Unusual neuroimaging in superficial siderosis

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Superficial CNS siderosis from bleeding into the subarachnoid space causes sensorineural hearing loss and cerebellar ataxia.\(^1\) Spinal origins of superficial siderosis are rare but can include nerve root pathology.\(^2\)

A 42-year-old man presented with an 8-year history of gait difficulty, dysarthria, and hearing loss. Past history was remarkable for a motor vehicle accident at age 10. Abnormalities on neurologic examination included decreased hearing, dysarthria, positive Babinski, and a wide-based gait. Head MRI demonstrated vermian atrophy and hypointense signal along the cerebellar surface and brainstem (figure, A). Postgadolinium MR revealed prominent vessels along the ventral brainstem. Cerebral angiography was negative. MRI of the spine revealed pial siderosis of the cord and cauda with peripheralization of roots suggesting arachnoiditis. Spine MRI showed a nonenhancing fluid collection ventral to the cervical cord (figure, B), tethered cord at T9 (figure, C), and enhancing vessels ventral to the cervical cord (figure, D). Invasive spinal angiography was negative. Surgery was performed with the intent of releasing a cord herniation at T9 but no dural defect was found. A left-sided nerve root avulsion was identified at T10.

Cerebrospinal fluid examination done after the surgery showed persisting subarachnoid hemorrhage. A dynamic CT myelogram identified a dural defect communicating with a fluid-filled cavity at T1–2. This defect was successfully repaired and a subsequent CSF study showed no red blood cells. The extensive siderosis may have contributed to sclerosis of the epidural plexus of veins, leading to venous hypertension.

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