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Review article

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

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ABSTRACT

Fibrocartilaginous embolism (FCE) is assumed to be caused by the migration of fibrocartilaginous nucleus pulposus components through retrograde embolization to the spinal cord artery. FCE is currently not well recognized among pediatricians due to its rarity. We present the case of a previously healthy 15-year-old soccer player who, after kicking a ball, developed progressive weakness in both legs and ischuria the next day. Magnetic resonance imaging revealed T2 hyperintensity in the anterior horn of the spinal cord at the Th12/L1 level with Schmorl's node at the level of L1/2. We also reviewed previous literature of FCE of the spinal cord in children (<18 years of age), and a total of 25 pediatric patients, including our case, were reviewed. The median age was 14 years and 64% of the reviewed patients were female. The most common trigger event was intense exercise or sports (52%). The neurological symptoms started within 1 day in most cases and time to symptom peak varied from a few hours to 2 weeks. The most common initial neurologic symptoms were weakness or plegia (100%), followed by paresthesia or numbness (44%). 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 (mostly mild to severe sequelae and 3 patients died). Although FCE is a very rare condition, pediatricians should be aware of the characteristics and include FCE of the spinal cord in their differential diagnosis, especially for physically active patients.

Keywords: clinically isolated syndrome; infarction; ischuria; transverse myelitis; multiple sclerosis; myelopathy; plegia

Introduction

Fibrocartilaginous embolism (FCE) of the spinal cord is a rare and underrecognized infarction. It 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 veterinary literature; recently, it has also been increasingly recognized in humans[3-7]. However, it is currently not well recognized among pediatricians due to 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 pediatric patients.

We present a case of 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. Comprehensive review of the literature includes epidemiology, clinical course, diagnosis, treatment, and prognosis of FCE among pediatric patients. We have obtained the patient's and his parents' consent to present this case.

Case Description

A previously healthy 15-year-old soccer player presented with weakness of both lower extremities and ischuria. He was a junior high school soccer player who practiced for more than 2 hours 4 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 ischuria 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 other vital signs were normal. On neurologic 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 and all other reflexes were intact; there was no Babinski sign. Sensory and sphincter examinations were unremarkable but he did have ischuria. 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 disc collapse with Schmorl's node at the L1/2 level, resulting in a vertical protrusion of the intervertebral disc cartilage into the adjacent upper and lower vertebral bodies (Figure). Blood tests revealed no abnormalities and the cerebrospinal fluid (CSF) analyses were also normal except for slightly elevated protein and myelin basic protein (Table 1). The nerve conduction velocity (NCV) test was normal.

Our differential diagnosis on admission included transverse myelitis and clinically isolated syndrome (CIS), but the MRI findings were atypical. After a Foley catheter was inserted, he was given two days of high-dose steroids (1 g/day) that were discontinued due to elevated blood sugar. Steroids was tapered to 1 mg/kg for 2 days and then to 0.5 mg/kg for 2 days. Although his right leg weakness did not completely improve with steroid pulse, ischuria completely disappeared and the Foley catheter was removed 8 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 (Figure). He was discharged 9 days after admission. Although the strength of his right leg remained at 4/5 at discharge, 3 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 CIS, literature review led to the consideration of FCE of the spinal cord as the most likely final diagnosis.

Method

Systematic review of the literature

Literature in English 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 papers were considered carefully, resulting in 18 case reports, 1 letter to the editor, and 1 case report and review of the literature of interest. After studying these articles carefully, we identified 20 literatures that described a case of FCE of the spinal cord in pediatric patients, and a total of 25 children younger than 18 years of age, including our case, were reviewed[1-20].

Result

Characteristics of patients with FCE from the literature

The characteristics of patients with FCE from the literature and our case are shown in Table 2. Twenty-five pediatric patients were reviewed. Only 3 cases were pathologically defined and the remaining 22 cases were suspected clinically by presentation, neurologic, and MRI findings. The median age at presentation was 14 years (interquartile range (IQR): 12–16). Of the 25 patients, there were more female patients than male (16 females vs 9 males). The trigger events included intense exercise or sports (n=13), trauma (n=6), motor vehicle accidents (n=1), riding a roller coaster (n=1), milking a cow (n=1), leaning forward (n=1), and unknown (n=2). The neurologic symptoms started within 2 hours in half of the cases (n=12), and within 1 day in most cases (n=18). The time to symptom peak varied from a few hours to a few weeks. Most cases showed pain (n=21), and back pain was the most common (n=17). Initial neurologic symptoms included weakness or plegia of the extremities (n=25), paresthesia or numbness (n=11), bladder or bowel incontinence (n=7), ischuria (n=4), and ventilatory failure (n=2). Abnormal findings of neurologic examination at presentation commonly included weakness of extremities ranging from complete plegia to mild weakness, diminished or compete loss of sensation, and bladder or bowel function problems. Affected spinal segments of the spine by MRI (n=23) were as follows (cervical (n=5), cervicothoracic (n=5), thoracic (n=6), thoracolumbar (n=5), and lumbar (n=2)). Two autopsy cases reported FCE in the spinal cord arteries from the lower medulla to T7 and in the lower medulla and C2-T7. Abnormal vertebral body or disc changes were reported in 15 cases and apparent Schmorl's node was reported in 6 cases. CSF was analyzed in 22 patients, and 13 were unremarkable. Elevated MBP, protein, and leukocytosis in CSF were observed in 3, 5, and 2 patients, respectively. Treatments were as follows: intravenous steroids (n=14), anticoagulant (n=7), IVIG (n=2), fluoxetine (n=1), plasma exchange (n=1), Vitamin B12 (n=1), cyclophosphamide (n=1), and 9 patients were not treated by any medicine. As for prognosis, only one patient fully recovered. Most patients (n=21) had mild to severe sequelae and 3 patients died.

DISCUSSION

FCE is a very rare cause of acute ischemic neuropathy and is not often suspected on presentation. In our case, the diagnosis was delayed because of the rare and underrecognized disorder. According to our review, only 25 cases of FCE of the spinal cord in pediatric patients younger than 18 years of age have been reported in the available English literature from 1961[1-20.] In addition, only 3 cases were pathologically defined[17,19,20]. It is possible that FCE is more common among children than expected.

Epidemiology of FCE of the spinal cord

According to a previous study, FCE shows bimodal distribution, with peaks in adolescence and late middle age[1]. Some review articles about FCE among adults or adults as well as a few children have been reported and sex differences differ depending on reports (male dominancy[21], and female dominancy[19,22,23]). According to our review, the median age of FCE of spinal cord among children was 14 years (IQR: 12-16) and the youngest was 8 months old. Our article showed female dominancy (64%) among children with FCE of the spinal cord. 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 dominance of FCE is unclear.

Pathophysiology of FCE of the spinal cord

The definitive pathophysiology of FCE remains unclear. However, some mechanisms have been proposed[1,2,23]. The most accepted hypothesis is that forceful herniation of the intervertebral disc 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 artery[1]. The nucleus pulposus may also extend into the vertebral body by Schmorl's node[2,23] or probably abnormal vertebral body or disc changes[9-12,15-17,19]. Schmorl's 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 6 cases (24%) had apparent Schmorl's node. In addition, abnormal vertebral body or disc changes were reported in 15 cases. Therefore, these abnormalities will also be helpful to diagnose FCE of the spinal cord among children. It is well known that the common blood supply to the spinal cord and nucleus pulposus, existing in infancy to childhood, generally becomes avascular after adolescence. The persistence of this vasculature has been postulated to increase the incidence of FCE around adolescence[23]. From our review, the median age of FCE of spinal cord among children was 14 years. Therefore, this result is compatible with the physiology of the vasculature of the spinal cord and nucleus pulposus. In the present case, we hypothesized that our patient developed FCE by minor trauma (kicking a soccer ball), which resulted in increased intervertebral pressure, which, in turn, caused the migration of fibrocartilaginous material (Schmorl's node may be involved) into this vasculature.

Symptoms of FCE of the spinal cord

FCE patients 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 events[21-23]. Although the clinical course usually involves back or neck pain at onset[22], some cases, including our case, did not experience such pain[9,10,15]. The time to symptom peak reported by previous articles including adults and few children varied from a few hours to 48 hours[21-23,26]. 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 soccer ball. He then developed left extremity weakness and felt tingling pain around both calves. Furthermore, he developed urinary incontinence later. Our review also showed that the neurological symptoms occurred within 2 hours from the event in half of the cases, and within 1 day in most cases. The time to symptom peak varied from a few hours to 2 weeks. These results are the same as those of previous articles on adults and a few children[21-23]. This temporal progression, like that in the present case, may be important in differentiating FCE from other mechanisms of arterial embolism[23]. Minor injuries and/or Valsalva maneuver have been often reported as the trigger of FCE[21-23]. Similarly, the trigger event in our review showed that only 6 patients (24%) had an apparent trauma event, and half of the patients (n=13) had intense exercise or sports. Therefore, progressive weakness, paresthesia, or bladder or bowel dysfunction after these events are highly indicative of FCE of the spinal cord.

Diagnosis of FCE of the spinal cord

FCE is often misdiagnosed with transverse myelitis, like in our patient[3]. Although a definitive diagnosis of FCE can only be made after an autopsy, diagnosis by clinical and radiological features is considered possible[1-18]. The number of clinically diagnosed cases has been exceeding the number of autopsy-proven cases, and we found only 3 pediatric cases of FCE of the spinal cord that were pathologically defined[17,19,20]. 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 disc or Schmorl's nodes strongly suggest the diagnosis of FCE[1,2,23]. Schmorl's nodes have been often found in the vertebral bodies in approximately 40% patients with FCE[20,26] but AbdelRazek et al.[23] reported that degenerative disc diseases and Schmorl's node were observed in only 32% and 17% of FCE patients, respectively. DWI of MRI is also reported as good modality because abnormalities can be found even within a few hours in patients with spinal cord ischemia[12]. However, MRI sometimes reveal normal results

in the early stage of FCE[2-4,13]. Therefore, in such a case, repeated MRI of T2WI or enhanced MRI will be needed later. Subacute MRI often shows "pencil-like" T2WI hyperintensity in the spinal cord[21-23]. CSF analysis is usually unremarkable. However, Mateen et al.[21] reported elevated CSF protein in 6 out of 8 patients. In our patient, the clinical course was characteristic, marked by a progressive neurologic deficit after physiological effort, increased T2WI signal intensity in MRI without early contrast enhancement, and the lesion was found in the anterior cord and was asymmetric (more on the right), corresponding with his right weakness, and the presence of Schmorl's node was highly suggestive of FCE. The differential diagnosis for progressive weakness is broad, including transverse myelitis, acute polyneuropathy, early multiple sclerosis, or neuromyelitis optica. Acute transverse myelitis usually involves multiple spinal cord levels on MRI and typically involves at least two-thirds of the cross section of the spinal cord[27], whereas in our patient, only a small portion of the anterior spinal cord was involved. In the present case, the patient had no prodromal illness or trauma, and had a negative thrombotic evaluation and aquaporin-4 immunoglobulin G (IgG) in the blood as well as lymphocytic pleocytosis, elevated protein level or IgG index, oligoclonal bands in the CSF, and a normal NCV test. Therefore, the clinical history in combination with these results and MRI features were highly suggestive of FCE, and the abovementioned differential diagnoses were excluded. Our study also showed that 23 patients were performed MRI and all cases showed T2WI hyperintensity in the spinal cord. Therefore, T2WI of MRI will be helpful tool to diagnose FCE of the spinal cord. The commonly affected sites are reported to be the cervical[1,22,23] or thoracic regions[21,23,26]. However, our review of the literature of FCE in children showed that the affected area was distributed evenly from the cervical to thoracolumbar regions.

Treatment and prognosis of FCE of the spinal cord

There is no specific treatment for FCE of the spinal cord. It varies from emergent surgery to steroid or anticoagulant therapy or blood pressure control, and supportive care and rehabilitative physical therapy are also important in long-term treatment[21-23]. However, there have been no report of significant clinical improvement after treatment. The prognosis of FCE of the spinal cord varies from poor to significant neurologic improvement due to the involved area in the spinal cord and the extent of spinal cord ischemia[21-23]. Involvement of the cervical spinal cord frequently shows an unfavorable progression due to respiratory failure[1,26]. In our case, we started steroid pulse therapy for unknown cause of myelitis and started rehabilitative physical therapy. His lower extremity weakness improved daily, and finally, his ischuria recovered. After 3 weeks

from onset, difficulty in ambulating on tiptoe has not improved. Although the degree of recovery of spinal cord ischemia is generally poor, our patient showed relatively good neurologic improvement, which may because the involved spinal cord area was the lumbosacral part and the extent of spinal cord ischemia was small compared with previous reports[2,5,23]. Our review showed half of the cases were treated by steroids and/or anticoagulant. However, most cases had finally mild to severe sequela. Therefore, the effect of these treatments on FCE are unknown. However, one completely recovered patient was treated by steroid and 3 patients who died were not treated by medicine. Further research will be necessary.

Limitation

Because 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.

Conclusion

In conclusion, although FCE of the spinal cord can only be diagnosed by histopathology, we finally diagnosed our patient with FCE according to clinical presentation, laboratory data (which failed to reveal any other plausible diagnoses), and MRI findings. Although FCE is a very rare condition, physicians should be aware of the characteristics of FCE and should include FCE of the spinal cord in their differential diagnosis, especially for physically active young pediatric patients.

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Conflict of Interests

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References

1. Tosi L, Rigoli G, Beltramello A. Fibrocartilaginous embolism of the spinal cord: a clinical and pathogenetic reconsideration. J Neurol Neurosurg Psychiatry. 1996;60:55-60.
2. Han JJ, Massagli TL, Jaffe KM. Fibrocartilaginous embolism--an uncommon cause of spinal cord infarction: a case report and review of the literature. Arch Phys Med Rehabil. 2004;85:153-157.
3. Shah S, Bryant P. Fibrocartilaginous emboli in the pediatric population: The role of rehabilitation in facilitating functional recovery. J Pediatr Rehabil Med. 2018;11:53-56.
4. Nagata K, Tanaka Y, Kanai H, Oshima Y. Acute complete paraplegia of 8-year-old girl caused by spinal cord infarction following minor trauma complicated with longitudinal signal change of spinal cord. Eur Spine J. 2017;26:1432-1435.
5. Jones DD, Watson RE, Heaton HA. Presentation and Medical Management of Fibrocartilaginous Embolism in the Emergency Department. J Emerg Med. 2016;51:315-318.
6. Eid R, Raj A, Farber D, Puri V, Bertolone S. Spinal Cord Infarction in Hemoglobin SC Disease as an Amusement Park Accident. Pediatrics. 2016;138.pii:e20154020.
7. Nelson JA, Ho CY, Golomb MR. Spinal Cord Stroke Presenting With Acute Monoplegia in a 17-Year-Old Tennis Player. Pediatr Neurol. 2016;56:76-79.
8. Rengarajan B, Venkateswaran S, McMillan HJ. Acute asymmetrical spinal infarct secondary to fibrocartilaginous embolism. Childs Nerv Syst. 2015;31:487-491.
9. Bansal S, Brown W, Dayal A, Carpenter JL. Posterior spinal cord infarction due to fibrocartilaginous embolization in a 16-year-old athlete. Pediatrics. 2014;134:e289-92.
10. Reisner A, Gary MF, Chern JJ, Grattan-Smith JD. Spinal cord infarction following minor trauma in children: fibrocartilaginous embolism as a putative cause. J Neurosurg Pediatr. 2013;11:445-450.
11. Holland NR.J. Acute myelopathy with normal imaging. Child Neurol. 2013;28:648-50.
12. Manara R, Calderone M, Severino MS, Citton V, Toldo I, Laverda AM, et al. Spinal cord infarction due to fibrocartilaginous embolization: the role of diffusion weighted imaging and short-tau inversion recovery sequences. J Child Neurol. 2010;25:1024-1028.
13. Tan K, Hammond ER, Kerr D, Nath A. Fibrocartilaginous embolism: a cause of acute ischemic myelopathy. Spinal Cord. 2009;47:643-645.

14. Raghavan A, Onikul E, Ryan MM, Prelog K, Taranath A, Chennapragada M. Anterior spinal cord infarction owing to possible fibrocartilaginous embolism. Pediatr Radiol. 2004;34:503-506.
15. Beer S, Kesselring J. Fibrocartilaginous embolisation of the spinal cord in a 7-year-old girl. J Neurol. 2002;249:936-937.
16. Davis GA, Klug GL. Acute-onset nontraumatic paraplegia in childhood: fibrocartilaginous embolism or acute myelitis? Childs Nerv Syst. 2000;16:551-554.
17. Yousef OM, Appenzeller P, Kornfeld M. Fibrocartilagenous embolism: an unusual cause of spinal cord infarction. Am J Forensic Med Pathol.1998;19:395-399.
18. McLean JM, Palagallo GL, Henderson JP, Kimm JA. Myelopathy associated with fibrocartilaginous emboli (FE): review and two suspected cases. Surg Neurol. 1995;44:228-34; discussion 234-235.
19. Toro G, Roman GC, Navarro-Roman L, Cantillo J, Serrano B, Vergara I. Natural history of spinal cord infarction caused by nucleus pulposus embolism. Spine (Phila Pa 1976). 1994;19:360-366.
20. Naiman JL, Donohue WL, Prichard JS. Fatal nucleus pulposus embolism of spinal cord after trauma. Neurology. 1961;11:83-87.
21. Mateen FJ, Monrad PA, Hunderfund AN, Robertson CE, Sorenson EJ. Clinically suspected fibrocartilaginous embolism: clinical characteristics, treatments, and outcomes. Eur J Neurol. 2011;18:218-225.
22. Cuello JP, Ortega-Gutierrez S, Linares G, Agarwal S, Cunningham A, Mohr JP, et al. Acute cervical myelopathy due to presumed fibrocartilaginous embolism: a case report and systematic review of the literature. J Spinal Disord Tech. 2014;27:E276-281.
23. AbdelRazek MA, Mowla A, Farooq S, Silvestri N, Sawyer R, Wolfe G. Fibrocartilaginous embolism: a comprehensive review of an under-studied cause of spinal cord infarction and proposed diagnostic criteria. J Spinal Cord Med. 2016;39:146-514.
24. Hilton RC, Ball J, Benn RT. Vertebral end-plate lesions (Schmorl's nodes) in the dorsolumbar spine. Ann Rheum Dis. 1976;35:127-132.
25. Kyere KA, Than KD, Wang AC, Rahman SU, Valdivia-Valdivia JM, La Marca F, et al. Schmorl's nodes. Eur Spine J. 2012;21:2115-2121.
26. Bockenek WL, Bach JR. Fibrocartilaginous emboli to the spinal cord: a review of the literature. J Am Paraplegia Soc. 1990;13:18-23. Review.
27. DeSanto J, Ross JS. Spine infection/inflammation. Radiol Clin North Am. 2011;49:105-27.

Figure 1.

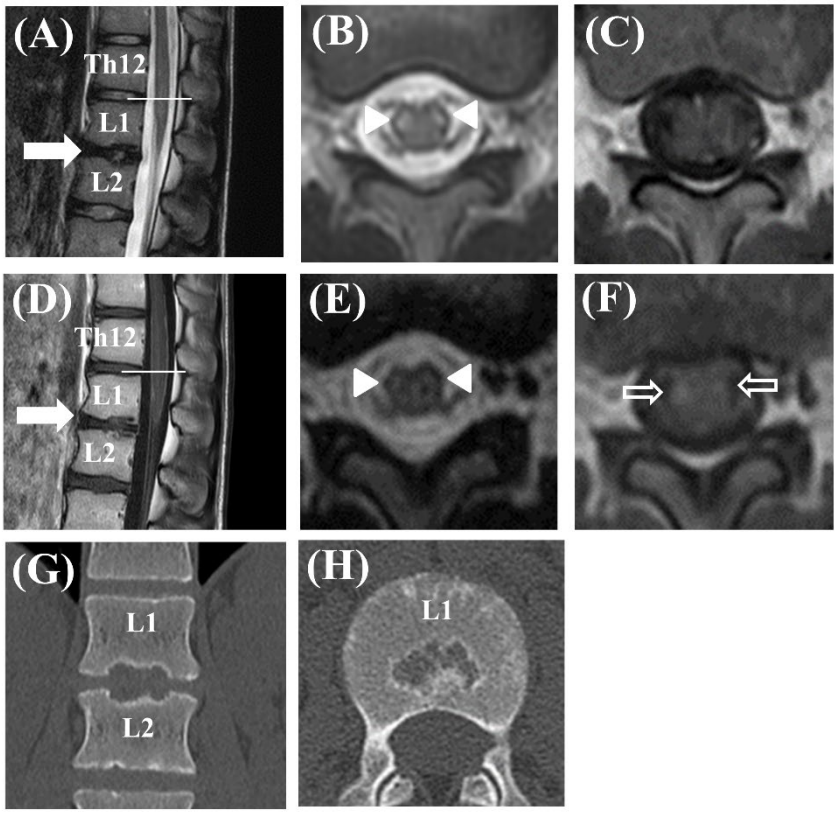

Figure legends

Figure 1. 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 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 disc collapse and Schmorl's node (white arrow) (A and D) were observed, along with adjacent vertebral end-plate destruction on computed tomography (G and H).

Tables

Table 1. Laboratory data of the patient

	Reference range	Value		Reference range	Value
CBC			**Antibody**		
WBC (/μL)	(3,900-9,800)	6,900	Toreponema antibody	(n.d.)	n.d.
Hb (g/dL)	(12.6-16.5)	12.2	Anti-nuclear (times)	(≦40)	< 40
Plt ($\times 10^4$/μL)	(17-41)	22	Anti-DNA (IU/mL)	(≦6)	≦2.0
			Anti-DS-DNA IgG (IU/mL)	(≦12)	< 10
Chemistry			anti-CL.B2GP1(U/mL)	(≦3.5)	≦1.2
AST (U/L)	(14-30)	23	anti-aquaporin4 (U/mL)	(≦3)	< 1.5
ALT (U/L)	(9-35)	11	MOG antibody	(n.d.)	n.d.
Na (mEq/L)	(138-144)	139	C3 (mg/dL)	(73-138)	93
K (mEq/L)	(3.7-4.7)	3.9	C4 (mg/dL)	(11-31)	25
Cl (mEq/L)	(102-109)	104	CH50 (/mL)	(25-48)	30.3
BUN (mg/dL)	(6.8-18.8)	10.4			
CRE (mg/dL)	(0.48-0.93)	0.80	**CSF**		
GLU (mg/dL)	(73-109)	85	Cells (/uL)	(0-5)	1
TP (g/dL)	(6.3-7.8)	7	Glu (mg/dL)	(47-69)	57
CK (U/L)	(50-275)	210	Protein (mg/dL)	(8.8-27.1)	31
TSH (μU/mL)	(0.55-4.78)	1.586	MBP (pg/mL)	(≦102)	≦40
Free T4 (ng/dL)	(0.95-1.74)	1.19	Oligoclonal band	(n.d.)	n.d.
ESR (mm) 1h	(2-10)	4	IgG index	(≦0.73)	0.38
2h	(4-20)	10			
Protein C activity (%)	(64-146)	90	**Others**		
Protein S activity (%)	(67-164)	76	T-SPOT	(n.d.)	n.d.
			Mycobacterium culture	(n.d.)	n.d.
Coagulation					
PT (%)	(70-130)	96			
PT (INR)	(0.9-1.1)	1.02			
APTT (sec)	(26.6-40.3)	27.4			
AT-III	(83-118)	111			
D-D	(≦0.9)	1			

Abbreviation: CBC, complete blood cell counts; WBC, white blood cells; Hb, Hemoglobin; Plt, platelet; AST, aspartate aminotransferase; ALT, alanine aminotransferase; BUN, blood urea nitrogen; CRE, creatinine; GLU, glucose; TP, total protein; CK, creatine kinase; TSH; thyroid-stimulating hormone; ESR, erythrocyte sedimentation rate; PT, prothrombin time; INR, international normalized ratio; APTT, activated partial thromboplastin time; AT, Antithrombin; D-D, D-dimer; MOG, myelin-oligodendrocyte glycoprotein; CSF, cerebrospinal fluid; MBP, myelin basic protein; n.d., not detected;

Table 2. Comprehensive review of our case and previous articles reporting fibrocartilaginous embolism in children younger than 18 years of age

Cases	Author	Publication (y)	Age	Sex (M/F)	Triger event	Time from trigger event to neurologic sympyoms/ Time to symptom peak	Initial neurologic symptoms	Pain	Abnormal findings of neurologic examination on admission	MRI findings (days after trigger event)	Presence of abnormal vertebral body or disc changes	Treatment	Lunber puncure	Suspected case/ Pathologically definite case	Prognosis at latest follow-up
Present case	Yamaguchi H, et al		15y	M	Kicked a ball during soccer practice	soon after the event/2d	Weakness of both lower extremities and ischuria	Tingling pain both calve	Mild distal weakness in right lower extremity Absentright Achilles DTR Umbulate difficulty without assistance and unable to bear any weight on his both legs Ischuria	T2WI hyperintensity in the anterior horn of the spinal cord at the Th12/L1 (d2) and enhanced lesion at T12/L1 (d8)	Schmorl's node (L1-L2)	iv high-dose steroids	Yes/elevated MBP and mildly elevated protein	Suspected	Steady gait after 3 wks; however, umbulation difficulty on tiptoe
1	Shah Sv, et al[3]	2018	12y	F	Unknown	-	Right-sided weakness, and paresthesia	Headache Neck	Moderate weakness in the right lower extremity Complete monoplesia in the right upper extremity Diminished sensation to light touch in the right forearm and upper arm	Normal (2d after admission) T2WI hyperintensity within the C2-C3 intervertebral disc posteriorly with a small focal protrusion (several d after admission)	No	Fluoxetine Aspirin IVIG iv steroids	Yes/unremarkable	Suspected	After several wks, able to walk with ankle foot orthotic for the right lower extremity
2	Nagata K, et al[4]	2017	8y	F	Hit back and neck with hyperflexion during a handstand	<2h/2h	Weakness in lower extremities	Back (minor)	Total paraplesia Complete loss of pain, temperture sensation and a sense of vibration below T11 region Bradycardia	Normal (d1) T2WI hyperintensity with the spinal cord T11-cranial level (d2) T2WI hyperintensity with longutudinal extension of the region between C6-T11 (d2) DWI hyperintensity below C6 (1wk) Minor bleeding at T11 vertebral region and spinal cord edema below C6 (T2 star)(2 wks)	No	None	Yes/unremarkable	Suspected	After 1y, complete paraplegia with sensory loss below T11
3	Jones DD, et al[5]	2016	14y	M	Run sprints and made quick turns at the gym	<1d/1d	Weakness of lower extremities and urinary incontinece	Lower back	Complete paraplesia in lower extremities with the exception of severe weakness in left extensor hallicus longus and dorsiflexion of left ankle Diminished bilateral sensation at L2-S1 region Absent patellar and Achilles DTR Absent volitional contraction of sphincter	T2WI hyperintensity over two-thirds of the spinal cord at T12-L1 with DWI restricted diffusion (d1)	Schmorl's node (Th12-L1)	Heparin iv steroids	No	Suspected	After 1 mo, bladder catheterization and able to walk with a front-wheeled walker
4	Eid R, et al[6]	2016	12y	F	Rode a roller coaster	<several h/several h	Left-sided weakness	Neck	Mild weaknesses in left sternocleidomastoid and trapezius and right deltoid Severe weakness in left upper extremity of deltoid Complete plesia in biceps, triceps, wrist flexion, wrist extension, and grip Mild weakness in hip flexion, foot dorsiflexion, and foot plantar flexion Moderate weakness in left lower extremity Mild slowing of coordination in the right upper extremity	T2WI hyperintensity along the midline portion of C2-C3 and swelling of the spinal cord between C2-C5 (d1)	No	iv high-dose steroids Enoxaparin	Yes/unremarkable	Suspected	After 1 y, left hemiparesis, spasticity of both upper and lower extremities, and limited motion of the right upper extremity with greater limitation in the right shoulder
5	Nelson JA, et al[7]	2016	17y	F	Intense tennis practice	<1d/5d	Left leg monoplegia and paresthesia	Back and left hip Bilateral lumber	Flaccid paralysis of the left lower extremity Mild weakness in righ lower extremity Decreased temperature sensation in the right lower extremity Highper DTR in right knee Urinary incontinence	A small disc extrusion with superior migration at T10-T11 with T2WI hyperintensity of the anterior spinal cord between T9-T11 (d3 and 5)	Schmorl's node (T11-T12)	Aspirin iv high-dose steroids Vitamin B12	Yes/elevated MBP	Suspected	After 18 mo, decreased pain, temperature, and pressure sensation and nearly baseline strength of right leg and left leg, and return to modified athletic activity
6	Rengarajan B, et al[8]	2016	14y/10y	M/F	Case1: Hammered fence posts with a sledgehammer Case2: Competed in a long jump event	Case1: 15min/several h Case2: <2h/a few h	Case1: Weakness of lower extremities, paraesthesia, and bowel and bladder incontinence Case2: Weakness of lower extremities, paraesthesia, and bowel and bladder incontinence	Back (center)/ Back and buttock	Case1: Mild weakness in right hip flexors and moderate weakness in distal muscles of the right lower extremity Complete plegia of left hip, knee, or ankle Absent DTR in the patella or ankle Decreased tone in both lower extremities Absent sensation of pin prick, light touch, temperature, and vibration below the T5 region Absent proprioception in lower extremities Case2: Moderate weakness in bilateral hip extensors, knee flexors and extensors No movement at the ankle and toes Absent DTR in patella and ankle Positive Babinski's reflexes Absent sensation to fine touch, pin prick, cold, vibration, or proprioception below L3 region	Case1: Long pencil-like segment of T2WI hyperintensity and restricted DWI in the anterior portion from C7/T1 to T6/T7 (d1) More apparent T2WI hyperintensity at same level (d10) Case2: T2WI hyperintensity and restricted DWI in the conus medullaris at the level of T12-L1 (d1) T1WI slightly hyperintensity at the anterior spinal artery at the T12-L1 and normal enhanced-MRI (d1)	No/No	Case1: iv high-dose steroids Enoxaparin Warfarin/ Case2: iv high-dose steroids Aspirin	Yes/unremarckable/ Yes/unremarckable	Suspected/ Suspected	Case1: After 13mo, able to walk, bike, and run normally. However, persistent right lower limb temperature hypoesthesia Case2: After 11mo, able to walk independently with a left ankle foot orthosis, and decreased sensation below knee

Table 2. *Continued*

Cases	Author	Publication (y)	Age	Sex (M/F)	Triger event	Time from trigger event to neurologic sympyoms/ Time to symptom peak	Initial neurologic symptoms	Pain	Abnormal findings of neurologic examination on admission	MRI findings (days after trigger event)	Presence of abnormal vertebral body or disc changes	Treatment	Lumber puncure	Suspected case/ Pathologically definite case	Prognosis at latest follow-up
7	Bamal S. et al[9]	2014	16y	F	Intense exercise	soon after the event/2d	Umbulate difficulty and paresthesia of lower extremities Tingling of the right leg	None	Mild weakness in the lower extremities in the bilateral iliopsoas and quadriceps Hightper DTR in the bilateral patella and positive Babinski's reflex in the right Diminished proprioception, temperature, and pinprick distal to T12 Dysmetriabilateral heel-shin-knee testing Wide-based gait and positive Romberg sign	Several T2WI hyperintensity in the dorsal spinal cord at T11-T12 (d4) restricted DWI at T11-T12 (d5)	Intervertebral disc desiccation and bulge at T11/T12	Anticoagulation	Yes/elevated MBP	Suspected	After 2mo, mild sensory changes in the right extremity
8	Reinner A. et al[10]	2013	8y/8m/12y	F/F/F	Case1: Danced at ballet class Case2: Fell off of a changing table Case3: Ground-level fall	Case1: soon after the event/ 2d Case2: 2h/2h Case3: soon after the event/ <1d	Case1: Paresthesia and numbness in hands and difficulty bending, and umbulation difficulty Case2: Paraplegia in upper extremities Case3: Complete quadriplegia and numbness over entire body	Neck/ None/ None/	Case1: Complete quadriplegia, diminished sensation to pain and temperature Case2: Paraplesia in her upper extremities Case3: Complete quadriplegia and diminished sensation over her entire body ventilatorary failure	Case1: T2WI hyperintensity in the C2-T1, and diminished T2WI hyperintensity in the C3-C4 (d2) Case2: Central spinal cord signal changes at C2-T1 (d1) Case3: T2WI hyperintensity and restricted DWI at C1-C4 (d1)	Case1: No Case2: T2WI hypointensity in multiple intervertebral discs (C2-C7) Case3: Diminished T2WI signal in C3-C4 disc space	iv steroids/ iv high-dose steroids/ iv high-dose steroids	Yes/unremarkable No/ Yes/unremarkable	Suspected/ Suspected/ Suspected	Case1: After 3mo, improved to an ASIA Grade D, ambulatory without assistance Case2: After 19mo, ASIA Grade C Case3: After 12mo, ASIA Grade D with residual weakness on the left side, and able to walk independently
9	Holland[11]	2012	17y	F	Unknown	-	Quadriparesis, ventilatorary failure and weakness in upper extremities	Back (between sholder blades)	Moderate weakness of deltoid and biceps, significant weakness of triceps and the wrists, and paraplegia Numbness in both legs coming up to the trunk Reduced DTR in the arms and absent in the legs Ischuria	T2WI and enhanced T1WI hyperintensity in the spinal cord at C5-7 (d7 after admission)	Small annular tear at C5-6	IVIG plasma exchange iv cyclophosphamide	Yes/unremarkable	Suspected	After several mo. weaned from the ventilator and regained some strength in the upper extremities. However, remained paraplegic and confined to a wheelchair
10	Manara. et al[12]	2010	13y/12y	M/F	Case1: Payed basketball, intense stretching, squanting and lifting exercises Case2: Fell backward, banging her back against a door	Case1: several d/several d Case2: a few h/a few h	Case1: Paraplegia, paresthesia in lower extremities Case2: Leg weakness, bowel and bladder incontinence	Case1: Back (interscapular) spreading to the epigastric region Case2: Back	Case1: Flaccid areflexic paraplegia with absent abdominal reflexes and hypoesthesia below T7 Bladder disfunction Case2: Flaccid areflexic paraplegia, bilateral mild hypoesthesia below T10	Case1: Slight intramedullary T2WI hyperintensity at the T7-T8 (several d) Intramedullary T2WI hyperintensity in the anterior spinal artery region at T4-T5 and T7 (several d-d2) Medullar T2WI hyperintensity in the anterior spinal artery territory (several d-d5) Case2: T2WI hypointensity of the L2-L3 disc (d1) T2WI and DWI hyperintensity "owl's eyes-like" within the spinal conus (d3)	Case1: Thiner intervertebral disc and T2WI hypointensity at T4-T5 Case2: Schmorl's node (L1-L2, L2-L3)	Case1: iv high-dose steroids Case2: iv high-dose steroids	Case1: Yes/unremarkable Case2: Yes/mild leukocytosis	suspected	Case1: After 1y, symptom-free Case2: After 2y, weakness in left lower extremity
11	Tan K. et al[13]	2009	16y	M	Elbowed in the face during gym class and wrestling	2d/2d	Weakness of lower extremities	Lower back	Complete paraplegia, sensory loss below T10 Urinary and bowel incontinence	Normal (d1) T2WI hyperintensity at T11-L2 (19mo)	Schmorl's node (L1-L2)	None	Yes/mildly elevated protein	Suspected	After 26mo, asymmetric spastic parparesis and reduced lower extremity sensation
12	Raghavan A, et al[14]	2004	14y	M	Trivial forced flexion of neck while playing with his dog	10d/10d	Weakness of all extremities	Back (interscapular)	Flaccid quadriplegia Absent DTR and abdominal cremasteric reflexes Absent pain and temperature sensation below C1 Urinary incontinence	T2WI hyperintensity with linear lesion at C1-T1 (d1)	No	None	No	Suspected	After 6wks, recovered antigravity strength of finger and wrist extension and flexion on the right, with a flicker of finger flexion on the left

Table 2. ***Continued***

Cases	Author	Publication (y)	Age	Sex (M/F)	Triger event	Time from trigger event to neurologic sympyoms/ Time to symptom peak	Initial neurologic symptoms	Pain	Abnormal findings of neurologic examination on admission	MRI findings (days after trigger event)	Presence of abnormal vertebral body or disc changes	Treatment	Lumber puncure	Suspected case/ Pathologically definite case	Prognosis at latest follow-up
13	Han JJ, et al[2]	2004	16y	M	Lifting exercises	1d/2d	Weakness in lower extremities, paraparesis and ischuria	Back (lumber)	Flaccid in left lower extremity, and severe weakness in right lower extremity Absent sensation to pinprick bilaterally below T6 level anteriorly and T8 level posteriorly Absent perianal pinprick sensation Trace DTR in the upper extremities and absent quadriceps and Achilles DTR Low sphincter tone	Normal (d1) T2WI hyperintensity at the T7-T8, localized within the left central and anterior spinal cord (2wks)	No	None	Yes/unremarkable	Suspected	After 8mo, normal strength in the upper extremities, abdominals, and right lower extremity Normal strength in left lower extremity except for mild weakness of knee flexion reduced pinprick and light touch on the right (T8-S5)
14	Beer, et al[15]	2002	7y	F	Car accident	<1d/1d	Paraplegia in lower extremities, paraparesis, ischuria, bowel incontinence	Chest (blunt trauma)	Flaccid paraplegia with a complete sensory loss below the T12 region, ischuria and bowel incontinence Absent Knee and ankle DTR	T2WI hyperintensity of the spinal cord from T4 down to the lumbar region (d3)	Narrowed intervertebral disc at T6/T7	None	No	Suspected	After 6mo, able to walk with ortheses Slight activity in all muscle groups of the lower extremities Marked spasticity of the lower extremities Impaired perception of touch, position sense and vibration below L3
15	Davis GA, et al[16]	2000	6y	F	Performed cartwheel	a few w/a few w	Paraplegia in lower extremities and bladder incontinence	Lower back	Flaccid paraplegia of lower extremities Absent DTR in lower extremities Absent pain and temperature sensation to T11	Multiseptated syrinx between T5 and T6-T10 (d1) T2WI hyperintensity in the anterior spinal cord from T10 to the conus medullaris (d2)	Narrowed intervertebral disc at L1/L2	iv steroids	Yes/unremarkable	Suspected	After 2mo, regained minimal strength in left quadriceps muscle, while the remainder of deficit not improved
16	Yousef, et al[17]	1998	14y	F	Leaning forward to pick up an object	Soon after the event/1d	Weakness and numbness in lower extremities and ischuria	Upper back	Flaccid paralysis of the lower extremities Decreased pain and light touch below level T6 Absent DTR in lower extremities	T8-T9 levels without evidence of spinal cord lesion (d1-2)	Irregularities of the end plates at T7-T8 and T8-T9	None	Yes/unremarkable	Defined	Death on the 3rd day
17	Tosi, et al[1]	1996	16y	F	Handstand	30min/1d	Left foot weakness	Back radiating to lower extremities	Absent flexion and extension of the left foot and weak on the right Absent ankle DTR Reduced sensation of touch, pain, and temperature on the left extremity below the knee	T2WI hyperintensity within the cord from T11-L1 (d3) and in the mid-dorsal region (d10) Pronounced atrophy of the epiconus at T11-T12 (T1WI) (5mo)	Schmorl's node (T12)	None	yes/elevated protein	Suspected	Not described
18	McLean, et al[18]	1995	17y	M	Weight lifting	12h/12h	Paresthesia in the lower extremities, paraplegia, and urinary incontinence	Burning pain in thighs, lower back, and lower extremity	Flaccid paralysis and absent sensation to all modalities at T12 Present priapism	T2WI hyperintensity at T7 (d3) Atrophy of the cord at the T11-12 (T1WI) (3mo)	No	iv high-dose steroids	Yes/unremarkable Yes/slight leukocytosis (Day5) Yes/elevated protein (3wks)	Suspected	After 24 mo, no neurological improvement
19	Toro, et al [19]	1994	16y	F	Milking a cow	15min/<1d	Paraplegia in lower extremities	Lower back	Flaccid paraplegia with completely sensory loss from the L1 down and Absent DTR in the lower extremities Ischuria Loss of perineal sensation	Not performed	Old fracture at L1-L2 vertebral body	None	Yes/elevated protein	Defined	Death after 6wks
20	Naiman, et al[20]	1961	15y	M	Basketball practice	40min/2h	Weakness of all extremities and ventilatorary failure	Epigastrium, back, neck and shoulders	Complete flaccid paralysis and Absent DTR in all extremities Respiratory paralysis Sensory loss extending up to T3-T4	Not performed	No	None	No	Defined	Death after 2h

Abbreviation: ASIA; American Spinal Injury Association, d; day, DTR; deep tendon reflex, DWI; diffusion weighted image, F; female, h; hour, iv; intravenous, IVIG; intravenous immunoglobulin, mo; month, min; minute, M; male, w: week, T1WI; T1 weighted image,T2WI; T2 weighted image, wk; week, y; year,-; not applicable

Weakness is categorized as follows (manual muscle test (MMT): 0/5 complete plesia, 1/5-2/5 severe, 3/5 moderate, 4/5 mild, 5/5 normal)

ASIA Impairment Scale is categorized as follows (Grade A; Complete lack of motor and sensory function below the level of injury, Grade B; Some sensation below the level of the injury, Grade C; some muscle movement is spared below the level of injury, Grade D; most of the muscles that are spared below the level of injury, Grade E; all neurologic function has returned to normal)