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LETTER TO THE EDITOR

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Superficial Siderosis Related to a Thoracic Disc Herniation with Associated Dural Injury

Jaime Toro,[1,2,3] Camilo Díaz,[1] Saúl Reyes,[1,2] Valerie Jeanneret[3] & Lisseth E. Burbano[1]

1 Department of Neurology, Hospital Universitario Fundación Santa Fe de Bogotá, Bogotá, Colombia
2 School of Medicine, Universidad El Bosque, Bogotá, Colombia
3 School of Medicine, Universidad de Los Andes, Bogotá, Colombia

Correspondence
Jaime Toro, M.D., Asociación Médica de Los Andes Avenida 9 No. 116–20 Oficina 409, Bogotá, Colombia.
Tel.: +57-1-2150169;
Fax: +57-1-2150205;
E-mail: jtoro@uniandes.edu.co
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All the listed authors have contributed equally to the development of the manuscript.

Superficial siderosis (SS) of the central nervous system leads to gradual neurological deterioration characterized by sensorineural hearing loss, cerebellar ataxia, and myelopathy [1,2]. SS has a prevalence of approximately 0.7% in the general population [3]. Males appear to be affected more often than females by a 2:1 ratio, and the average age of presentation is between the 5th and 6th decades of life [1]. Chronic bleeding into the subarachnoid space can cause deposition of hemosiderin in neuronal tissues and the characteristic hypointensity seen on T2-weighted magnetic resonance imaging (MRI) [1,2]. Causes of bleeding include a past history of intradural surgery, head/back trauma, central nervous system tumors, arteriovenous malformation, nerve root avulsion, dural injury [1,2], and cerebral amyloid angiopathy [4]. SS has been rarely reported in association with dural defects caused by protruding intervertebral discs [5,6]. We present an unusual case of chronic pial siderosis related to a thoracic disc herniation with associated dural injury. Heightened suspicion and appropriate use of imaging can lead to diagnosis and prompt treatment of this condition.

A 66-year-old, right-handed man presented to our neurology clinic with a four-year history of leg weakness and progressive gait impairment. Symptoms progressed over time, and he started to use a cane 6 months previously. The patient also reported bilateral hearing loss and urinary incontinence. He denied tremor, fasciculations, back pain, falls, dysphagia, or dysarthria. His past medical history was otherwise unremarkable.

Neurological exam revealed bilateral sensorineural hypoacusis. Muscle strength was 5/5 in both upper extremities and 4/5 in the lower extremities. Deep tendon reflexes were mildly hyperactive in the lower extremities while normal in the upper limbs. The plantar reflex was extensor on the right and flexor on the left. Sensory examination was normal. The patient had a spastic–ataxic gait with bilateral foot drop.

Complete blood count, serum glucose, and electrolytes were normal. A lumbar puncture was performed with an opening pressure of 7 cm of water. Cerebrospinal fluid (CSF) was xanthochromic and had hyperproteinorachia (59.8 mg/dL [normal < 50 mg/dL]), 2 leukocytes/mm^3, 117 erythrocytes/mm^3, and a normal glucose. A brain MRI showed extensive hypointensity around the brainstem and along the cerebellar folia (Figure 1A) as well as within the cerebral fissures (Figure 1B). Cervical (Figure 1C) and thoracic (Figure 2A) MRIs also showed hypointensity along the entire pial surface of the spinal cord and a disc herniation at the T8–T9 level. Cerebral and spinal angiography showed no signs of extravasation of contrast agent representing bleeding. A dynamic CT myelogram was also performed and showed a ventral dural defect at the level of the thoracic disc herniation (Figure 2B). These findings were consistent with chronic pial siderosis related to a thoracic disc herniation with associated dural injury.

Physical therapy was started, and surgical repair of the dural defect was offered but declined by the patient. The patient has remained stable without increasing disability after 6 months of follow-up. Repeat MRI 6 months later showed persistence of hemosiderin deposits around central nervous system (CNS) structures.

The classic triad of sensorineural hearing loss, cerebellar ataxia, and myelopathy is present in 39% of cases [1]. Clinical findings

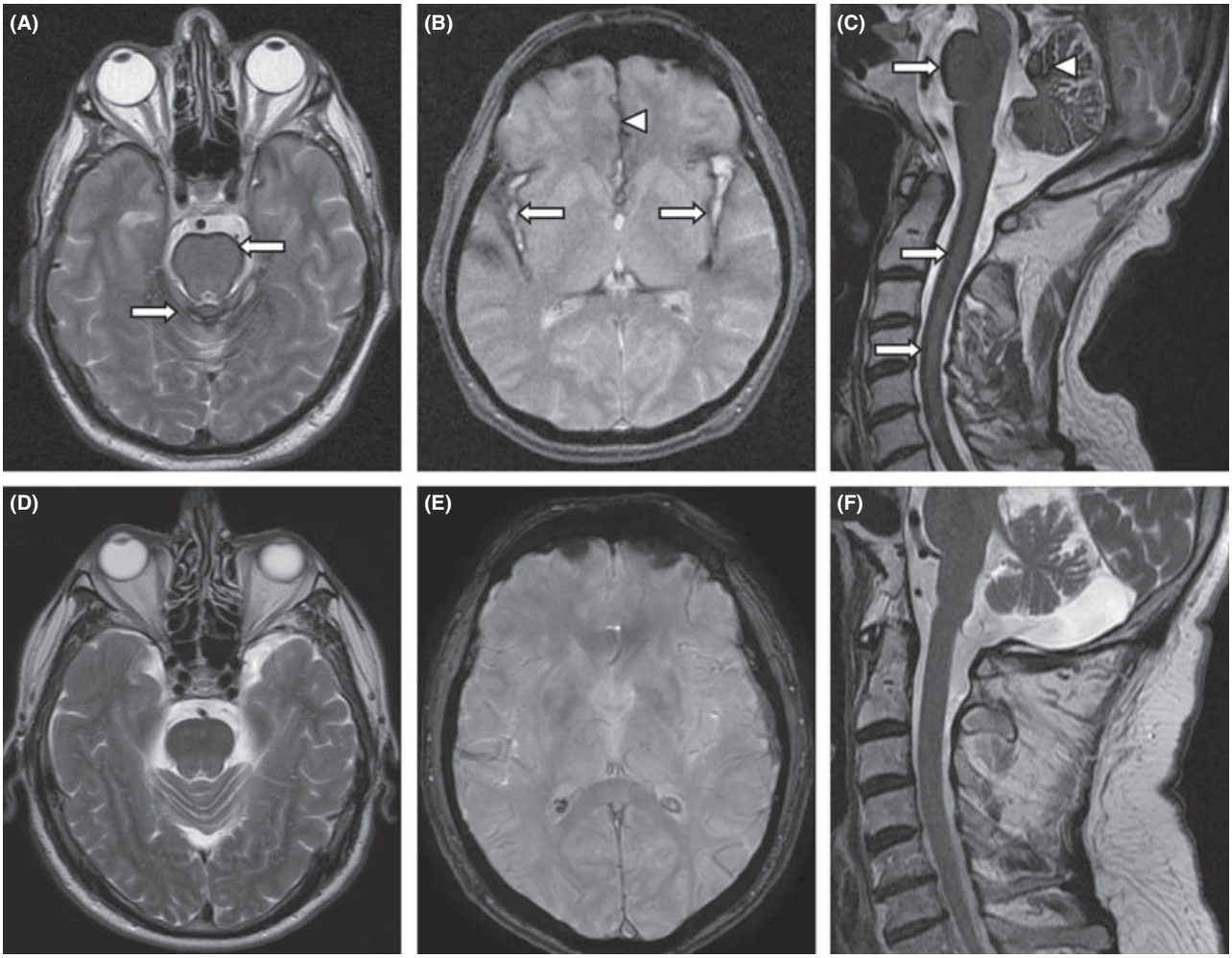

Figure 1 Brain and spinal cord MRI. (**A**) Axial T2-weighted brain MRI shows hemosiderin deposits around the pons and cerebellar folia (arrows). (**B**) Hypointensity along the interhemispheric (arrowhead) and Sylvian fissures (arrows) is best demonstrated by gradient-echo T2-weighted MR imaging. (**C**) Sagittal T2-weighted cervical MR imaging shows the characteristic low signal intensity along the surface of the brainstem, spinal cord (arrows), and cerebellum (arrowhead). (**D**) to (**F**) MRI images correspond to a 72-year-old male without SS or thoracic disc herniation, in which the absence of hemosiderin deposits is clear. (**D**) Axial T2-weighted brain MRI. (**E**) Gradient-echo T2-weighted MRI. (**F**) Sagittal T2-weighted cervical MRI.

such as hyposmia, nystagmus, cognitive impairment, and lower motor neuron signs are found less often [1]. In 2007, a review of 270 cases reported that SS was attributed to CNS tumors in 21% of cases, 13% to trauma, and 9% to vascular malformations. Other causes were history of neurosurgery (7%), brachial plexus injury (6%), and cerebral amyloid angiopathy (3%). Most cases remained idiopathic (35%) [1].

Dural injury is an important mechanism to explain the chronic bleeding into the subarachnoid space [3, 5]. Dural defects, as seen in our patient, can be caused by protruding discs, osteophytes, or disc-osteophyte complexes [5]. These defects are responsible for a CSF leak into the epidural space, which eventually produces a fluid-filled collection and CSF hypovolemia. It has been proposed that increasing pressure in the epidural space produces trauma of the internal venous plexus, which causes chronic microhemorrhage [7]. Blood products circulate back through the leakage point into the subarachnoid space, leading to hemosiderin deposition in the subpial layers of the CNS [5, 7].

Characteristic T2-weighted MRI findings of SS include a rim of hypointensity around the spinal cord, cerebellum, brainstem, and cortical fissures. Gradient-echo images have a higher sensitivity for the detection of hemosiderin deposits [2,8]. Dynamic CT myelogram and digital substraction myelogram are helpful in identifying dural defects, determining a potential site for surgery [7,8]. Angiographic studies are also useful for identifying any source of bleeding [8].

Cerebrospinal fluid analysis, although not necessary for establishing a diagnosis, can reveal xanthochromia, increased red blood cell count, hyperproteinorachia, erythrophages, and siderophages [1,5]. A low opening pressure can be found in patients with CSF leaks, as seen in our case [1,2].

The goal of treatment is to prevent progression of the condition by localizing and surgically repairing the bleeding source [2].

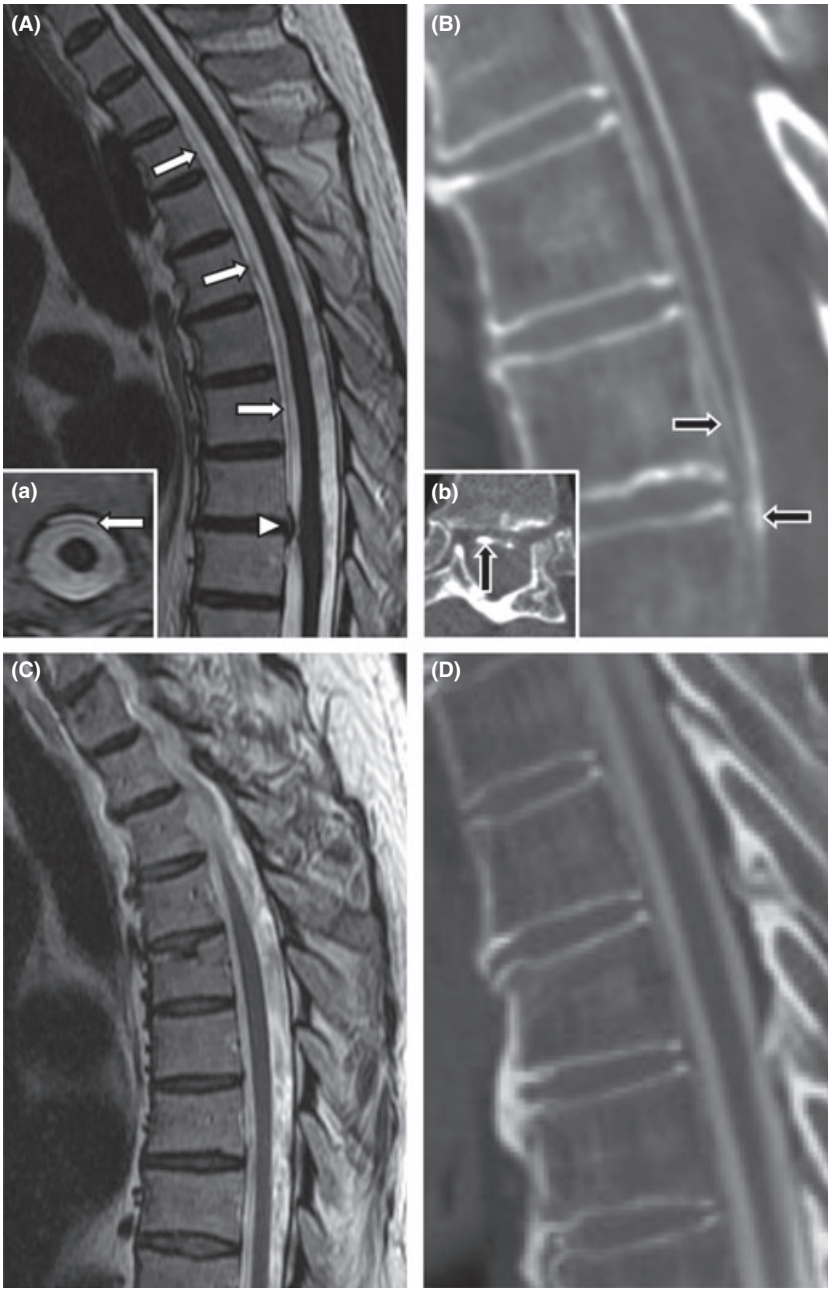

Figure 2 Thoracic MRI and dynamic CT myelogram. (**A**) Sagittal and axial (a) T2-weighted thoracic MR images show a thoracic epidural fluid-filled collection (arrows) and a disc herniation at T8–T9 level (arrowhead). Sagittal (**B**) and axial (b) dynamic CT myelogram shows a contrast leak (arrows) through a ventral dural defect at the level of the disc herniation. (**C**) T2-weighted thoracic spinal cord MRI in which the absence of either hemosiderin deposits or thoracic disc herniation is clear and (**D**) normal dynamic CT myelogram of a 71-year-old female.

Unfortunately, there is no known effective therapy that has been proven to reverse the effects of siderin deposits in the brain [2,9]. A pilot safety study of iron-chelator agents has been conducted, but further studies are needed to evaluate their efficacy and clinical benefit [9,10].

In conclusion, dural injury due to disc herniation should be considered as a cause of SS. Although advanced imaging may be needed to determine the source of bleeding and establish a therapeutic approach, a detailed history and careful neurological examination are essential in distinguishing common causes such as tumors and trauma. Therapy is tailored toward identification and treatment of the underlying cause.

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Conflict of Interest

The authors declare no conflict of interest.

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