Fibrocartilaginous Embolism Spinal Cord Infarction, Mistaken for Glial Fibrillary Acidic Protein Autoimmune Transverse Myelitis: A Case Report

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ABSTRACT

Introduction: Fibrocartilaginous embolism causing spinal cord infarct is rare, and a high index of clinical suspicion is needed for accurate diagnosis.

Case Presentation: A 65-year-old woman presented to our hospital with back pain, paraparesis, and neurogenic bladder. Magnetic resonance imaging showed a T4-T7 signal abnormality that was misdiagnosed initially and treated as autoimmune myelitis.

Discussion: Fibrocartilaginous spinal cord infarction is rare and remains a clinical diagnosis with supportive imaging findings. The imaging findings may be nonspecific, and other etiologic diagnostic considerations must be excluded.

Conclusion: Fibrocartilaginous embolism causing spinal cord infarct can be mistaken for transverse myelitis. A high index of clinical suspicion with clinical and radiologic correlation is necessary to make accurate diagnosis and avoid unnecessary treatment.

INTRODUCTION

Spinal cord infarctions are uncommon when compared to their cerebral counterpart. Unlike cerebral infarctions, spinal cord infarct due to fibrocartilaginous embolism (FCE) from intervertebral nucleus pulposus is a rare but well-recognized phenomenon. It was first described by Naiman in 1961.[1] This condition often affects a broader age range, including young adults and pediatric patients without obvious stroke risk factors.

The spinal cord has a complex arterial blood supply with significant individual variability. A single large anterior spinal artery runs ventrally in the midline from vertebrobasilar junction to the filum terminale. The spinal cord receives auxiliary supply from an inconsistent array of variable numbers of radiculomedullary and descending aorta branches.[2] Posteriorly, there are paired posterior spinal arteries that primarily supply the dorsal spinal cord, which are fed by similar radicular branches at every spinal level.[2] Spinal cord blood supply can be divided into 4 semi-distinct territories. The first extends from C1 to T3 and derives blood supply from the vertebral artery. The second region, which extends from T3 to T7, is often supplied by the left intercostal artery. The third region, which extends from T8 to T12, receives supply from the artery of Adamkiewicz; and the fourth region, which includes the conus, sometimes is supplied by branches from the internal iliac artery.[3] FCE causes spinal cord infarction when dislodged material from fibrocartilaginous nucleus pulposus causes occlusion of a spinal radicular artery at or near the level of disc extrusion. Identification of an associated vertebral body infarction is suggested to be a confirmatory sign of FCE-associated spinal cord ischemia.[4]

CASE PRESENTATION

A 65-year-old woman presented to our hospital with a 1-day history of mid back pain, numbness from the upper thorax to bilateral feet, subjective bilateral leg weakness, and difficulty emptying her bladder.

A few days prior to presentation, she was engaged in house painting but otherwise reported no physical trauma. Her prior medical history included stage IV low grade B-cell lymphoma diagnosed in 2022 but now in remission, diet-controlled diabetes, hypertension, tobacco use, obstructive sleep apnea, and

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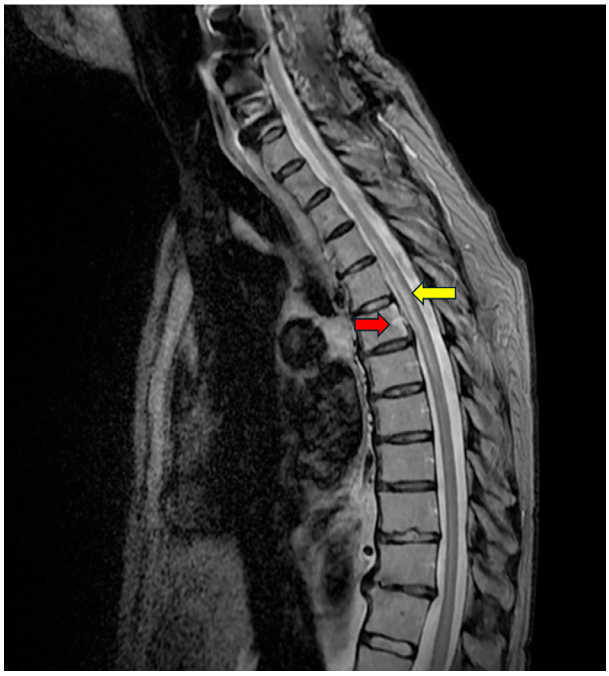
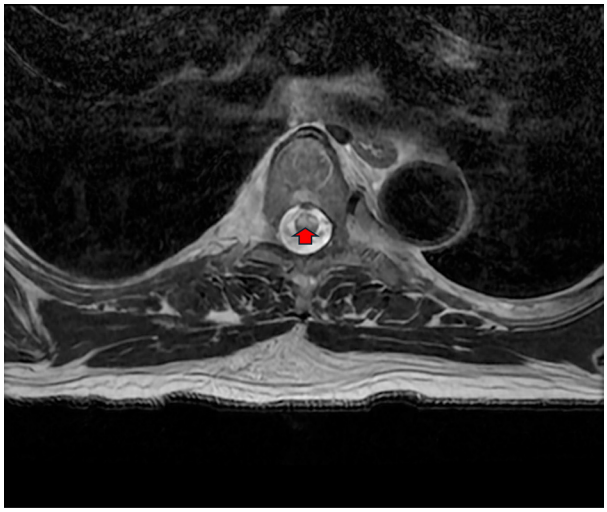
Figure 1. Sagittal T2 Magnetic Resonance Imaging Scan



Scan showing increased signal abnormality involving the ventral aspect of the cord from T4-T7 (yellow solid arrow) and T2 prolongation signal abnormality of the T6 vertebral body (solid red arrow).

Figure 2. Axial T2 Thoracic Spine Magnetic Resonance Imaging



Scan showing hyperintense signal abnormality predominantly involving the ventral aspect of the cord at T6 (red arrow).

cervical laminectomy with fusion from C4-C5 and C5-C7 14 years earlier. She had a retained bone growth stimulator from this procedure.

Initial neurologic examination showed normal cranial nerve testing, normal strength of proximal and distal upper and lower extremity muscles, diffuse hypoactive reflexes, sensory deficit to mild noxious stimulation to thoracic spinal cord level 4 (T4) with preserved position and vibratory sensation (proprioception), as well as urinary retention with a 220 cc post-void residual volume. Planned magnetic resonance imaging (MRI) was delayed due to patient safety concerns related to the bone stimulator, and the patient was provided supportive care.

On hospital day 3, she continued to report mid back pain and diffuse weakness. Neurologic examination revealed mild bilateral proximal lower extremities with Medical Research Council (MRC) grade 4/5 weakness and normal deep tendon reflexes but persistent T4 sensory level to mild noxious stimuli with preserved proprioception.

MRI of the cervical spine showed expected postsurgical changes without spinal canal narrowing. Thoracic spine MRI showed a T2-signal hyperintensity ventrally from T4-T7, with associated disc extrusion at T6-T7. There was no cord compression. A linear T2-signal prolongation abnormality involving the posterior aspect of the T6 vertebral body with marrow enhancement was noted (Figures 1 and 2). Preliminary cerebrospinal fluid (CSF) study showed albuminocytologic dissociation (red blood cells 2, nucleated cells 4, and protein 102 mg/dl). Infectious, inflammatory, and autoimmune laboratory markers were ordered. Brain MRI performed on hospital day 4 was normal. Based on the patient's clinical presentation of subacute motor deficits, sensory level, autonomic dysfunction, and initial MRI thoracic spine results and CSF findings, a preliminary diagnosis of transverse myelitis was made. Viral etiologies returned negative. She was started on high-dose intravenous methylprednisolone, 1 gram daily for 3 days. Her symptoms of proximal leg weakness improved to normal, and she was discharged home with a plan for outpatient follow-up.

The patient was re-admitted to the hospital 3 days postdischarge for progressively worsening bilateral lower extremity weakness and numbness. Neurologic examination now revealed MRC grade 2/5 proximal and 3/5 distal bilateral lower extremity weakness with hyperactive reflexes associate with nonsustained (2 beats) ankle clonus on the right side. Also, sensory level to T4 persisted. Upper extremity strength and reflexes were normal. An additional 3 days of high-dose methylprednisolone was initiated.

Further testing included a negative lumbar spine MRI. Computer tomography (CT) scan of chest, abdomen, and pelvis showed mild prominent axillary and mediastinal adenopathy. Fluorodeoxyglucose positron emission (FDG-PET) scan was stable, without signs of recurrent lymphoma. Extensive laboratory testing was negative for human immunodeficiency viruses 1 and 2, leukemia/lymphoma immunotyping by flow cytometry, hepatitis panel, Powassan virus, West Nile virus, and Lyme serologies. CSF bacteria, fungal, and viral cultures/serologies were negative. Multiple sclerosis panel showed elevated immunoglobulin kappa free light chain at 0.4760 (reference < 0.1000) and oligoclonal

bands at 3 (reference <2). Myelopathy autoimmune panel was negative except for positive glial fibrillary acidic protein (GFAP) antibodies by immunofluorescence assay. A preliminary diagnosis of GFAP-positive autoimmune transverse myelitis was made pending confirmatory cell-based assay. The patient was started on a 5-day plasmapheresis regimen due to persistent paraparesis. Several days later, the GFAP confirmatory cell-based assay returned negative. Repeat thoracic MRI 17 days after the initial study showed stable findings. However, upon further careful review of both thoracic MRI studies by Neuroradiology, it was observed that the T2-signal abnormality involving the T6 vertebral body represented a subacute ischemic bone infarct. Given the proximity and overlapping vascular distribution to the spinal cord lesion and the associated disc extrusion at or near the same level, a final diagnosis of FCE spinal cord infarction was made. The patient was started on aspirin 81 mg and rosuvastatin 20 mg daily. Alternative stroke mechanism was excluded with negative hypercoagulable profile, transthoracic echocardiogram, and a 30-day cardiac event monitor.

Upon hospital discharge 25 days after the initial admission, the patient's lower extremity paraparesis had improved. The sensory deficit descended to T6 level with preserved proprioception. On outpatient follow-up evaluation 1 month after discharge, she required intermittent catheterization for neurogenic bladder, and her lower extremity showed trace proximal muscle weakness with mild ataxic gait on rapid turns only.

Table. Proposed Schematic Approach to Diagnosis of Fibrocartilaginous Embolism (FCE) Causing Spinal Cord Infarction

Steps	Description
1	Establish clinical syndrome of myelopathy, sensory level most important
2	Exclude traumatic and compressive etiologies of myelopathy by history and imaging using MRI (preferred) or CT scan with and without contrast
3	Exclude inflammatory etiologies of myelopathy using CSF. Absence of pleocytosis or increased IgG index
4	Establish the diagnosis of spinal cord infarction. Requires steps 1-3 plus 1 "major" criterion or 2 "minor" criteria
	Major Criteria • Clear vascular distribution by exam such as sparing of proprioception • Clear vascular distribution on imaging, axial views MRI • Radiologic changes, MRI T2-hyperintensity in the vertebral body or inter-vertebral disc adjacent to the cord infarction
	Minor Criteria • Accompanying new onset neck or back pain • Symptom progression to nadir or near nadir in 4-8 hours • Initial unremarkable MRI of the spinal cord with subsequent evolution of an intra-parenchymal lesion
5	Establish the high likelihood of FCE. This requires the absence of other more common etiologies of spinal cord infarction, mainly being aortic pathologies, plus presence of one or more of the following: • Temporal relation to heavy lifting or minor neck or back injury or any event that can cause increase intra-disc or intra-vertebral pressure like axial falls, or events that can reverse the venous drainage of the spinal column away from the heart and to the spinal cord instead such as Valsalva maneuver. • Presence of degenerative disc disease especially protrusions or Schmorl's nodes at or near the infarction • Absence of more than 1 vascular risk factor.

Abbreviation: MRI, magetic resonance imaging; CT, computed tomography; CSF, cerebrospinal fluid; IgG, immunoglobulin.

DISCUSSION

Histopathologic evidence for thrombosis of radicular spinal arteries due to fibrocartilaginous nucleus pulposus leading to spinal cord infarction have been well-documented.[5,6] Our patient presented with back pain without specific trauma, followed by subacute onset of fluctuating bilateral lower extremities paraparesis, persistent sensory level, and neurogenic bladder. The initial fluctuating weakness made this case more difficult to identify as an ischemic process.

A study of 41 histopathologically confirmed cases of spinal cord infarctions due to FCE showed 64% were female. Average age was 41 (range 14–78 years) with clinical picture at presentation of transient neck or back pain, followed by syndrome of myelopathy with abnormal sensory level, neurogenic bladder, and paraplegia in cases involving the thoraco-lumbar spinal cord or quadriplegia in cases of cervical spinal cord involvement. Time to symptom peak ranged from 15 minutes to 21 days.[6]

Our case was notably different from previous reports regarding fluctuating paraparesis. Our patient received high-dose methylprednisolone on admission with presumptive diagnosis of transverse myelitis. The steroid effect may have played some role in the fluctuating nature of the initial symptoms. Additionally, clinical presentation of spinal cord ischemia can vary depending on location and extent of infarction. A rapid decline of function within 12 hours of onset with severe neurologic deficit is characteristic; however, up to 17% of patients have biphasic syndrome with transient or mild symptoms followed by deterioration.[7] We also observed initial hyporeflexia in our patient, as was the case in multiple other studies.[4-6,8] Flaccid paraparesis with hyporeflexia is the dominant initial finding in spinal cord ischemia due to spinal shock. Over time, hyperreflexia, spasticity, and extensor plantar reflexes prevail.

Our patient had elevated immunoglobulin CSF free kappa light chain and 3 oligoclonal bands, suggesting possible intrathecal immunoglobulin synthesis commonly found in multiple sclerosis, as well as other infectious, autoimmune, and inflammatory central nervous system pathologic states known to trigger a humoral immune response. She also has a history of B-cell lymphoma, although in remission. Interestingly, elevated immunoglobulin free light chain is present in 27% of patients with diffuse B-cell lymphoma,[9] and systemic immunoglobulins may cross the blood-brain-barrier by passive transfer,[10] necessitating a cautious and careful interpretation of abnormal levels in CSF.

Spinal cord ischemic infarct can be mistaken for and treated as an inflammatory or autoimmune transverse myelitis. FCE-

associated spinal cord infarction is rare and remains a clinical diagnosis with supportive imaging findings. The imaging findings may be nonspecific, however, and other etiologic diagnostic considerations need to be excluded, such as autoimmune, inflammatory, infectious, and metabolic transverse myelitis.

A schematic approach to the diagnosis of FCE (Table) as proposed by AbdelRazek et al[6] serves as a useful template for the diagnosis of this elusive disorder. Our case meets the major steps and all 3 elements of the major criteria of this schematic, including evidence of vertebral body infarction.

CONCLUSIONS

As demonstrated by this case, spinal cord infarction due to FCE can be mistaken for an inflammatory or autoimmune transverse myelitis. A high index of clinical suspicion, cautious interpretation of laboratory findings, and multidisciplinary review of the spinal cord imaging is essential. Correlation with a detailed patient history and clinical symptoms as they evolve are needed to arrive at the correct diagnosis.

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