Surgical treatment of superficial siderosis associated with a spinal arteriovenous malformation

Case report

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SUPERFICIAL siderosis of the central nervous system (CNS) is characterized by the deposition of hemosiderin in the leptomeninges and subpial layers of the brain and spinal cord. Sensorineural hearing loss and ataxia are the main clinical manifestations of superficial siderosis and are present in at least 90% of cases. Other features include dementia, myelopathy, and various cranial nerve disorders, such as anosmia, oculomotor nerve palsies, and facial nerve palsy. Superficial siderosis is caused by chronic bleeding into the subarachnoid space, but the source of the hemorrhage is identified in only approximately 60% of cases. In the remaining patients, the superficial siderosis is considered to be idiopathic. We report on a patient who presented with superficial siderosis and a spinal arteriovenous malformation (AVM) that had been diagnosed on angiographic studies, in whom extensive examinations including spinal magnetic resonance (MR) imaging and myelography studies were normal. Obliteration of spinal AVMs may successfully prevent the progression of superficial siderosis.

KEY WORDS • arteriovenous malformation • spinal angiography • subarachnoid hemorrhage • superficial siderosis

In many patients with superficial siderosis of the central nervous system (CNS) no source of bleeding can be established, despite extensive examinations. The authors report a patient with superficial siderosis and a spinal arteriovenous malformation (AVM) that was not visible on magnetic resonance (MR) imaging or myelography but was identified on angiographic studies.

This 71-year-old man presented with a 2-year history of progressive gait difficulties and hearing loss. Examination showed ataxia, hearing loss, and quadriparesis. On MR imaging superficial siderosis of the brain and spinal cord was seen; however, MR imaging of the CNS, as well as cerebral angiography and myelography studies, did not reveal the source of hemorrhage. Spinal angiography revealed a small slow-flow pial AVM at the C-5 level originating from the anterior spinal artery. A C-5 corpectomy was performed and the AVM was obliterated. The patient did well and reported no further progression of his symptoms during 3 months of follow up.

Spinal angiography is indicated to complete the evaluation of patients with superficial siderosis, even if results of spinal MR imaging and myelography studies are normal. Obliteration of spinal AVMs may successfully prevent the progression of superficial siderosis.

Case Report

History. This 71-year-old man presented to a tertiary care center at another institution with a 2-year history of progressive gait difficulties, hearing loss, dizziness, and ringing in the right ear. There was no history of trauma, and the patient did not complain of any neck or back pain. Examination revealed bilateral neurosensory hearing loss, truncal and appendicular ataxia, and a mild spastic quadriparesis. On MR imaging of the brain extensive superficial siderosis was found (Fig. 1). A lumbar puncture was performed and examination of cerebrospinal fluid showed xanthochromia, 9108 erythrocytes/mm³, 15 nu-
cleated cells/mm³, and a total protein level of 107 mg/dl. On MR imaging of the entire CNS, with and without gadolinium enhancement, four-vessel cerebral angiography, and computerized tomography myelography, the results were normal and no source of hemorrhage was identified. A diagnosis of idiopathic superficial siderosis of the CNS was made. The patient sought another opinion.

Examination. Our neurological examination of the patient confirmed the previous observations, but we recommended spinal angiography to complete his evaluation. This study showed a very small, slow-flow vascular malformation adjacent to the anterior spinal artery at the C-5 level (Fig. 2). The abnormality was only visible on the anteroposterior projections and could not be detected on the oblique and lateral films. On careful review of the sagittal and axial MR images of the cervical spinal cord and the myelogram we did not detect any abnormality.

Operation. A C-5 corpectomy and C4–5 and C5–6 discectomies were performed. The dura was opened and a small pial AVM was identified arising from the anterior spinal artery. The main draining vein had an area of stenosis between two areas of venous dilation. The anterior surface of the spinal cord showed intense xanthochromic discoloration. The abnormal fistulous connections were coagulated, and the corpectomy defect was reconstructed with a fibular allograft and an anterior cervical plate.

Postoperative Course. The patient made an uneventful recovery. An angiogram obtained postoperatively demonstrated complete obliteration of the AVM. During 3 months of follow up the patient has not noted any further progression of symptoms.

Discussion

Any disease process along the neuraxis that causes repeated episodes of minor bleeding into the subarachnoid space can induce superficial siderosis. The most commonly identified causes of superficial siderosis are: 1) tumors, particularly ependymomas; 2) vascular anomalies, including AVMs and cavernous angiomas; and 3) various dural abnormalities, such as cervical root avulsions and pseudomeningoceles. Spinal AVMs have been identified only rarely as the source of hemorrhage in patients with superficial siderosis.

Superficial siderosis is a progressive disease for which no effective medical treatment is available. Therefore, evaluation for an underlying, potentially treatable, cause should be thorough. The initial study of choice is MR imaging of the brain and spinal cord. If these studies fail to reveal a source, cerebral angiography is indicated. In addition, myelography is often performed to rule out the presence of a spinal AVM, as it was in our patient. Finally, spinal angiography is advocated to complete the evaluation of a patient with superficial siderosis, although the yield of this study is reported to be very low. In fact, we have not been able to find any report in the literature in which spinal angiography was the only diagnostic study that revealed the source of the hemorrhage in patients with superficial siderosis. However, in our patient it is clearly demonstrated that spinal angiography is indicated in the evaluation of superficial siderosis, even if spinal MR imaging and myelographic results are normal. The spinal AVM in our patient was very small, had slow flow, and could only be demonstrated on a single angiographic pro-

FIG. 1. Axial T₂-weighted MR image obtained in a 71-year-old man, demonstrating a hypointense rim of hemosiderin deposition surrounding the mesencephalic, cerebellar folia, and sylvian fissures.

FIG. 2. Anteroposterior left vertebral artery angiogram revealing the anterior spinal artery (straight arrow) and contrast material entering an abnormal draining vein (curved arrow) at the C-5 level.

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jection. These factors may explain the apparent low yield of spinal angiography in the diagnosis of causes of superficial siderosis.

References


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