Pediatric Neurology 99 (2019) 3–6

Contents lists available at ScienceDirect

Pediatric Neurology

journal homepage: www.elsevier.com/locate/pnu

Topical Review

Fibrocartilaginous Embolism of the Spinal Cord in Children: A Case Report and Review of Literature

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ARTICLE INFO

Article history:
Received 12 February 2019
Accepted 20 April 2019
Available online 26 April 2019

Keywords:
Clinically isolated syndrome
Infarction
Urinary retention
Transverse myelitis
Myelopathy
Plegia
Spinal cord infarction
Ischemic myelopathy

ABSTRACT

Fibrocartilaginous embolism is assumed to be caused by fibrocartilaginous nucleus pulposus component migration through retrograde embolization to the spinal cord artery. Fibrocartilaginous embolism is currently not well recognized among pediatricians because of its rarity. We describe a previously healthy 15-year-old soccer player who, after kicking a ball, developed progressive weakness in both legs and urinary retention the next day. Magnetic resonance imaging revealed T2 hyperintensity in the anterior horn of the spinal cord at the Th12/L1 level with Schmorl node at the level of L1/2. We also review the previous literature on fibrocartilaginous embolism of the spinal cord in children (less than18 years age); a total of 25 pediatric patients, including our patient, were identified. The median age was 14 years, and 64% of the reviewed patients were female. The most common trigger event was intense exercise or sports. The neurological symptoms started within one day in most cases, and the time to symptom peak varied from a few hours to two weeks. The most common initial neurological symptoms were weakness or plegia (100%), followed by paresthesia or numbness (48%). Affected areas of the spinal cord were distributed evenly from the cervical to thoracolumbar regions. Although steroids and anticoagulants were most commonly used, the prognosis was quite poor (mild to severe sequelae with three deaths). Although fibrocartilaginous embolism is a very rare condition, physicians should be aware of the characteristics and include fibrocartilaginous embolism of the spinal cord in their differential diagnosis, especially for physically active patients.

Introduction

Fibrocartilaginous embolism (FCE) of the spinal cord is a rare and underrecognized condition. FCE is characterized by ischemic myelopathy, which is assumed to be caused by the migration of fibrocartilaginous nucleus pulposus components through retrograde embolization to the spinal artery.[1] Symptoms include rapidly progressive plegia, paresthesia, and bladder or bowel dysfunction typically following back or neck pain.[2]

FCE is well described in the veterinary literature, and it has been increasingly recognized in humans.[3-7] However, it is currently not well recognized by pediatricians because of its rarity. Recognizing the clinical features that distinguish FCE from other causes of spinal cord disorders will be helpful in the proper management of these patients.

We describe a 15-year-old soccer player with ischemic myelopathy secondary to suspected FCE based on clinical and radiological findings. In addition, we also reviewed previous reports of FCE of the spinal cord in children younger than 18 years to elucidate the characteristics in pediatric patients. A comprehensive review of the literature includes epidemiology, clinical course, diagnosis, treatment, and prognosis of FCE among pediatric patients.

Conflict of interests: None.

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https://doi.org/10.1016/j.pediatrneurol.2019.04.013
0887-8994/

Patient description

This previously healthy 15-year-old soccer player presented with weakness of both legs and urinary retention. He was a junior-high-school soccer player who practiced for more than two hours four times a week. Two days before admission, he felt progressive heaviness on his right leg without back pain after kicking a soccer ball. A day before admission, in addition to right lower extremity weakness, left extremity weakness developed and he started to feel tingling pain around both calves. Furthermore, he developed urinary retention on the same day. He denied any trauma, infectious symptoms, or fever. He had no significant medical or family histories.

On admission, he was afebrile and his other vital signs were normal. On neurological examination, his mental status and cranial nerves were normal. He had no tenderness over his back, and he had 5/5 strength in his upper extremities. Although he described weakness of both lower extremities, he only demonstrated mild distal right leg weakness (ankle dorsiflexion and plantar flexion, 4/5). The right Achilles reflex was absent, but all other reflexes were intact; there was no Babinski sign. Sensory and rectal sphincter examinations were unremarkable, but he did have urinary retention. He was able to walk without assistance. However, his gait was unsteady; he could not walk on tiptoe.

Magnetic resonance imaging (MRI) studies of the brain and cervical and thoracic spinal cord were unremarkable. However, a lumbar MRI showed a hyperintense lesion on T2-weighted images (T2WI) in the anterior horn of the spinal cord at the Th12/L1 level without enhancement and a disk collapse with Schmorl node at the L1/2 level, resulting in a vertical protrusion of the intervertebral disk cartilage into the adjacent upper and lower vertebral bodies (Fig). Blood tests including complete blood count, serum chemistry, coagulation (protein C and S levels, prothrombin, activated partial thromboplastin time), lupus anticoagulant, anticardiolipin antibodies, anti-aquaporin 4, and myelin-oligodendrocyte glycoprotein antibody revealed no abnormalities.

Cerebrospinal fluid (CSF) analysis was also normal except for slightly elevated protein (31 mg/dL [normal range for lab: 8.8 to 27.1 mg/dL]) and myelin basic protein (237 pg/mL [normal range: ≤102]). The nerve conduction velocity test was normal.

Our differential diagnosis on admission included transverse myelitis and clinically isolated syndrome, but the MRI findings were atypical. After a Foley catheter was inserted, he was given high-dose steroid (1 g/day) for two days, which was discontinued due to elevated blood sugar. The steroid was tapered to 1 mg/kg for two days and then to 0.5 mg/kg for two days. Although his right leg weakness did not completely improve with steroid pulse, urinary retention completely disappeared and the Foley catheter was removed eight days after admission. A repeat MRI on the same day showed a new finding of enhanced lesion at T12/L1, and the previous high-intensity T2WI at T12/L1 was reduced (Fig). He was discharged nine days after admission. Although the strength of his right leg remained at 4/5 at discharge, three weeks later his right muscle strength returned to 5/5, his gait was steady, and he could walk on tiptoe. Although his likely diagnosis on admission was transverse myelitis or clinically isolated syndrome, a literature review led to the consideration of FCE of the spinal cord as the most likely final diagnosis.

Method

Systematic review of the literature

The English medical literature was reviewed for cases reported as FCE of the spinal cord in pediatric patients. The PubMed database, up to January 29, 2019, was searched using the following search terms: “fibrocartilaginous emboli” or “fibrocartilagenous emboli” or “nucleus pulposus emboli.” A total of 123 articles were found; the abstracts or manuscripts of these articles were considered carefully, resulting in 18 case reports, one letter to the editor, and one case report and review of the literature of interest. After studying these articles carefully, we identified 20 articles that described pediatric patients with FCE of the spinal cord, and a total of 25 children younger than 18 years, including our patient, were reviewed.[1-20]

Results

Characteristics of patients with FCE from the literature

The characteristics of patients with FCE from the literature and our case are shown in the Supplementary Table. Only three cases of FCE were pathologically defined, and FCE was suspected in the remaining 22 cases clinically by presentation, neurological findings, and MRI findings. The median age of the patients at presentation was 14 years (interquartile range: 12 to 16 years). Of the 25 patients, there were more female patients than male (16 female versus nine male). Half of the cases were associated with heavy exercise. The neurological symptoms started within two hours in half of the cases and within one day in most cases. The time to symptom peak ranged from a few hours to a few weeks. Pain was observed in most cases, and back pain was the most common (n = 17). All patients showed weakness or plegia of the extremities as initial neurological symptoms, and half of the patients showed paresthesia or bladder or bowel dysfunction. In seven cases, the first MRI was normal, but repeat MRI conducted several days later revealed T2WI hyperintensity in the spinal cord in all cases. The affected spinal segments, as determined by MRI, were the cervical (n = 2), cervicothoracic (n = 6), thoracic (n = 4), thoracolumbar (n = 7), and lumbar (n = 1) vertebra. Abnormal vertebral body or disk changes were reported in 18 cases, and apparent Schmorl node was reported in six cases. CSF results in half of the cases were unremarkable. Elevated myelin basic protein, protein, and leukocytosis in CSF were observed in three, five, and two patients, respectively. Intravenous steroids (n = 14) and anticoagulants (n = 7) were the usual treatments, and nine patients were not given any medication. As for the prognosis, only one patient fully recovered, most patients had mild to severe sequelae (n = 21), and three patients died.

Discussion

FCE is a very rare cause of acute ischemic myelopathy and is not often suspected on presentation. We identified only 25 cases of FCE of the spinal cord in pediatric patients younger than 18 years in the available English literature from 1961.[1-20] In our patient, the diagnosis was delayed because the disorder is rare and underrecognized.

FCE shows a bimodal distribution, with peaks in adolescence and late middle age.[1] Sex differences differ depending on the reports (male predominance[21] and female predominance[19,22,23]). Our review showed a female predominance. Although vertebral body damage by osteoporosis or cartilage degeneration, often found in postmenopausal women, is a possible risk factor for the occurrence of FCE of the spinal cord, it is rare in female children. Therefore the reason for female predominance of FCE is unclear.

The pathophysiology of FCE remains unclear. The most accepted hypothesis is 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 cartilaginous material into the spinal

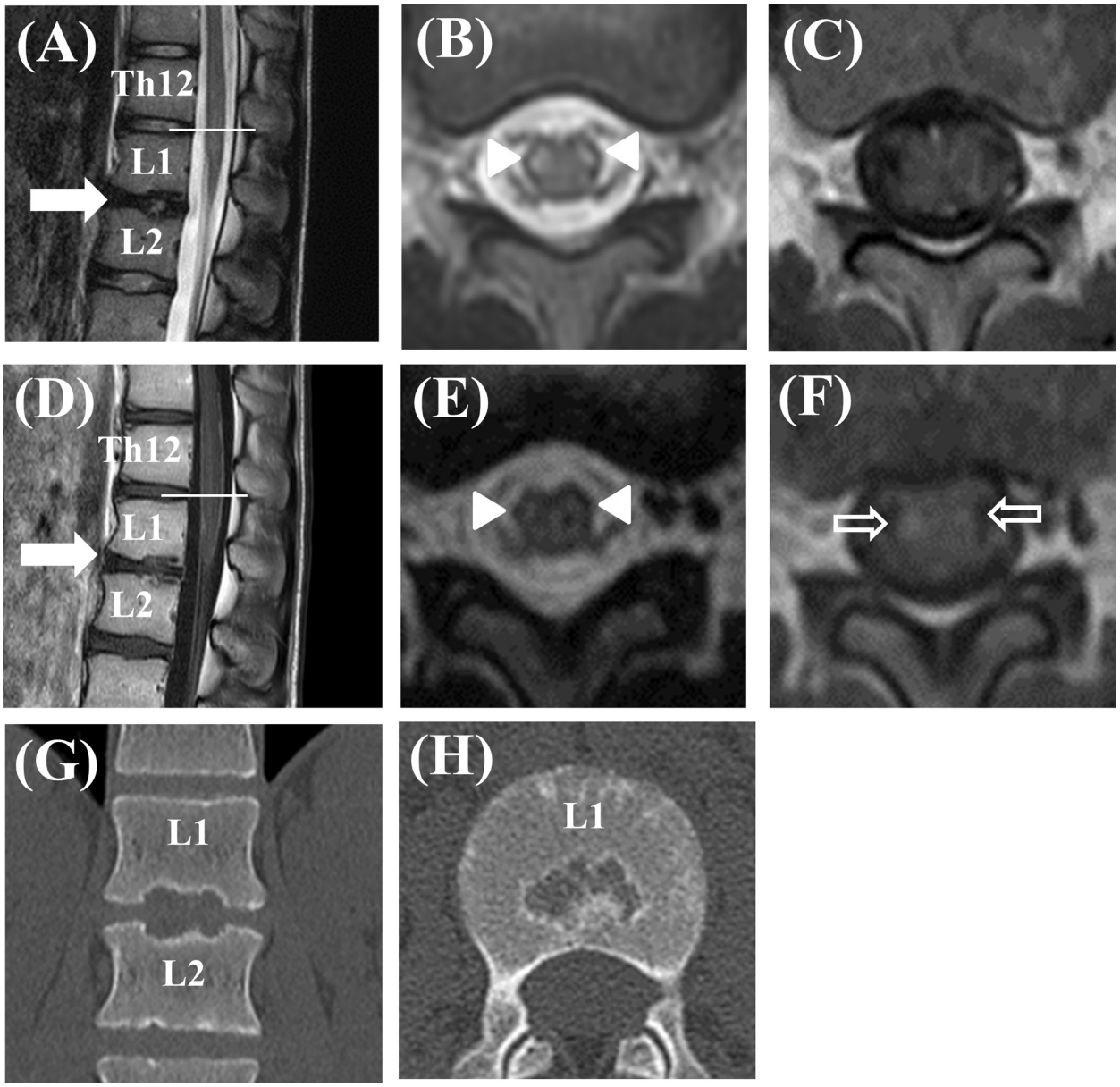

FIGURE. Sagittal (A), axial T2-weighted (B), and contrast-enhanced (C) images of the lumbar spinal cord on day 1. The sagittal and contrast-enhanced images of Th12/L1 showed no abnormalities. The axial T2-weighted image showed an increased signal bilaterally at the Th12/L1 level in the anterior horn of the spinal cord (B) (arrowhead). The sagittal image revealed no abnormality on day 13 (D). However, a high-intensity area of the axial T2-weighted image at Th12/L1 appeared diminished (E) (arrowhead), and new findings were noted on a contrast-enhanced image at Th12/L1 (F) (the right side is more strongly enhanced than the left side; empty white arrow). In addition, L1/L2 disk collapse and Schmorl node (white arrow) (A and D) were observed, along with adjacent vertebral end-plate destruction on computed tomography (G and H).

artery.[1] The nucleus pulposus may also extend into the vertebral body by Schmorl node[2,23] or probably abnormal vertebral body or disk changes.[9-12,15-17,19] Schmorl nodes, focal masses of fibrocartilage within the vertebral body, are commonly found in adults but are rare among children.[24,25] However, our review showed that six patients (24%) had an apparent Schmorl node. In addition, abnormal vertebral body or disk changes were reported in 18 cases (72%). Therefore these abnormalities will also be helpful to diagnose FCE of the spinal cord in children.

Although a definitive diagnosis of FCE can only be made after an autopsy, diagnosis by clinical and radiological features is considered possible.[1-18] Patients with FCE typically present with weakness that progresses to paralysis over a period of minutes to hours, often with sudden back pain, sensory problems, or bladder dysfunction after a suggestive history of a minor traumatic event.[21-23] The characteristic finding of difference between spinal cord infarction and inflammatory cord disease is a rapid course of symptoms to equilibrium state, mostly over hours.[23] Our patient developed sudden progressive right lower extremity weakness after kicking a ball. He then felt tingling pain around both calves. Although the clinical course usually involves back or neck pain at onset,[22] some patients, including our own, do not experience such pain.[9,10,15] Minor injuries and Valsalva maneuver have been often reported as the trigger of FCE.[21-23] Similarly, the trigger event in our review showed that only 24% had an apparent traumatic event and half of the patients had completed intense exercise. Therefore progressive weakness, paresthesia, and bladder or bowel dysfunction after these events are suggestive of FCE of the spinal cord.

The MRI findings of expansion of the spinal cord with increased signal on T2WI involving the spinal cord without early contrast enhancement in association with a narrowed disk or Schmorl nodes strongly suggest the diagnosis of FCE.[1,2,23] Diffusion-weighted imaging sequence on MRI is also reported as a good modality because abnormalities can be found even within a few hours in patients with spinal cord ischemia.[12] However, MRI sometimes reveals normal results in the early stage of FCE.[2–4,13] Therefore, repeat MRI with a T2WI sequence or an enhanced MRI may be needed later. The commonly affected sites are the cervical[1,22,23] or thoracic regions.[21,23,26] However, our review in children showed that the affected areas were distributed evenly from the cervicothoracic to the thoracolumbar regions.

There is no specific treatment for FCE of the spinal cord. Emergency surgery, corticosteroids, and anticoagulant therapy have all been utilized acutely. Blood pressure control, supportive care, and rehabilitative physical therapy are important in the long-term management.[21-23] Significant clinical improvement after treatment is often not evident, so the effect of these treatments are unknown. However, our review showed that one patient who completely recovered was treated with cortico steroids and three patients who died were not treated with medicine. Further research will be necessary. The prognosis of FCE of the spinal cord varies from poor to significant neurological improvement, depending on the involved area in the spinal cord and the extent of spinal cord ischemia.[21-23] Involvement of the cervical spinal cord often has an unfavorable prognosis because of respiratory failure.[1,26] Although the degree of recovery of spinal cord ischemia is generally poor, our patient regained relatively good neurological function, which may be because the spinal cord infarction involved the the lumbosacral area and the extent of spinal cord ischemia was small compared with that of previous reports.[2,5,23]

There are limitations to our report. As FCE of the spinal cord can be definitively diagnosed only by autopsy, some previously reported cases of spinal cord infarction might not have been caused by FCE. In addition, as we only included articles in English, we might have missed some important articles written in other languages.

In conclusion, the clinical course of our patient was characteristic, marked by a progressive neurological deficit after physiological effort and increased T2WI signal intensity in MRI without early contrast enhancement; the lesion was found in the anterior cord and was asymmetric (more on the right), corresponding with his right leg weakness, and the presence of Schmorl node was highly suggestive of FCE.

Acknowledgments

We thank Drs. Norio Chihara, Division of Neurology, Kobe University Graduate School of Medicine, and Tomonori Kanda, Division of Radiology, Kobe University Graduate School of Medicine, for their advice regarding the patient's MRI findings and diagnosis. We also thank Dr. Toshiyuki Takahashi, Department of Multiple Sclerosis Therapeutics, Tohoku University Graduate School of Medicine, for measuring myelin oligodendrocyte glycoprotein. This study was supported by a Grants-in-Aid for Scientific Research (KAKENHI) from the Ministry of Education, Culture, Sports, Science and Technology of Japan (subject ID: 18K089181 8 to Hiroaki Nagase).

Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.pediatrneurol.2019.04.013.

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