

SURGICAL RESECTION OF A CEREBRAL ARTERIOVENOUS MALFORMATION FOR TREATMENT OF SUPERFICIAL SIDEROSIS: CASE REPORT

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BACKGROUND

To our knowledge, there are only two reported cases of cerebral arteriovenous malformation associated with superficial siderosis. In both cases, the patients were asymptomatic and were discovered on retrospective review of magnetic resonance imaging.

CASE DESCRIPTION

We describe a case of superficial siderosis in a 47-year-old male with a remote history of closed head injury that presented with progressive hearing loss, cerebellar ataxia, and urinary incontinence. Lumbar puncture was indicative of active subarachnoid bleeding. Cerebral angiography revealed a small vascular malformation that was resected.

RESULTS

Pathological examination confirmed the diagnosis of the arteriovenous malformation. Six months after surgery the patient is neurologically stable with no further progression of clinical signs or symptoms.

CONCLUSIONS

We report the first case of surgical resection of an intracranial arteriovenous malformation for the treatment of superficial siderosis. We emphasize the necessity of a detailed evaluation when superficial siderosis is suspected to localize and resect potential bleeding sources, because the disease is progressive and often irreversible.
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KEY WORDS

Arteriovenous malformation, subarachnoid hemorrhage, superficial siderosis.

Superficial siderosis is a rare clinical syndrome that develops from recurrent subarachnoid hemorrhage. In cases where a source of bleeding has been identified, dural abnormalities (47%) and tumors (35%) are the most commonly associated

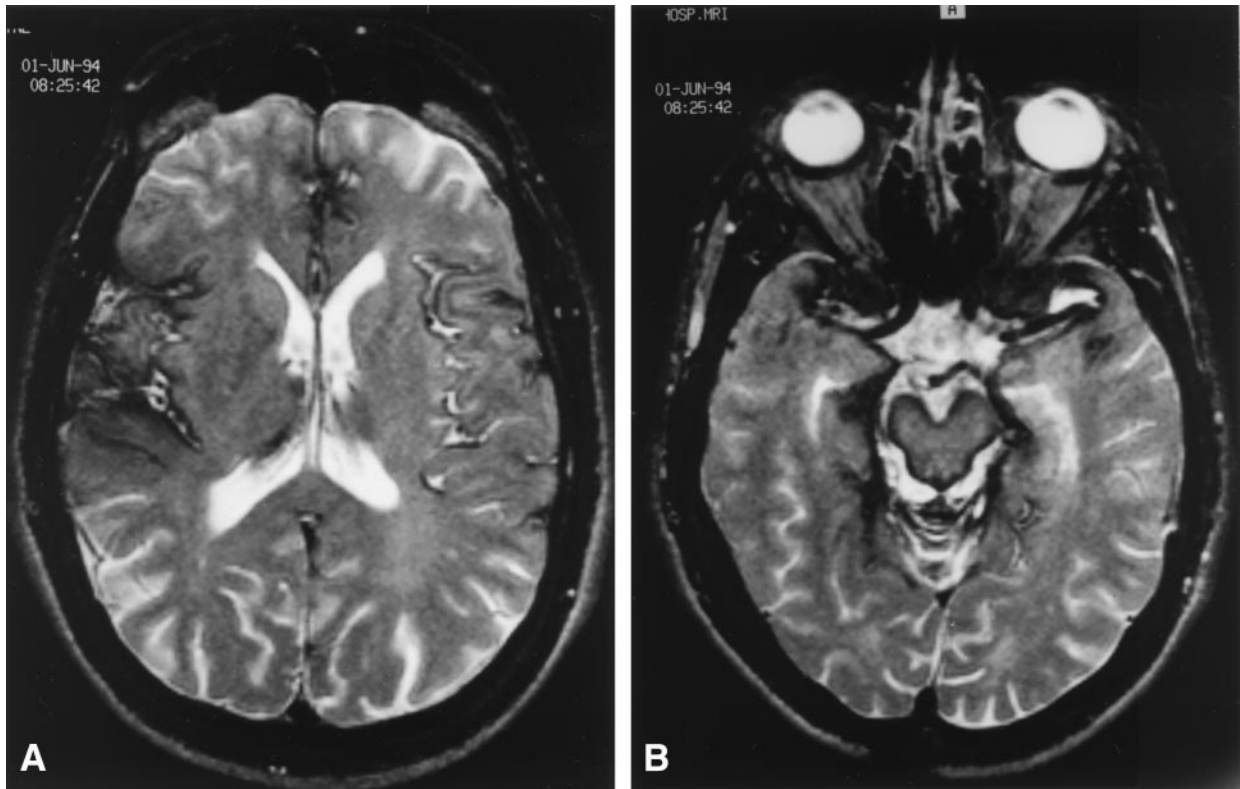
pathology [4]. Less often, vascular malformations (18%) have been found to be the source of hemorrhage [4-8,10,14,15]. One report in the literature briefly mentions two cases of cerebral arteriovenous malformations (AVM) associated with superficial siderosis [12]. In both cases, the patients had no symptoms or signs of the clinical syndrome and were diagnosed by a retrospective review of magnetic resonance imaging (MRI) studies.

We report a case of superficial siderosis in a middle-aged man who presented with progressive hearing loss, cerebellar ataxia, pyramidal signs, and urinary incontinence. Characteristic hemosiderin deposition on magnetic resonance imaging confirmed the diagnosis. Cerebral angiography identified a small right frontal vascular malformation, which was surgically resected. To our knowledge, this is the first report of surgical resection of a cerebral vascular malformation for treatment of superficial siderosis.

CASE REPORT

A 47-year-old man presented to our institution with an 8-year history of progressive hearing loss and a 5-year history of progressive gait difficulty and urinary incontinence. Past medical history was significant for severe closed head injury from a motor vehicle accident at 18 years of age that kept him hospitalized for several months. At that time, the patient had loss of consciousness and complex orthopedic trauma complicated by a left ulnar neuropathy. No intracranial or spinal procedures were performed. Initial evaluation of the patient's hearing loss at a tertiary care center did not reveal the etiology of the patient's symptoms. Further diagnostic studies between 1994 and 1996 included MRI of the brain and spinal cord that showed an incidental small cervical syrinx at C6-7, enlargement of

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1 Axial T2-weighted MR images at the level of the lateral ventricles (A) and midbrain (B) showing extensive hypointensity around the brainstem, cerebellar folia, and sylvian fissure, which represents superficial siderosis.

the fourth ventricle, and hypoplastic cerebellar tonsils. Moreover, the T2-weighted MRI of the brain in 1994 revealed superficial hypointensity along the brainstem, cerebellum, and Sylvian fissures, which was consistent with superficial siderosis (Figure 1A, B). The superficial siderosis was thought to be a result of the patient's previous trauma and no further diagnostic evaluation or intervention was pursued. In 1998, the patient was referred to our institution for evaluation of his myelopathy.

EXAMINATION

Neurological examination revealed bilateral ptosis, miosis on the left, and bilateral hearing loss, left worse than right. In addition, there was loss of sensation in the lower cervical and upper thoracic dermatomes, prominent gait ataxia, and hyperreflexia of the lower extremities. A repeat MRI of the brain and spinal cord revealed extensive hemosiderin deposition in the sylvian fissures and along the pial surface of the brainstem, cerebellum, and spinal cord. The previously noted cervical syrinx at C6-7 was unchanged (Figure 2). A diagnosis of superficial siderosis was entertained and further diagnostic evaluation included cerebral angiography, which revealed a small vascular malformation in

the posterior aspect of the right frontal lobe (Figure 3). A CT myelogram and a detailed spinal angiogram excluded other potential sources of bleeding (spinal AVM, nerve root avulsion). Lumbar puncture revealed xanthochromic CSF with 2000 erythrocytes/mm³ and an elevated ferritin level. On the basis of these studies, the patient was diagnosed with superficial siderosis, secondary to recurrent bleeding of a cerebral vascular malformation.

OPERATION

Preoperatively, stereotactic cerebral angiography was used to localize the lesion. A right frontal craniotomy was performed over the lesion and revealed thickened dura, which was reflected. A xanthochromic arachnoid membrane lay over the lesion. Two arterial feeders entered the lesion on its lateral aspect and converged into one large distal trunk. On its medial aspect a single draining vein ran superiorly into the sagittal sinus. After careful dissection, the feeding and draining vessels were ligated and the lesion was resected in a core of tissue measuring 2 × 1.5 × 1.5 cm. Intraoperative angiography confirmed the complete obliteration of the lesion. Surgical pathology confirmed the diagnosis of arteriovenous malformation.



2 Sagittal T2-weighted MR image of the cervical spinal cord showing a hypointense rim of hemosiderin around the spinal cord. Note the atrophy of the cerebellar folia and the hyperintense syrinx at C6-7 (arrow).

POSTOPERATIVE COURSE

The patient had no complications postoperatively. At 6 months after surgery, the patient has had no further progression of clinical signs or symptoms. Subjectively, the patient feels that his symptoms are improving.

DISCUSSION

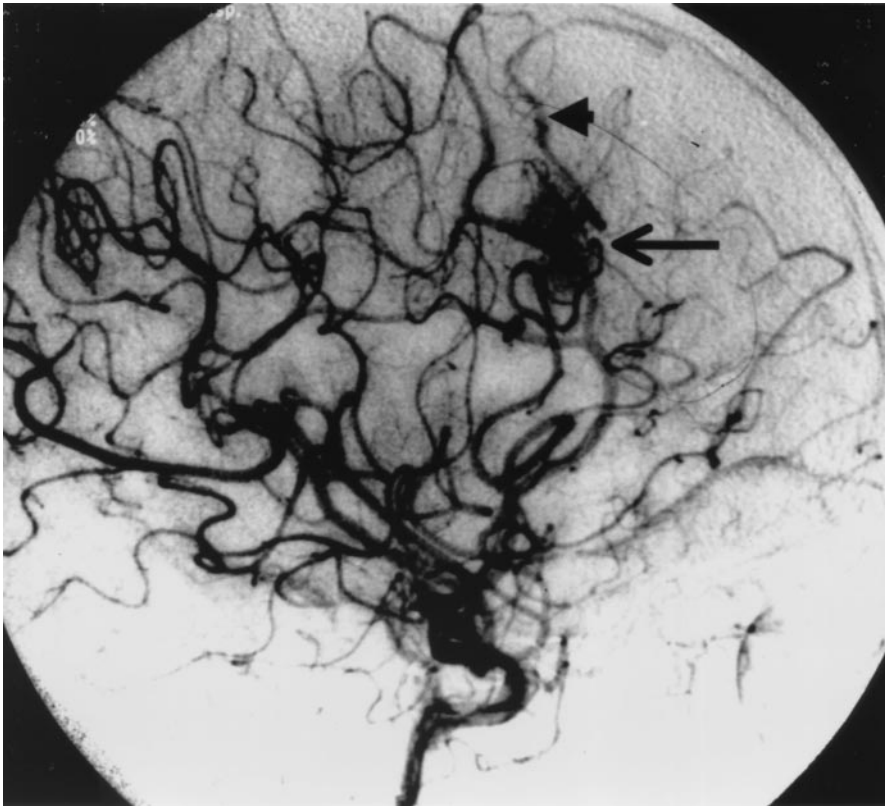
Superficial siderosis is caused by recurrent bleeding into the subarachnoid space that results in the deposition of hemosiderin in the leptomeninges and subpial layers of the brain and spinal cord. A source of bleeding is found in more than half of all cases [4]. The syndrome is characterized by the clinical triad of sensorineural deafness (95%), cerebellar ataxia (88%), and pyramidal signs (76%) [4,13]. Less common findings include dementia, bladder dysfunction, anosmia, and sensory deficits [4]. Although our patient's history of hearing loss, ataxia, and urinary incontinence is classic for the syndrome, the long interval between initial onset of symptoms in 1990 and the establishment of a diagnosis in 1998 emphasizes the slow progression of the disease and its insidious nature. More-

over, some of the patient's clinical findings were attributed to a remote history of trauma from a motor vehicle accident, such as the cervical syrinx that may have caused the suspended sensory levels at C7-T4.

The diagnosis of superficial siderosis is most easily confirmed by MRI. In our patient, a T2-weighted image revealed the presence of a hypointense rim along the pial surface of the brainstem, cerebellum, and spinal cord, that was consistent with the deposition of hemosiderin. This led to cerebral angiography and the localization of the source of bleeding, a right frontal vascular malformation. Because of the patient's history of trauma and the rarity of cerebral vascular malformations associated with superficial siderosis, the patient underwent computed tomographic (CT) myelography and complete spinal angiography to exclude other sources of bleeding. There have been several reports in the literature of superficial siderosis associated with spinal AVMs [5,14,15] and traumatic dural injury from nerve root avulsions [2,17].

An aggressive approach to the localization of potential sources of bleeding is warranted for several reasons. First, the neurological deficits of superficial siderosis are progressive and often irreversible [4]. Second, there is no effective medical therapy currently available for the treatment of superficial siderosis [1,8,13,16]. Desferrioxamine, an iron chelator, and trientine, a copper chelator, have been used in the treatment of superficial siderosis. Unfortunately, their administration over varying time periods did little to improve symptoms [4,8,13,16]. Third, surgical ablation of a potential bleeding source may arrest intermittent subarachnoid hemorrhage [17] and normalize the CSF profile [9]. Although our patient has a remote history of trauma, the results of the lumbar puncture and evidence of recurrent bleeding from the cerebral vascular malformation as seen at surgery confirm that this AVM was the reason for the patient's superficial siderosis. We recommended surgery as the most likely therapeutic intervention capable of preventing further decline in the patient's neurological status.

To our knowledge, there only are 11 reported cases of aneurysm or vascular malformation causing superficial siderosis in the world literature (Table 1). In three of these cases, the patients had no clinical symptoms [7,11] and in a fourth case insufficient data were presented to confirm the diagnosis [3]. The remaining seven cases of symptomatic superficial siderosis are comprised of three patients with spinal AVMs (two cervical, one thoracic), three cases of periventricular venous malformations (two third ventricle, one fourth ventricle), and one case with an anterior communicating artery aneurysm.



3 A lateral angiogram after injection of the right internal carotid artery revealing a posterior frontal vascular malformation (arrow) with a draining vein to the sagittal sinus (arrowhead).

Given the propensity of many CNS lesions to bleed, it is surprising that symptomatic superficial siderosis is not more common. The rarity of symptomatic superficial siderosis is emphasized by Offenbacher's radiographic review of 8843 consecutive head MRIs covering a seven-year time period. In that study, only 13 patients (0.15%) had MRI evidence of superficial siderosis, and of those patients only two (0.023%) were symptomatic [12]. The rarity of superficial siderosis is explained by its etiology—chronic low-grade intermittent bleeding

over many years leading to the deposition of hemosiderin in the subpial layers of the CNS.

Unfortunately, it is impossible to determine which patients with CNS lesions are likely to develop superficial siderosis. Given the rarity of the disease, our case illustrates that an aggressive approach to the localization and resection of potential bleeding sources is warranted in the treatment of superficial siderosis. This case also is unique in that a vascular malformation of the cerebral convexity is the etiology of the superficial siderosis. Moreover,

1 Vascular Malformations and Aneurysms Associated with Superficial Siderosis

YEAR	AUTHOR	TYPE	LOCATION	NOTES
1. 1947	Cammermeyer	Aneurysm	Vertebral	Insufficient clinical data
2. 1962	McGee et al.	Venous malformation	Thalamus-periventricular	
3. 1967	Foncin et al.	Spinal AVM	Thoracic	
4. 1969	Hughes, Oppenheimer	Aneurysm	Anterior comm. artery	
5. 1969	Hughes, Oppenheimer	Aneurysm	Middle cerebral artery	Asymptomatic
6. 1983	Pinkston et al.	Spinal AVM	Cervical	
7. 1988	Koepfen, Dentinger	Posterior fossa AVM	Inferior medullary velum	Same case reported
7. 1991	Parnes, Weaver	Posterior fossa AVM	Inferior medullary velum	in two articles.
8. 1996	Offenbacher et al.	Cerebral AVM	Lateral ventricle	Asymptomatic
9. 1996	Offenbacher et al.	Cerebral AVM	Not specified	Asymptomatic
10. 1996	Hashimoto, Hoyt	Cavernous angioma	Lateral ventricle	
11. 1998	Schievink et al.	Spinal AVM	Cervical	

it is the first report of resection of a cerebral arteriovenous malformation for the treatment of superficial siderosis.

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COMMENTARY

This case report by Haroun et al provides a concise review of superficial siderosis. The authors illustrate the comprehensive radiographic assessment (MRI studies of the brain and spinal cord, cerebral and spinal angiography, and CT myelography) that is necessary for the evaluation of these patients. However, radiographic testing may be unrevealing in nearly 50% of patients, and other causes should be considered [2]. Superficial siderosis may be asymptomatic. This report serves as a reminder that superficial CNS siderosis needs to be considered in patients with bilateral sensorineural hearing loss [1] as well as ataxia and pyramidal signs.

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