Case Report

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An ischemic myelopathy case series: Flaccid paraplegia following a spike ball save and numbness while walking normally

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Abstract:

Spinal cord infarctions in children are rare and early magnetic resonance imaging studies are often negative. A high clinical suspicion must be maintained to identify stroke and initiate workup for underlying etiology to suggest appropriate treatment. We present two cases of spinal cord infarction without major preceding trauma. The first was caused by disc herniation and external impingement of a radiculomedullary artery and the second was due to fibrocartilaginous embolism with classic imaging findings of ventral and dorsal cord infarctions, respectively. These cases were treated conservatively with diagnostic workup and aspirin, though additional treatments which can be considered with prompt diagnosis are also explored in our discussion. Both cases recovered the ability to ambulate independently within months. Case 1 is attending college and ambulates campus with a single-point cane. Case 2 ambulates independently, though has some difficulty with proprioception of the feet so uses wheelchairs for long-distance ambulation.

Keywords:

Artery of Adamkiewicz, fibrocartilaginous embolism, radiculomedullary artery, stroke, surfer's myelopathy

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Introduction

Spinal cord stroke presents as an acute or hyperacute weakness or sensory deficits below the level of the lesion which can be symmetric or asymmetric. Etiologies of spinal cord infarct are many, including traumatic and atraumatic causes. We present two cases of atraumatic spinal cord stroke of the anterior spinal artery distribution and posterior spinal artery distribution, respectively, with classic clinical and imaging presentations. Anterior spinal artery infarcts result in weakness below the level of the lesion due to involvement of the ventral horn of the spinal cord containing the motor neurons, loss of pain and temperature sensation due to involvement of the decussating anterolateral system, and autonomic dysfunction early with late findings including spasticity, hyperreflexia, long-tract signs, neurogenic bowel and bladder, and sexual dysfunction. Posterior spinal artery distribution infarct involves loss of the dorsal column-medial lemniscus system which results in loss of epicritic sensory modalities such as vibratory sense, two-point discrimination, proprioception, and fine touch, dorsal horn sensory neurons, and portions of the corticospinal tract resulting in weakness in some cases. We present two cases with workup and treatment along with exploration of the differential for atraumatic spinal cord infarcts.

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Case Reports

Case 1

While playing spike ball, a previously healthy 17-year-old boy experienced acute onset back pain and weakness and decreased sensation in both legs. He had bent forward to hit a low ball underhanded and felt a pop sensation in his back immediately at the onset of symptoms. He did not fall or sustain any trauma. A magnetic resonance imaging (MRI) study of the spine without contrast performed within 2 h of the injury was interpreted as normal. He was subsequently transferred to Riley Hospital for Children, a tertiary care facility in Indianapolis, IN. Upon arrival, his sensation had improved significantly; however, he remained densely, and symmetrically plegic in both lower extremities. He retained full strength and sensation in his upper extremities. His associated symptoms included transient lightheadedness, dizziness, and large-volume urinary retention of up to 900 mL. He had no encephalopathy, neck stiffness, seizures, fever, or headaches. MRI of the whole spine with and without contrast was repeated 3 days after the initial injury and showed spinal cord infarction in the territory of the anterior spinal artery (ASA) territory from T5 to T8 with multi-level Schmorl's nodes from T7 to L3 [Figure 1].

The workup for spinal cord stroke included a complete blood count with differential, demonstrating mild thrombocytopenia of 149 K/mm^3 (Ref 150–450) and basic metabolic panel, which was unremarkable. C reactive protein was elevated at 3.9 mg/dl (Ref <1.0), while other inflammatory markers including erythrocyte sedimentation rate, compliment 3, complement 4, and ANA titer were within normal limits. Hypercoagulability workup showed slightly elevated homocysteine of 13.9 mCmol/L (Ref 4.3–9.9) and factor VIII assay of 185% (Ref 43–155), although additional workup with Factor V Leiden 1691 G >A polymerase chain reaction (PCR), Prothrombin gene mutation 20210 G >A PCR, Cardiolipin immunoglobulin G (IgG) and IgM, Beta 2 glycoprotein IgG and IgM, Dilute Russel viper venom time (DRVVT), antithrombin III functional assay, Protein C activity, Protein S clottable, and Lipoprotein (a) were all unremarkable. In consultation with hematology, and neurology, and neurointerventional radiology, the patient started aspirin 81 mg orally daily and enoxaparin subcutaneously at prophylaxis dose enoxaparin due to immobility.

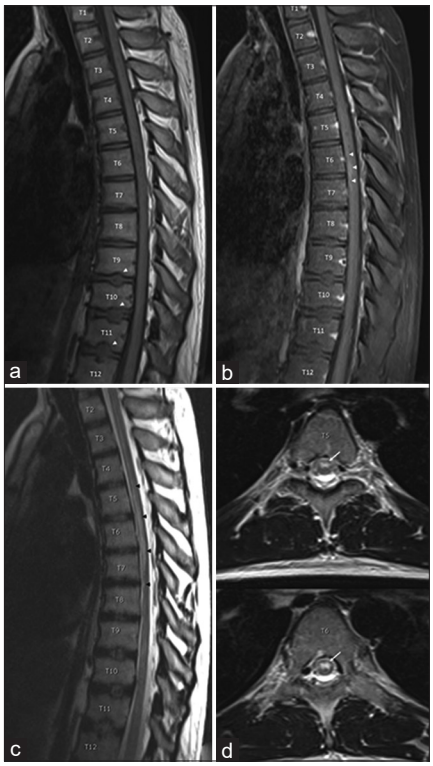

Figure 1: MRI Thoracic Spine without and with gadolinium contrast performed 3 days after symptom onset. (a) Noncontrast sagittal T1 Thoracic Spine MRI: Arrowheads show superior border of multilevel Schmorl's nodes. (b) Postgadolinium sagittal T1 Thoracic Spine MRI: Arrowheads show posterior border of faint enhancement of the anterior spinal cord. (c) Noncontrast sagittal Thoracic Spine T2 MRI: Arrowheads show corresponding edema of the anterior cord from T5 to T8. (d) Noncontrast Axial Thoracic Spine T2 MRI: Arrows show edema of the anterior cord in the distribution of the ASA at the levels of T5 and T6. MRI: Magnetic resonance imaging, ASA: Anterior Spinal Artery

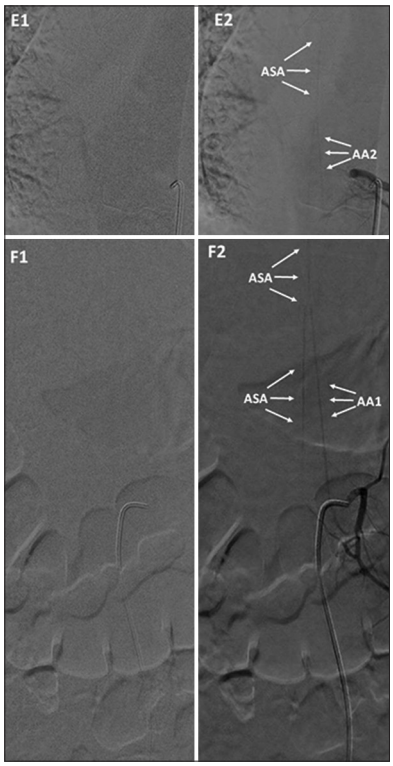

Figure 2: Digital subtraction angiography of the spine. (E1) Catheterization of left T8 intersegmental artery. (E2) Contrast reveals a prominent radiculomedullary artery, a duplicate AA2 with severely decreased contrast opacification of both AA2 and the ASA. (F1) Catheterization of left L1 intersegmental artery. (F2) Contrast reveals prominent radiculomedullary artery, the AA1 with robust contrast opacification and normal filling of the ASA. ASA: Anterior Spinal Artery, AA1: Artery of Adamkiewicz 1, AA2: Artery of Adamkiewicz 2

Additional imaging including computed tomography angiography (CTA) of the chest, abdomen, and pelvis was normal and showed no aortic abnormality. Echocardiogram with an agitated saline study revealed normal cardiac anatomy and function without right-to-left shunt. Conventional spinal angiography (CSA) demonstrated stagnant flow in the ASA with evidence of external compression of an accessory artery of Adamkiewicz (AA), arising from the left T8 level intersegmental artery. There was no noted compression of the left T8 radiculomedullary artery without definite filling defect or complete occlusion and lack of annular fissure on MRI imaging. The etiology of this stroke is likely suggestive of extrinsic compression of the radiculomedullary artery due to disc herniation rather than fibrocartilaginous embolism (FCE) or embolus, though FCE remains a diagnosis based on autopsy and the absence of complete filling defect does not completely exclude embolism as emboli may pass if small enough [Figure 2].

Over the course of 2 months, the weakness improved with acute rehabilitation, and he walked up to 1000 feet with crutches. Evolving spasticity of both legs was treated with low dose baclofen 5 mg orally twice daily, and he is now ambulatory without assist devices. However, he continues to have neurogenic bladder and impotence. He has since weaned from baclofen and now ambulates with a single-point cane around his college campus.

Case 2

A 15-year-old previously healthy boy presented with decreased sensation in his legs, left greater than right, without significant weakness, upon waking and walking. He reported a preceding 4 days of back pain after getting up from a bath, which was exacerbated when he picked up his younger brother the day before onset. He reported numbness and tingling in the left foot, with spasms throughout the left lower extremity, which worsened during the ride to the hospital. When he arrived, he also had pain that spread up the left leg, involving his genitals, buttocks, the right groin, and right thigh. He retained full strength and sensation in the upper extremities. Fine touch was decreased over the left leg and a sensory level was detected in the left hip, though proprioception and crude touch were intact bilaterally. Superficial abdominal reflexes were decreased over the left lower quadrant. Upper motor neuron signs were absent. There was no bowel or bladder involvement. He had no encephalopathy, neck stiffness, seizures, fever, or headaches. An MRI of the whole spine without contrast on the 1st day demonstrated an abnormal T2 hyperintense signal within the dorsal spinal cord at T10-T11, though it was motion limited. There was adjacent intervertebral disc herniation at T10-T11 and Schmorl's nodes, which raised suspicion for FCE leading to a dorsal spinal cord stroke. He did not have CSA at the time of presentation.

Lumbar puncture showed slight glycorrhachia with total nucleated cells of 3/mm^3 (Ref 0–5), red blood cells of 44/mm^3 (Ref 0–5), glucose of 75 mg/dl (Ref 40–70), protein of 29 mg/dl (Ref 15–45), negative culture and stain, and elevated myelin basic protein of 60.4 ng/ml (Ref 0–5.5). Additional cerebrospinal fluid studies including Aquaporin 4 IgG titer, Myelin Oligodendrocyte Protein IgG titer, IgG Index, and isoelectric focusing and immunoblotting were all unremarkable.

Hypercoagulability workup was performed with complete blood count, complete metabolic panel, prothrombin time, international normalized ratio, DRVVT, Protein C Ag, Protein S Ag, Antithrombin Ag, Homocysteine, Factor V Leiden 1691 G >A PCR, Prothrombin Gene 20,210 G >A PCR, Cardiolipin IgG, IgA, and IgM, and Beta 2 Glycoprotein IgG and IgM were all unremarkable. Iron studies showed low serum iron of 34 mCg/dl (Ref 50–212) though low total iron binding capacity and ferritin were normal. In consultation with hematology and neurology, the patient started aspirin 81 mg orally daily.

The patient underwent outpatient physical and occupational therapy for his abnormal gait. He returned to the hospital approximately 3 weeks later with worsening back pain. MRI of the spine was stable, and he was started on gabapentin 300 mg orally twice daily [Figure 3]. One month after injury, he was able to walk without assistive devices, although he uses a wheelchair for longer distances due to continued sensory

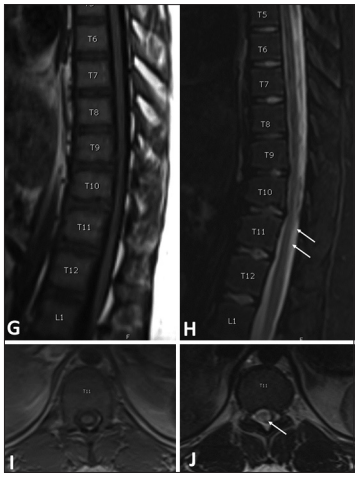

Figure 3: MRI Thoracic spine without gadolinium contrast performed 3 weeks after symptom onset. (g) Noncontrast sagittal T1 Thoracic Spine MRI, (h) Noncontrast sagittal Thoracic Spine T2 MRI: arrows show edema of the dorsal cord inferior to T10, (i) Noncontrast axial T1 Thoracic Spine MRI, (j) Noncontrast axial T2 Thoracic Spine MRI: arrow shows edema of the dorsal cord at T11. MRI: Magnetic resonance imaging

deficits. The patient estimated that sensation improved about 70% at 2 months from injury.

Discussion

Spinal cord infarcts in adults are estimated at 3.1/100,000 persons, though are more rare in children with only small cohort estimates made postmortem with broad differential of both traumatic and atraumatic causes.[1-4] Traumatic causes of spinal cord infarction are often seen with high-impact injuries with resultant systemic hypotension, vasospasm secondary to traumatic hemorrhage, dissection, or expansile mediastinal or retroperitoneal hematomas causing hemodynamic compromise, subluxation, intervertebral disc herniation or stretching and fracture of the vertebral column.[3] Nontraumatic causes of spinal cord infarction include embolic, thrombotic, inflammatory, neoplastic, systemic hypotension with vasospasm without preceding trauma reversible hemodynamic compromise (i.e. vasospasm), external compression, vascular malformations, and Surfer's myelopathy.[4-9] Spinal cord infarcts most frequently present with weakness and numbness below the level of the lesion, neurogenic bladder/bowel with associated incontinence, and sexual dysfunction are common; however, cases of unilateral presentations have also been described.[3,10,11] Our patients had no significant trauma before their infarctions, so an atraumatic differential will be explored.

Surfer's myelopathy, also known as acute hyperextension myelopathy, is an ischemic spinal cord injury thought to be secondary to hyperextension of the back with vascular compromise of the cord.[4,9,11] The most common presenting symptom in Surfer's myelopathy is back pain, which occurs frequently after a first surfing lesson in young healthy persons.[4,9,11] Surfer's myelopathy has been suspected in other sports involving hyperextension of the spine, and case 1 likely hyperextended the spine due to his spike ball save. This could have predisposed him to vascular compromise through disc compression externally.[9,11] Additional risk factors for Surfer's myelopathy include long sedentary travel periods, putting patients at risk to develop deep venous thrombosis.[4,9,11]

FCE occurs when the annulus fibrosus or nucleus pulposus of intervertebral discs migrates into adjacent vasculature leading to infarct, often associated with a vascular malformation or minor trauma such as Valsalva maneuver.[7,12] FCE is increasingly recognized postmortem as a cause of spinal cord infarct through histopathology, although remains a diagnosis of exclusion.[12] Radiographic findings suggesting FCE include endplate herniations into the vertebral bodies known as Schmorl's nodes, annular fissure, intervertebral disc herniation, or other vertebral body abnormalities.[7,9,12] Schmorl's nodes in isolation are suggestive though not specific to FCE as they are present with any degenerative change to the spine.[7] Cases 1 and 2 demonstrated Schmorl's nodes, though case 1 additionally demonstrated intervertebral disc herniation on the MRI of the spine [Figure 1]. The angiogram for case 1 did not demonstrate a filling defect which is generally inconsistent with embolic etiologies except in the case of very small emboli which may pass through undetected, and, interestingly, he had an anatomic variant with two Arteries of Adamkiewicz, which has been previously reported, though only occurs in 11.3% of cases in a large meta-analysis of anatomic variants.[13]

Any case of atraumatic spinal cord stroke should also be evaluated for reversible treatable causes such as vasculopathy, hypercoagulability, coagulopathy, inflammatory cause, extrinsic compression amenable to surgery, infectious etiologies, or embolic sources. Workup should include vascular imaging and spine imaging with magnetic resonance, CTA, conventional angiogram, survey for clot burden, hypercoagulability testing, and cerebrospinal fluid analysis.[4-6,8] More subacute presentations should be worked up for demyelinating syndromes, infectious causes, vascular malformations, and neoplastic lesions.[6,8,10,14]

While there is no consensus for acute disease modifying pharmacotherapy for idiopathic spinal cord infarctions not amenable to surgical management or without comorbid treatable causes, permissive hypertension, hydration, antithrombotic therapy, and early and aggressive rehabilitation may impact the long-term varied and guarded prognosis, ranging from nearly complete recovery to minimal or no recovery.[4,5,7,11] More generally, tissue plasminogen activator (tPA) given intravenously and thrombectomy are rarely utilized within the permissible window in ischemic cerebral pediatric stroke due to difficulty with early recognition.[15] tPA and thrombectomy have been used in pediatric cerebral ischemic stroke.[4,11,15-17] Corticosteroids at a high dose with 30 mg/kg methylprednisolone intravenously followed by maintenance dosing of 5.4 mg/kg/h for 23 h have shown benefit for traumatic spinal cord injury previously in a randomized controlled trial.[17] Corticosteroid use, intravenous tPA, lumbar cerebrospinal drain, and volume expansion and pressor therapy to maintain mean arterial pressures over 85 mmHg in addition to supportive care has previously been suggested for Surfer's myelopathy.[4,11,17] In the presence of prothrombotic disorders or embolic stroke with measurable clot burden, there is consensus that anticoagulation is needed for secondary stroke prevention until the resolution of existing clot, though in the absence of detectable clot burden, daily aspirin generally dosed 1–5 mg/kg/day is sufficient.[4,6,11,18]

In summary, atraumatic and traumatic spinal cord infarctions are hyperacute or delayed onset lesions resulting in severe disability. To prevent the recurrence and extension of disease, extensive workups are required to identify surgical candidates, thrombophilia, inflammatory etiologies, and infectious causes. We present two cases of atraumatic spinal cord infarctions treated conservatively with diagnostic workup and antiplatelet therapy, though prompt diagnosis and exclusion of contraindications may result in treatment with additional therapeutics such as corticosteroids, tPA, volume expansion and pressor therapy, and surgical drainage can be considered.

Ethical statements

The research was approved by the Indiana University HRPP Internal Review Board on 10-18-2022 (protocol number 16995) and conformed to the ethical principles regarding human experimentation developed by the World Health Organization as outlined in the Declaration of Helsinki.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Informed consent

Before acquiring internal review board approval for this manuscript, verbal consent was acquired from the families of each case detailed in this manuscript over the phone.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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