Sudden thoracic pain and paraplegia: a case of spinal cord infarction

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Spinal cord infarction (SCI) is exceptionally uncommon, accounting for 0.3–1% of all strokes [1]. The relationship between SCI and vascular surgical interventions, like aortic aneurism surgery, is well-established in adults [2], however spontaneous spinal cord strokes are extremely rare, especially in pediatric patients. Fibrocartilaginous embolism (FCE) has been reported to represent 5.5% of spinal cord infarctions and is under-diagnosed due to its vague clinical presentation [3]. The spinal cord receives its vascular support from two arteries: anterior and posterior spinal arteries (ASA and PSA), and ASA is the most affected (~95%). Symptoms can vary from minor weakness to paraparesis with urinary retention and back pain is the most common symptom.

Case report. A previously-healthy acrobatic gymnastic 12-year-old girl presented to the emergency room for a sudden thoracic back pain that woke her up at 6 a.m. When she tried to stand up, she realized she also had pain in the right leg, lower extremities weakness and was unable to walk. She had her usual acrobatic gymnastics training the day before, where she performed various jumps with knee landing. Other symptoms were denied, such as fever, infectious symptoms, urinary retention or numbness. No significant medical or family histories were found.

On admission, her vital signs were normal for her age and she was afebrile. On neurological examination, normal mental status and cranial nerves. The upper extremities had 5/5 strength and no sensitive alterations. She demonstrated grade 4 muscle strength of the lower limbs and lowering of both legs in the Mingazzini test. Hypoesthesia of the left lower limb was found. There were no proprioceptive alterations. Knee reflexes were present bilaterally with grade 2 response but we could not get any response bilaterally on the ankle reflexes. Babinski sign was absent. She could not walk without assistance. The remaining neurological examination was unremarkable.

Bloods tests including complete blood count, serum chemistry, coagulation and the remaining prothrombic study (protein C and S levels, lupus anticoagulant, anticardiolipin and Beta-2 glycoprotein antibodies and antithrombin III) were normal. A c.667C>T heterozygotic variant on the methylenetetrahydrofolate reductase (MTHFR) and a heterozygotic variant 4G in position 675 on the plasminogen activator inhibitor 1 (PAI-1) gene were found, but the homocysteine level was normal. The spinal column and chest x-rays were normal. The thoracic and lumbar computed tomography (CT) were unremarkable. No cerebrospinal fluid (CSF) analysis was done. She was admitted for further investigation and an urgent thoracic and lumbar magnetic resonance imaging (MRI) was ordered, that showed a prominent high T_2 signal resulting in pencil-like hyperintensity extending between T2-3 and T6-7 levels on the sagittal plane, and two bright dots, on T_2 weighted images on the axial plane (the so called 'owl eye appearance'), corresponding to the involvement of the anterior spinal artery territory. (Fig. 1) The cerebral-MRI was normal, as well as the remaining spinal cord imaging. The most likely diagnosis was an ischemic SCI. A thoracic echocardiogram revealed no evidence of cardiac emboli or congenital cardiomyopathy.

Treatment consisted of supportive care, including monitoring the patient's respiratory, cardiovascular, hydration and neurological status and she was started on aspirin and physical rehabilitation.

By day six she was able to stand on her own and could gait without support a week later. She maintained sensitive alterations, mainly thermic. The remaining hospitalization was uneventful and she was discharged by day 14 to a rehabilitation facility as an impatient to continue intensive treatment, where she continued treatment for one month.

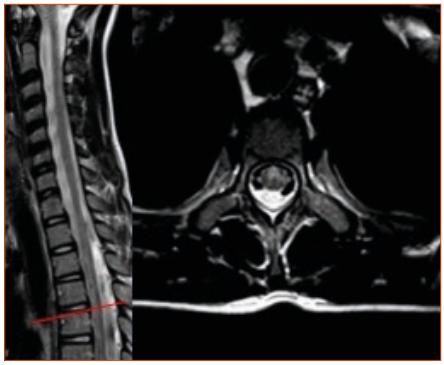

Figura 1. Lumbar-MRI showing prominent high T_2 signal resulting in pencil-like hyperintensity extending between T2-3 and T6-7 levels on the sagittal plane, and two bright dots, on T_2 weighted images on the axial plane (the so called 'owl eye appearance'), corresponding to the involvement of the anterior spinal artery territory.

On her 6-month follow-up she denied feeling any pain and could walk without support. She complained about sensitive thermic alterations on both lower limbs, worse on the proximal left leg (she was unable to distinguish cold from hot water). No other neurological deficits were found on physical examination. She still maintained aspirin. A thoracic-MRI six months after the injury showed evolution to chronic spinal cord infarction with T_2-weight hyperintense signal from T2-3 to T6-7, with no significant spinal atrophy and no new lesions (Fig. 2).

One year later, she has a normal life, with slight sequelae, persisting only hypoesthesia of the lower left limb.

Spinal cord infarction is extremely rare in children and has a different pathogenesis than in adults [4]. Fibrocartilaginous embolism is a type of ischemic myelopathy and was first described in 1961 by Naiman in a 15-year-old boy who died of multiple fatal microscopic arterial emboli of the nucleus pulposus involving the spinal cord and brain steam, suddenly developing quadriplegia, after a trivial fall on his back during a basketball game [5].

The spinal cord receives its vascular support from one anterior spinal artery (ASA) and two posterior spinal arteries (PSA), that only supply the posterior one-third region of the spinal cord. There is a high level of collateral circulation thus decreasing the spinal cord susceptibility to vascular injury. The lower thoracic spinal cord has a lesser degree of collateral circulation and is less vascularized, therefore is at a higher risk of infarction [6]. The pathophysiology of

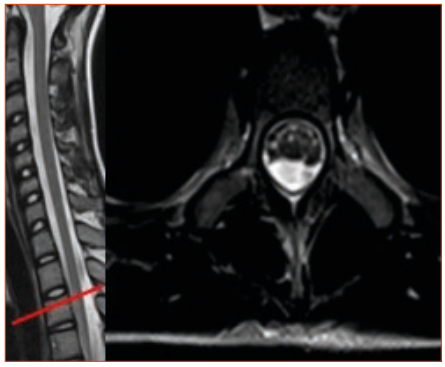

Figura 2. Lumbar-MRI showing evolution to chronic spinal cord infarction with T_2-weight hyperintense signal from T2-3 to T6-7, with no significant spinal atrophy and no new lesions.

FCE remains unclear. It is though that forceful herniation of the intervertebral disk nucleus pulposus material into the intradiscal or vertebral body vessels secondary to minor trauma or some axial loading forces induces the prolapse of the cartilaginous material into the spinal artery [7]. Both sexes appear to be affected equally [8], although some studies show a female predominance [3,7,8].

The presenting and evolving signs and symptoms will depend on the location of the infarction [9]. Fifty percent of patients with SCI reaches maximum symptoms within 12 hours and within 72h for virtually every patient [10]. The most common presentation of FCE is acute back pain on the level of the lesion (70 to 77%), rapid progression of neurologic deficits (71%) and physical examination consistent with vascular-distribution myelopathy (75%), such as dissociative anesthesia and weakness of upper or lower limbs [8,10]. Other symptoms can be present, like respiratory distress and decreased forced vital capacity, urinary and bowel incontinence, loss of deep tendon reflexes and loss of anal tone [9]. The typical ASA infarction pattern is bilateral weakness, dissociative sensory loss with or without autonomic symptoms, because of the involvement of the anterior two-thirds of the cord involving gray and white matter [10]. A pathognomonic finding for ASA infarction is the sparing of proprioception and vibratory sensation below the sensory level. PSA infarction is typically unilateral and less severe, for the presence of two posterior arteries [10].

If SCI if suspected, MRI is essential for diagnosis and should include various sequences [10]. Typically, MRI shows a T_2 hyperintense lesion in a vascular distribution that does not enhance with gadolinium [3]. It also can exclude other causes of acute spinal syndrome, such as compressive myelopathies, vascular malformations, myelitis and tumors. The imaging can be normal at the beginning, but if the clinical suspicion is high, the MRI should be repeated within the next two to seven days [9]. Laboratory studies for a general evaluation, infection markers and cerebral spinal fluid (CSF) have no specific markers for FCE and are usually normal.

A definitive diagnosis can only be made by autopsy, but clinical and radiological features can provide a very likely diagnosis of FCE [7]. Abdel-Razek at al proposed a schematic approach to diagnosing FCE that involves establishing the presence of myelopathy based on neurological examination; excluding traumatic and compressive etiologies of myelopathy by history and imaging; normal CSF studies and no gadolinium enhancement in MRI thereby excluding inflammatory etiologies; the presence of one major criterion (clear vascular distribution by physical exam or imaging modalities or MRI T_2 hyperintensity) or two minor criteria (accompanying new onset neck or back pain, symptom progression within 4-8 hours or initial unremarkable MRI); and that no other cause of SCI has been found [3].

No consensus guidelines exist for FCE. Blood pressure control, supportive care and rehabilitation therapy are important in the long-term management [7]. Corticosteroids, anticoagulant therapy and immunoglobulin have been used in pediatric patients, without any evidence to support them [7,9]. The only treatment that has proven to be successful in achieving good outcomes is rehabilitation therapy [6].

The prognosis depends mainly on the location and extension of the lesion, varying from poor to significant neurological improvement. Severe neurological deficits at onset and involvement of proprioception are associated with poorer outcomes [10]. Our patient was a success case, perceived she has almost no neurological deficits, which is rare in FCE patients, to our knowledge [3,10].

In conclusion, SCI is rare in children and in patients with characteristic clinical and radiological findings, such as our patient, FCE should be a possible diagnosis since a good functional outcome is attainable with prompt recognition and adequate management. An MRI must be performed, and the T_2 hyperintense lesion will confirm the diagnosis of SCI.

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