

Superficial Siderosis of the Central Nervous System Due to Bilateral Jugular Vein Thrombosis

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Objective: To describe a novel cause of meningeal siderosis due to intermittent subarachnoid bleeding caused by chronic bilateral jugular vein thrombosis.

Design: Case report and review of literature.

Patient: A 51-year-old man with a distant history of cervical injury who presented with transient aphasia in the setting of progressive cognitive decline.

Intervention: Neurological examination, magnetic resonance imaging, lumbar puncture, and angiogram.

Results: The patient had intermittent subarachnoid bleeding resulting from extensive venous collaterals in the neck and cervical spine due to chronic bilateral jugular vein thrombosis.

Conclusion: Unexplained neurological deterioration and history of cervical trauma warrants diagnostic consideration of superficial siderosis and jugular vein thrombosis.

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SUPERFICIAL SIDEROSIS OF THE central nervous system, an increasingly recognized cause of insidious neurological deterioration, results from hemosiderin deposition in the leptomeninges, subpial layers of brain, and spinal cord following recurrent subarachnoid bleeding. Patients with superficial siderosis most frequently present with sensorineural deafness, ataxia, myelopathy, pyramidal signs, and cognitive impairment.^{1,2} The source of subarachnoid hemorrhage is not identified in almost half of the patients. Repeated intrathecal bleeding is generally caused by intradural surgery, dural injury following cervical root avulsion, neoplasia, and vascular lesions. We describe a novel cause of meningeal siderosis due to intermittent subarachnoid bleeding from chronic bilateral jugular vein thrombosis.

REPORT OF A CASE

A 51-year-old man was admitted for transient aphasia and dizziness. During the 10 years prior to admission, he had slowly progressive cognitive decline with increasing disorientation, slowness of thought, and inability to cope with multiple tasks at work. The patient also had hearing loss, balance difficulties, and decreased sense of smell for several months. His medical history was notable for a recent diagnosis of vitamin B₁₂ deficiency, history of smok-

ing, and alcohol use. He also had a remote history of C2 fracture 33 years prior due to a motor vehicle accident—as a consequence, the patient was paralyzed for 2 days and in cervical traction for 4 months with complete recovery. At presentation, his neurologic examination revealed slowness of speech, mixed aphasia, apraxia, broad-based gait, mild ataxia, and hyperreflexia with bilateral extensor plantar responses. An electroencephalogram showed left temporal slowing. Initial magnetic resonance imaging showed cerebellar atrophy and T2 hypointensity along the surface of the brain (including the spinal cord, brainstem, cerebellum, and cerebrum) most prominent on susceptibility sequences consistent with meningeal siderosis (**Figure 1**). Magnetic resonance imaging of the spine showed prominent venous engorgement involving the ventral aspect of the brainstem and cervical cord with no intrinsic cord abnormalities (**Figure 2**). The patient started taking phenytoin for suspicion of focal left temporal seizures and nimodipine for prophylaxis for vasospasm, and he did not have any further episodes of aphasia. A lumbar puncture showed opening pressure of 10 mm Hg; xanthochromia; total protein, 0.107 g/dL (to convert to grams per liter, multiply by 10.0); glucose, 84 mg/dL (to convert to millimoles per liter, multiply by 0.0555); white blood cell count, 2 to 3/cm³ (100% neutrophils); and red blood cell count, 383/cm³, which did

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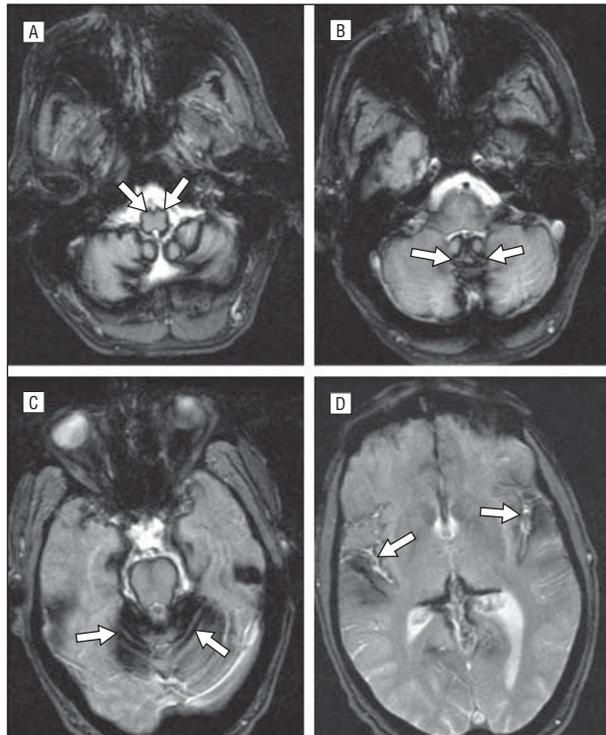


Figure 1. Magnetic resonance imaging susceptibility sequences (axial) showing diffuse hemosiderosis (low signal, arrows) throughout the spinal and cerebral meninges, especially prominent around brainstem (A), cerebellar folia (B and C), and sylvian fissures (D).

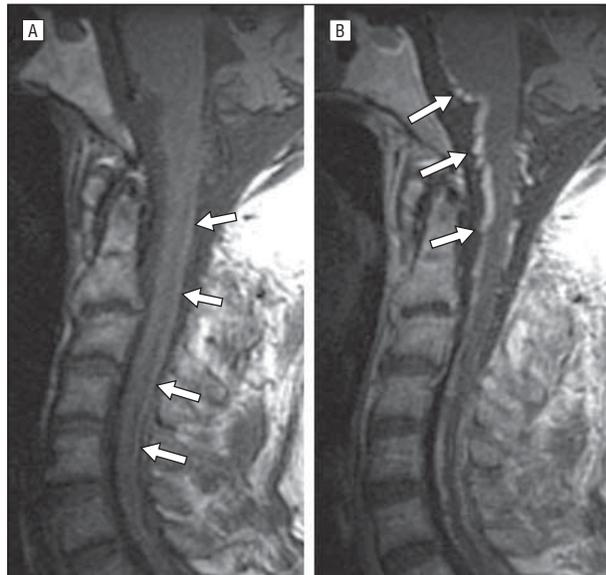


Figure 2. Magnetic resonance imaging of the cervical spine showing abnormal, high-intensity signal around the whole spinal cord (A; arrows, sagittal T1 precontrast) and abnormal vessel engorgement anterior to the cervical spine (B; arrows, sagittal T1 postcontrast).

not clear. Gram stain and culture did not reveal any infection. Cerebral and spinal angiograms were then performed and showed no angiographic evidence of arterial venous malformation, fistula, or aneurysm. However, there was bilateral venous occlusion at the level of the jugular bulbs (**Figure 3**, A and B). Below the jugular bulbs, there was an extensive venous collateral network

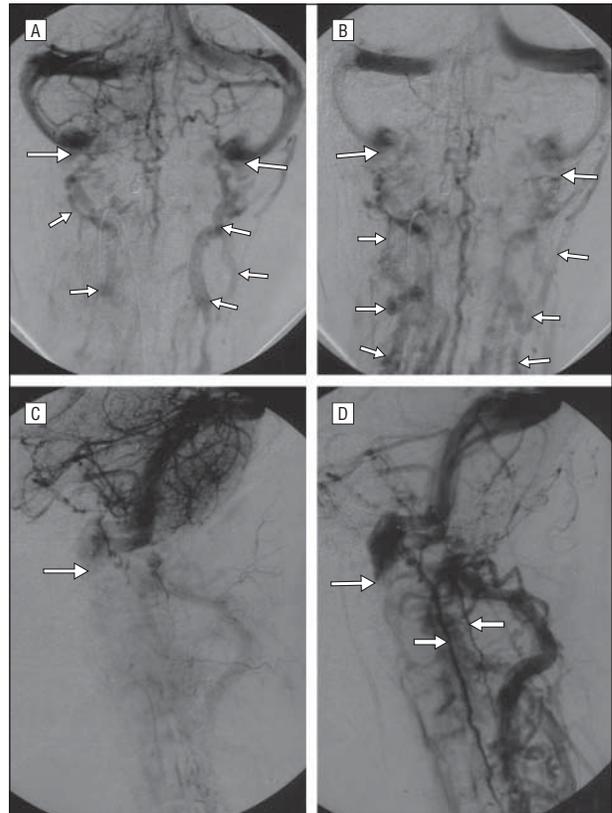


Figure 3. Cerebral and spinal angiogram. Early (A) and late (B) anterior-posterior and early (C) and late (D) lateral projections show bilateral occlusion of internal jugular veins at the level of the jugular bulbs (large arrows) with dilatation of surrounding cervical veins (A and B, small arrows) which now serve as venous outflow. D, Small arrows indicate severely dilated anterior and posterior spinal veins.

in the neck that included engorged anterior and posterior spinal veins (Figure 3, C and D). The patient had a tagged (Tc-99M) red blood cell study that did not reveal a source of active bleeding. A chest computed tomogram did not show any evidence of malignancy or superior vena cava syndrome. A repeated lumbar puncture 2 weeks later showed pressure within the reference range; protein, 0.98 g/dL; glucose, 90 mg/dL; and no xanthochromia. Results of laboratory evaluation were normal including negative anti-Purkinje cell, antineuronal nuclear, lupus anticoagulant, anticardiolipin antibodies, and activated protein C resistance screening. The homocysteine level was slightly elevated at 1.93 mg/L (to convert to micromoles per liter, multiply by 7.397). No active source of bleeding was found, and the patient was discharged while taking phenytoin and nimodipine. Owing to the chronicity of the jugular vein thrombosis and the extensive venous collateralization, no specific treatment was recommended. He continued to be neurologically stable without recurrence of seizures during 8 months of follow-up.

COMMENT

Superficial siderosis of the central nervous system (CNS) is a progressive disorder associated with recurrent sub-arachnoid bleeding, leading to deposition of blood deriva-

tives along the leptomeninges, spinal cord, and cranial nerves.²⁻⁴ Superficial siderosis is being increasingly recognized as a cause of insidious neurological deterioration.^{5,6} However, the source of subarachnoid hemorrhage remains unidentified in up to 50% of patients. Chronic hemorrhage is often the result of trauma (root avulsions), dural surgical procedures, cystic lesions (eg, meningoceles, pseudomeningoceles), tumors (eg, ependymomas, meningiomas, oligodendrogliomas), and vascular lesions (eg, aneurysms, arteriovenous malformations). Undetected subarachnoid hemorrhage may also occur owing to microscopic venous or capillary leakage, especially while receiving anticoagulant treatment. As far as the authors are aware, this is the first reported patient with superficial siderosis due to enlarged intrathecal venous collaterals from bilateral jugular venous thrombosis (JVT).

The most common contemporary cause of JVT is jugular venous cannulation, either iatrogenic or related to patient intravenous drug use.⁷ Other rare causes of JVT have been described: dehydration and marasmus, metastatic obstruction, intravenous hyperalimentation via indwelling venous catheters in the subclavian position, hemoglobinopathies, occlusion of the lateral sinuses, and infection (eg, pharyngeal abscess), and the hypercoagulable state associated with autoimmune disease and malignancy.⁷⁻¹² Although posttraumatic dural sinus thrombosis is not uncommon, we are unable to identify a reference in the literature to posttraumatic JVT.

Our patient developed a number of symptoms including sensorineural deafness, cognitive impairment, reduced sense of smell, dizziness, and balance difficulties during a 10-year period. All of these symptoms were consistent with previously described, slowly progressive signs of superficial siderosis. Our patient experienced headaches as well. Headache has been described as a frequent presentation of superficial siderosis.¹³ Additionally, our patient presented with aphasia, which has not been described as a symptom of superficial siderosis. Slowing over the left temporal lobe on electroencephalogram led us to suspect focal temporal seizures as the cause of the aphasia that resolved with the administration of phenytoin. Seizures have only been described in 2 patients,^{4,13} one of whom was described as having focal seizures. Sensorineural deafness has been found to be the leading manifestation of cranial nerve deficit and has been described in about 95% of patients.¹⁴ It may lead to deafness in 1 to 12 years.

The neurological deterioration occurs slowly over many years or decades, as in our patient, and therefore may not be linked with the inciting injury.¹⁴ The mean age at presentation of symptoms is 50 years (range, 14-77 years).⁵ The latency period between the causative event and the presentation of symptoms may vary from 8 to 22 years (mean, 8.7 years) with the longest period ever reported being 36 years. In our patient, there was a distant history of C2 fracture followed by prolonged cervical traction in bed 33 years before presentation and 23 years prior to onset of symptoms. This was likely the inciting event that led to bilateral JVT.

Early surgical intervention to stop the source of bleeding is crucial in halting disease progression. Surgical coagulation of the bleeding site is thus far the only means of treating superficial siderosis with proven effective-

ness. However, the source of bleeding remains undetected in nearly half of patients, most likely owing to the intermittent and slow nature of bleeding.

Superficial siderosis is becoming increasingly recognized as a cause of unexplained progressive neurological deterioration. Premorbid diagnosis is possible with magnetic resonance imaging. If possible, repair of a bleeding site may prevent further neurological decline. We present an unusual case with superficial siderosis resulting from cervical cord venous collateralization due to bilateral jugular vein thrombosis from a distant history of C2 fracture. Diagnostic consideration of superficial siderosis is warranted in patients with insidious neurological decline and a history of cervical trauma.

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