer outcome, comparable to those with severe extensive ischemic injury, for example at the lower thoracic and lumbar levels. However, a functional recovery can be hoped. As noticed by Robertson et al. [3] without established correlation with ASIA-score, the need of wheelchair can decrease for 81% to 42% after 6 months of recovery, while 26% of patients use a gait aid and 33% walk unassisted. The need of bladder catheterization remains high (45%) in the long-term. Median survival is estimated at 6years. Estimated survival of 55% at 5years and 42% at 10years. Considering this data, clinicians should emphasize the possibility of functional recovery.

Conclusion

Although it is a rare entity, physician when faced with a sudden loss of muscle strength in the limbs even in young adult without predisposing factor must consider the diagnosis of SCI. The limitation of early MRI imaging must be taken in consideration. A recent study [5] suggests, as effective acute treatment, CSF drainage and elevated mean arterial pressure (above 90mmHg). Antiplatelet and anticoagulation need larger studies to prove their efficiency in recovery, while using steroid should be avoided. The rehabilitation remains the cornerstone of treatment. Long-term results remain poor in most of patients. However, unexpected findings [3] reveal that functional recovery may be possible, providing some hope.

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Conflict of interest: No author has any conflict of interest in writing the manuscript.

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