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A patient with superficial siderosis, intraspinal cyst, low-pressure headache and low-lying cord

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Abstract

Superficial siderosis is a rare condition caused by deposition of hemosiderin in the central nervous system. In recent years, it has been used to identify dural defects connecting the intrathecal space with the intraspinal fluid-filled collection seen in patients with superficial siderosis. However, rare reports describe the association of low-lying cord in superficial siderosis patients.

We present a patient with progressive cerebellar ataxia, sensorineural hearing loss, low-pressure headache, pyramidal signs and additional cervical and lumbar radiculopathy with neurogenic bladder which might be related to cervical disc herniation and tethered cord. CSF and Magnetic resonance imaging (MRI) studies confirmed the diagnosis of superficial siderosis. Dural defect is indirectly suspected by existence of fluid-filled collection ventral to the spinal cord and low-pressure headache in our patient. Underlying causes of low-pressure headache and possible dural defect should be searched for as an associated and possibly treatable condition in superficial siderosis and provide further insights into the mechanism of bleeding in our patient who have an intraspinal fluid-filled collection.

Key words: Superficial siderosis; intraspinal cyst; low-pressure headache; low-lying cord.

Introduction

Superficial siderosis of central nervous system results from hemosiderin deposition in the subpial layers of the brain and spinal cord due to chronic subarachnoid hemorrhage. A history of previous intradural surgery or trauma is common. It is either due to dural defect pathology, a vascular tumor or a vascular abnormality. In recent years, an extra-arachnoid longitudinally extensive intraspinal fluid-filled collection has been frequently noted in patients with superficial siderosis (1, 2). Some reports have also noted an association between superficial siderosis and CSF hypovolemia and low-pressure headache (3, 4). We describe a patient of superficial siderosis with intraspinal cyst, low-pressure headache and additional imaging finding of low-lying cord.

Case report

This 55-year-old female was evaluated for a 2-year history of unsteady gait and decreased left hearing. She experienced a motor vehicle accident 10 years ago and had a sequel of backache with radicular pain from right thigh to lateral leg. Two years ago, she had bumped frontal head against the ground. After the accident, she began to suffer from intermittent headache and dizziness which was aggravated by upright position and alleviated by lying down. Her symptoms review was also remarkable for 3 months history of urinary urgency and frequency, slurred speech and episodes of right fingers numbness. The numbness was accompanied by radicular component with neck pain. The onset of these symptoms was gradual and the course was slowly progressive. Her examination was remarkable for sensorineural hearing loss in left ear, an ataxic dysarthria, increased lower limb tone, hyperreflexia, bilateral extensor plantar response and bilateral Hoffmann sign. She had a wide-based ataxic gait and was unable to walk on her toes and heel, or tandem. CSF study showed borderline-low open pressure (7 cm H_2O) and evidence of chronic hemorrhage (cell count: RBC: 520/ul, WBC: 0/ul; ferritin: 93.5 ug/L). Brain and spinal cord MRI (Fig. 1, 2) revealed hypointense coating along the bilateral sylvian fissures, tentorium, sulci of superior aspect of cerebellum, brainstem and whole spinal cord on the T2W gradient images, which was identified as

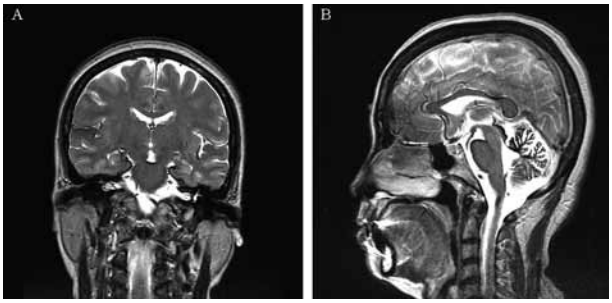

FIG. 1. — (A) Coronal T2-weighted MR image reveals marginal hypointensity involving the Sylvian fissure, interhemispheric fissure and basal cerebral surface cortical sulci. (B) Sagittal T2-weighted MR image shows diffuse hypointensities over the cerebellar folia, superior vermis, quadrigeminal plate and ventral pons with severe cerebellar atrophy.

hemosiderin deposition. Conus medullaris was placed at low end of L3 vertebral body with tethering of the filum terminale to dorsal dural sac without spinal dysraphism (Fig. 2B). Extradural fluid anterior to the spinal cord from C8 to T8 level was noted in spinal cord MRI (Fig. 2 C,D). Gadolinium-enhanced MRI of the spine (Fig. 2E) suggested mildly prominent vessels along the ventral aspect of the spinal cord, however, angiography was negative. With longitudinally extensive fluid collections, a CT myelogram may help localize the defect. We discussed with patient and her family for further treatment plan. They decided to receive chelation therapy with Deferiprone without further invasive study.

Discussion

The current patient represents typical triad signs of progressive cerebellar ataxia, sensorineural hearing loss, pyramidal signs and additional cervical and lumbar radiculopathy with neurogenic bladder which might be related to cervical disc herniation and tethered cord. A potential source of bleeding should be carefully investigated by imaging the entire neuraxis, including cranial and spinal cord MRI with enhancement and MR angiography. Further techniques such as dynamic CT myelogram or digital substraction myelography may be needed to identify the dural defect, but they are invasive techniques of time-intensive and high radiation expose (2). The patient refuses further invasive investigation and surgical management of epidural fluid accumulation.

Orthostatic headache and borderline-low open CSF pressure supported the diagnosis of craniospinal hypovolemia or intracranial hypotension. Some similarities between craniospinal hypovolemia and superficial siderosis with intraspinal fluid collection were speculated by Kumar *et al.* (3). Dura tear is indirectly suspected by existence of fluid-filled collection ventral to the spinal cord and low-pressure headache in our patient. The calcified disc protrusion and trauma history could have caused the dural defect and is most commonly near the cervicothoracic junction or in the mid to upper thoracic spine by previous reports (1, 5, 6). Bleeding source might origin from dural tear with friable vessels, though the mechanism is still not very clear. Tethered cord

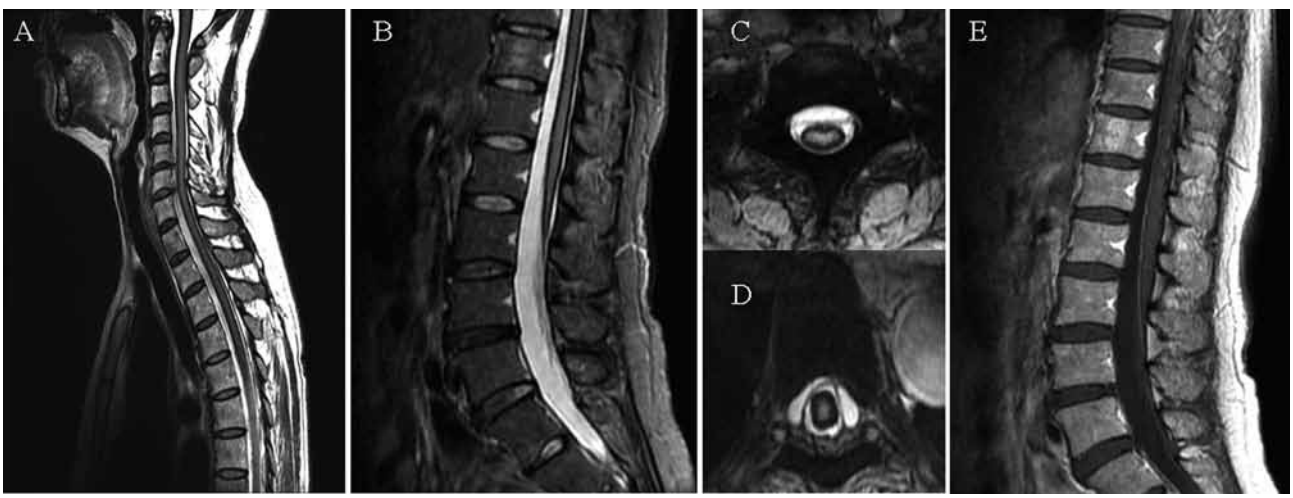

FIG. 2. — Marginal hypointensity around whole spinal cord is significantly noted in T2 weighted MR image. (A) Sagittal T2-weighted MR image shows disc protrusion between C5/C6 and C6/C7 (B) Sagittal T2-weighted MR image shows low tethered cord with conus medullaris placed at L3. (C, D) Axial T2-weighted thoracic cord MR image respectively shows a fluid-filled collection anterior to the spinal cord at C8 and at T8. (E) Sagittal T2-weighted image of the lumbar spine with contrast enhancement showing leptomeningeal enhancement, possibly due to prominent pial vascularity.

syndrome is less common causes of superficial siderosis. Vera et al reported a superficial cerebral and spinal hemosiderosis caused by secondary tethered cord syndrome after resection of a spinal lymphoma (7). Spinal cord tethering with low-placed conus medullaris in our patient might be either related to possible arachnoiditis (1) or developmental defect. We suppose that spinal cord tethering reaches a threshold level of tension under the precipitation of cervical disc herniation or traction the root. This causes a reduction in spinal cord blood flow and dysfunction of neuronal mitochondrial terminal oxidase which increases burdens of iron toxicity and oxidative damage after episodic nature of subarachnoid hemorrhage from dura tear and trauma (8, 9).

As our best knowledge, there are no similar reports about low-lying cord in superficial siderosis patients. The patient reported here is a representative example of the typical clinical history of superficial siderosis and the association with low-pressure headache, low-lying cord and intraspinal fluid collection raises an interesting question about pathological mechanisms in this rare disorder.

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