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Original article

Childhood idiopathic spinal cord infarction: Description of 7 cases and review of the literature

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Abstract

Objectives: To describe the clinical course, neuroimaging findings and functional outcome of idiopathic spinal cord infarction (SCI) in adolescents.

Methods: Retrospective and descriptive analyses of seven patients with idiopathic SCI and 50 additional cases from the literature were included. Data collected concerned clinical presentation, MRI findings, initial diagnosis, treatments and functional outcome at the last medical visit.

Results: Mean age at presentation was 13.2 years (range 13–15). All patients presented a sudden and painful acute myelopathy with <24 h time to maximal symptoms manifestation. A suspected trigger related to a minor effort was reported in 3/7 cases. Six patients presented with paraplegia, one with paraparesis. All had bladder dysfunction needing catheterization. Three patients had an initial misdiagnosis. Initial MRI was considered as normal in 2 cases. In the 5 other cases, T2-weighted-MR images showed hyperintensity within the thoracolumbar spinal cord, affecting mostly the anterior spinal artery territory. Evidence for associated spinal growth dystrophy were present in 6/7 cases. Mean follow-up time was 27.4 months (range 3–46): 2 patients recovered autonomous ambulation, 4 patients regained walking ability with aids and one child (the shortest follow-up) remained wheelchair-dependent. A neurogenic bladder was still reported in 6/7 children at the last visit. Complementary analyses with literature cases were consistent with the findings obtained in our cohort.

Conclusion: Idiopathic SCI typically occurs in adolescence with a rapid onset and painful acute myelopathy. The MRI shows a T2-hyperintense signal within the spinal cord and provides evidence for an ischemic mechanism. Etiology remains unclear in most cases even though some specific risk factors for this age must play an important role in the pathogenesis, such as mechanical constraints on the immature spine.

Keywords: Spinal cord infarction; Ischemic stroke; Anterior spinal artery syndrome; Idiopathic; Magnetic resonance imaging (MRI); Spinal growth dystrophy; Child

1. Introduction

Spinal cord infarction (SCI) is a dramatic acute event, associated with long-term sequelae impacting a patient's quality of life over the long term [1,2]. No epidemiologic data are available for childhood SCI even

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though it is considered to be much less frequent than a cerebral ischemic stroke. Due to its rarity, knowledge of the natural history of childhood SCI is mostly based on reports of pediatric cases or studies that mix children and adults.

Blood supply to the spinal cord is mostly provided by one anterior and two posterior spinal arteries. The anterior spinal artery (ASA) supplies about two thirds of the cross-sectional area of the spinal cord, including the anterior and lateral corticospinal tracts, the spinothalamic tracts and the anterior horn cells [3]. Therefore, clinical manifestations of SCI depend upon the vascular territory involved. The anterior territory is more vulnerable than the posterior region, which contains many anastomoses. Anterior spinal artery syndrome (ASAS) is an acute myelopathy with sudden back pain followed by the appearance of a rapid bilateral motor deficit and sensory impairment, as well as bladder and/or bowel dysfunction. Typically, temperature and pain sensation are altered, whereas vibration, light touch, and proprioceptive sensations are preserved (due to sparing of the lemniscus pathway) [4]. Magnetic resonance imaging (MRI) usually confirms diagnosis with hyperintense signal on T2-weighted images restricted to a vascular territory [5].

As for cerebrovascular ischemic stroke, the pathogenesis of SCI is different between children and adults. Apart from traumatic causes and common etiologies (including aortic surgery, systemic hypotension, parainfectious vasculopathies, cardioembolic disease, tumor embolism or radiotherapy), it can also occur in an otherwise healthy child without any explanation despite extensive investigation [1,6]. A minor and unnoticed trauma is sometimes reported and thought to constitute as a risk factor [1,7–10], just as the presence of an isolated thrombotic risk factor [11–15].

No study has focused on these idiopathic cases of SCI so that the clinical course, MRI findings and the pathogenesis remain largely unknown. In this study, we described 7 cases of idiopathic SCI in children. We also carried out an exhaustive review of the literature that permitted, along with our 7 cases, to analyse 57 cases of acute idiopathic SCI.

As it may be misdiagnosis with other causes of acute myelopathy, and especially acute transverse myelitis, the purpose of this retrospective study was to point out typical features and specificities of the clinical presentation and MRI findings of SCI. We also studied the follow-up data of these children and discussed the potential underlying pathogenesis.

2. Patients and methods

2.1. Identification of patient study

Patient cases were retrospectively identified by survey among the clinicians of pediatric neurology departments of four different French university hospitals, representing tertiary reference centers (Bordeaux, Toulouse, Tours, Montpellier). Inclusion criteria were: (1) Clinical diagnosis of acute transverse myelopathy, total or partial, including sudden and rapid-onset (<72 h) of sensory, motor, or autonomic dysfunction attributable to the spinal cord [4], (2) MRI evidence for ischemic lesions on imaging (vascular territory, extension of the lesion, restricted diffusion), (3) Age one month to 18 years. Patients were excluded from the study if the SCI was related to a specific etiology known to possibly induce systemic hypotension (aortic diseases, any kind of surgery, arteriovenous malformation) or any other obvious cause like inflammatory diseases (infectious, systemic) or radiotherapy. Thrombotic disorders were considered as a risk factor, not as a direct cause, and were thus included in the study.

2.2. Data collection

The local neuro pediatricians were asked to retrieve all cases of SCI by data base search. For each patient included, we collected information from medical charts regarding past history, suspected trigger, clinical manifestation at presentation, initial diagnosis, imaging and other diagnostic investigations, treatment and functional outcome. As a rapid progression of symptoms (<4 h) argues in favor of ischemia, the delay between onset of deficit and maximal severity was also collected [4]. Functional status was evaluated from the clinical report at the last visit taking into account the level of motor improvement (complete, partial, none), ambulation/locomotion (independent, with aids, or wheelchair), sensory impairment (persistent or not) and bladder disorder (need for catheterization or not). All reports of MRI scans were collected and images were all reviewed by the same radiologist (PB). We tabulated the timing of the scan from the onset of deficits, signal changes on axial and sagittal T2-weighted sequences and diffusion-weighted imaging, the precise location of the lesion, the number of spinal levels involved, the presence of enhancement after chelate of gadolinium administration and associated spine modifications. Premature disc degeneration was detected as disk space narrowing with decrease of signal intensity within the nucleus pulposus, related to an altered disk hydration [16]. Schmorl's node corresponds to a central herniation of the nucleus pulposus throughout the cartilaginous and bony end plate into the body of the adjacent vertebra [16]. This study has been approved by the local Ethical Committee. Informed consent from all patients and/or their legal representatives has been obtained.

2.3. Literature review

The Online database Pubmed.com was used to search additional cases by combining the terms "spinal cord