Pearls & Oy-sters: Fibrocartilaginous embolism myelopathy

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CLINICAL PEARL Fibrocartilaginous embolism myelopathy is a stroke syndrome, characterized by rapidly progressive paraplegia (hours to 2–3 days) following an episode of back pain (mostly after a minor trauma).[1] CSF studies are normal and MRI shows T2 hyperintensity in the spinal cord with associated swelling, diffusion restriction, and often presence of Schmorl's nodes at the level of injury.[1]

CASE REPORT A 25-year-old man with no significant past medical history and an active lifestyle presented to the hospital with a complaint of bilateral lower extremity numbness and weakness. Three days prior, the patient slipped while getting out of his truck and fell into a split position. Immediately afterwards he felt a sudden pain in his left groin with associated paraesthesia and weakness initially in his left thigh. Within several hours, the weakness and paraesthesia spread over the entire left leg and 24 hours later to the right leg. He also felt his bladder was full and lost control of his flatulence.

The patient was a well-developed man in mild distress with normal vital signs. On neurologic examination, mental status and cranial nerves were normal. Manual motor testing of his upper extremities was unremarkable. Testing of his lower extremities revealed increased tone bilaterally. In the right lower extremity his iliopsoas was 4/5, hamstrings were 4+/5, quadriceps were 4+/5, tibialis anterior was 5/5, and gastrocnemius was 5/5. In the left lower extremity, his iliopsoas was 1/5, hamstrings were 4−/5, quadriceps were 4−/5, tibialis anterior was 4/5, and gastrocnemius was 4+/5. Muscle bulk was normal. Rectal sphincter tone was diminished. He had no adventitial movements. Deep tendon reflexes were 2+/4 in the upper extremities and 4+/4 in the lower extremities bilaterally, with several beats of ankle clonus and bilateral extensor response in the toes. Sensory examination revealed intact vibration, proprioception, and pain and temperature sensation in the upper extremities bilaterally. There was an L1 sensory level involving pinprick and temperature testing. Proprioception and vibration were intact. Light touch elicited dysesthesias in the left lower extremity. Coordination was intact. The patient was able to sit up with assistance but was not able to bear any weight on his legs.

The patient was admitted to a telemetry unit and had a thoracolumbar spine MRI, which showed a hyperintense T2 signal in the central and left posterior aspect of the cord from T9 through T11, slightly greater in the craniocaudal dimension. There were Schmorl's nodes at the inferior T10, superior T11, inferior T11, and superior T12 end plates (figure). Axial and sagittal MRI T1 pre- and post-gadolinium images did not show any evidence of enhancement at the level corresponding to the T2 hyperintensity. A lumbar puncture revealed an opening pressure of 29 cm of water with CSF analysis showing leukocytes 2/μL, erythrocytes 4/μL, glucose 63 mg/dL, and protein 44 mg/dL. CSF Lyme antibody was negative. CSF myelin basic protein was elevated at 307 ng/mL. CSF Venereal Disease Research Laboratory was nonreactive. CSF angiotensin converting enzyme level was normal. CSF protein electrophoresis was normal. Fluid culture and Gram stain were negative. The patient had normal bedside electromyography/nerve conduction tests and somatosensory evoked potentials.

Based on the above findings, a diffusion-weighted MRI of the thoracolumbar spine was ordered, which revealed restricted diffusion corresponding to the signal abnormality seen on T2-weighted sequences at the level of T9–T11 (figure). The patient was treated for spasticity and constipation and was provided with physical therapy. He was discharged with the presumptive diagnosis of fibrocartilaginous embolism myelopathy to a rehabilitation facility with improvement in his lower extremity strength and bladder function.

DISCUSSION In 1961, the first case of fibrocartilaginous embolism myelopathy was reported in a 15-year-old boy who became progressively tetraplegic

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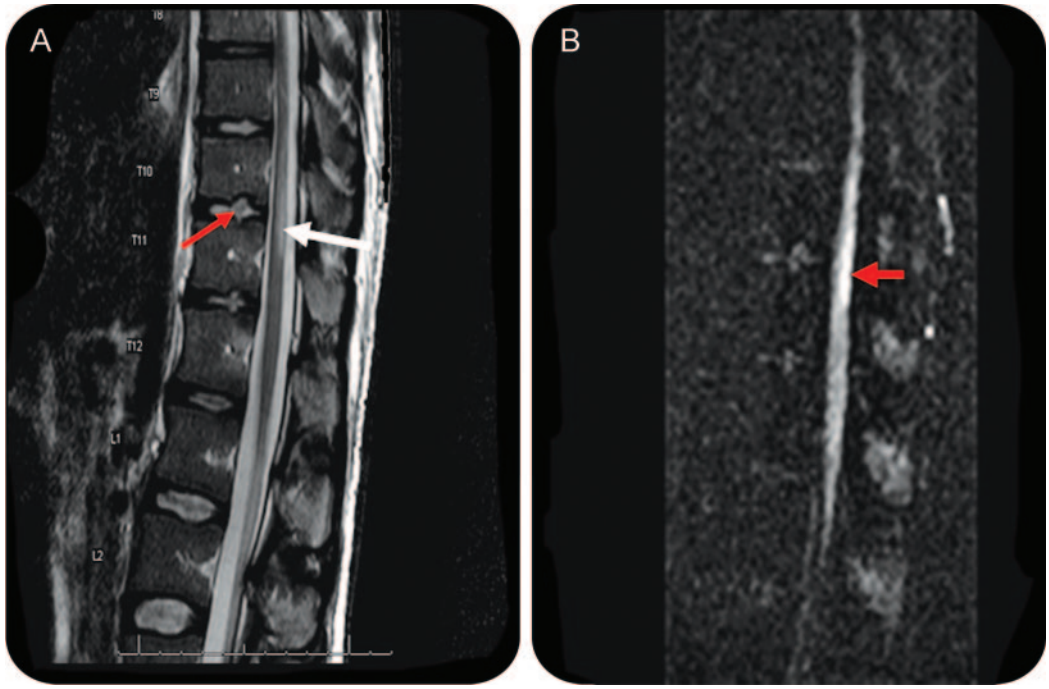
Figure Sagittal MRI T2 and diffusion-weighted imaging sequences of the thoracolumbar spine



(A) Sagittal T2-weighted SPIR sequence with large white arrow pointing to the intraaxial hyperintense signal abnormality (T9–T11) and small red arrow pointing to a Schmorl's node at the level of the injury. (B) Sagittal DWI sequence with large red arrow showing diffusion restriction at T9–T11.

with breathing difficulties and died after a seemingly minor fall on his coccyx during a basketball game.[2] Since then, over 40 cases have been reported in the human literature and many more in the veterinary literature. The ages reported for this clinical condition ranged from the first to the sixth decade of life with peaks in adolescence and in late middle age.[3] This condition affects mostly young women and nearly always involves the cervical cord.[4]

The finding of fibrocartilaginous material in spinal cord arterioles and venules on postmortem biopsies in conjunction with abnormal signals and subsequent atrophy on MRI of affected spinal cord segments suggests that fibrocartilaginous embolism is a probable cause of the acute spinal cord injury.[5] Several mechanisms describing how these cartilaginous particles travel to the spinal cord vasculature have been proposed. One suggestion is that central pressure to the spinal column from minor trauma results in breaking of the nucleus pulposus with secondary infiltration of the ruptured material into the small arteries present in degenerating disks in the adult population and as embryologic remnants from childhood, followed by retrograde flow into the perforating branches of the anterior spinal artery.[6] Another proposed mechanism suggests that herniated intervertebral disk material (Schmorl's nodes) is propelled into the spinal veins of the vertebral bone marrow by retrograde flow during Valsalva maneuvers.[6] Finally, the presence of a spinal arteriovenous connection may serve as a channel by which cartilaginous material from the spinal veins can propagate to the anterior spinal artery circulation.[6]

A common blood supply to the spinal cord and the nucleus pulposus is present in early life and normally becomes avascular after adolescence.[3] This may explain the increased incidence of fibrocartilaginous embolism myelopathy around adolescence. The incidence of fibrocartilaginous embolism myelopathy may be increased in later life as a result of the neovascularization that takes place in a degenerating disk, which can reestablish the common blood supply described above.[3]

The clinical outcomes of patients diagnosed with this condition are dictated by the localization of the cord infarct. There appears to be a higher reported mortality in patients with cervical cord involvement.[7] At this time, there are no clear recommendations for follow-up for this condition; however, in our case we recommended to the patient to avoid heavy lifting and frequent Valsalva maneuvers.

DISCLOSURE

Dr. Roshal and Dr. Gutierrez report no disclosures. Dr. Brock serves on a speakers' bureau for Sanofi-Aventis. Dr. Kremens has received honoraria for lectures and educational activities not funded by industry; serves as a consultant to Teva Pharmaceutical Industries Ltd. and Gershon Lehman Group; and has served/serves on speakers' bureaus for Teva Pharmaceutical Industries Ltd., Novartis, Allergan, Inc., GlaxoSmithKline, and UCB.

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