Superficial siderosis of the central nervous system after brachial plexus injury

Case report

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 \checkmark Superficial siderosis is a rare condition characterized by deposition of hemosiderin in the leptomeninges and in the subpial layers of the brain and spinal cord. It is associated with cerebrospinal fluid abnormalities consistent with recurrent bleeding into the subarachnoid space. The usual symptoms are hearing loss, ataxia, spastic paraparesis, sensory and sphincter deficits, and mental deterioration.

A case is presented of severe superficial siderosis of the central nervous system in a 51-year-old man who had suffered a brachial plexus injury at the age of 20 years. The diagnosis was made by means of magnetic resonance imaging 16 years after the initial symptoms, which comprised bilateral hearing loss and anosmia. Subarachnoid bleeding was due to traumatic pseudomeningocele of the brachial plexus, a very unusual cause of superficial siderosis. This case is interesting insofar as the surgical treatment prevented further bleeding and possibly progression of the disease.

KEY WORDS · hemosiderin · superficial siderosis · chronic subarachnoid hemorrhage · magnetic resonance imaging

UPERFICIAL siderosis of the central nervous system (CNS) is a rare entity, consisting of the deposition of an iron-containing pigment (hemosiderin) in the leptomeninges and in the subpial layers of the CNS.⁵ Clinically, this condition is characterized by slowly progressive symptoms and signs, including bilateral neurosensory hearing loss, ataxia, spastic paraparesis, somatosensory loss, and sphincter disturbances, with possible mental decay. The diagnosis of superficial siderosis may be suggested on obtaining xanthochromic cerebrospinal fluid (CSF) or a history of repeated episodes of subarachnoid bleeding. In the past, this diagnosis was possible only at autopsy, from a finding of deposits of pigment; before 1980, about 40 cases had been described.¹¹⁻¹⁴ Now magnetic resonance (MR) imaging can reveal the presence of hemosiderin in meningeal and subpial tissues, thus allowing the diagnosis during the patient's lifetime.¹⁻⁸ To our knowledge, this is the only reported case of superficial siderosis in which the source of chronic bleeding has been identified and removed at surgery, with normalization of the CSF.

Case Report

This 51-year-old man, who had been deaf since the age of 35 years, came to our attention because of gradually progressive gait ataxia and genitourinary dysfunction. He had been involved in a car crash at the age of 20 years, causing a stretch injury of the left brachial plexus with complete sensorimotor loss in that arm. While in his 30's, he suffered peripheral paralysis of the left facial nerve which resolved completely. In the same period, the patient was admitted to an ear, nose, and throat department because of bilateral neurosensory hypacusis, which became total within a few years. At the age of 38 years, he began to complain of a decrease in the sense of smell, which also slowly progressed toward bilateral anosmia. At 42 years of age, he was hospitalized because of repeated episodes of urinary retention in the absence of obstructive uropathy; on this occasion, a neurological examination revealed a slight spastic paraparesis with ataxia. Myelography demonstrated a posttraumatic meningocele with avulsion of the left C-8 spinal nerve root (Fig. 1). Computerized tomography (CT) of the brain, CSF iso-



FIG. 1. On myelography, no root filaments were visible at the level of the left C-8 and first dorsal root (*arrows*). At the C-7 level, contrast medium fills an extradural cystic cavity (pseudomeningocele) corresponding to the avulsion of the C-8 root.

electrical studies, and Schilling test were all within normal limits, excluding the diagnoses of multiple sclerosis and a nutritional myelopathy. During the following years, the patient's paraparesis gradually worsened.

Examination. In April, 1991, the patient was admitted to our department of neurology. He walked with difficulty, and was unable to go up or down a step; he also complained of urinary frequency, urgency, and impotence. Neurological examination revealed good preservation of cognitive function. Visual acuity was normal. The severity of his hypacusis required communication by means of written messages. Anosmia, bilateral first-degree nystagmus, sensitive ataxia of the lower limbs, spastic paraparesis with bilateral hyperreflexia, clonus, and bilateral Babinski signs were also present. The left upper limb was markedly hypotrophic, with complete anesthesia, whereas the right arm was normal.

On MR imaging of the brain and spinal cord on a 1.5-tesla system, T_2 -weighted axial and coronal images documented diffuse hypointensity along the surface of the brain stem, the cerebellum, and the eighth cranial nerve (Fig. 2). A thick rim of low signal intensity was observed around the optic chiasm, the optic nerves, and the entire spinal cord (Fig. 3). The brain stem, cerebellum, and spinal cord were severely atrophic, especially in the superior vermis region, whereas no atrophy was present in the supratentorial compartment. Gadolinium-enhanced T_1 -weighted MR imaging sequences showed no evidence of enhanced lesions, not even at the level of the meningocele. A CT scan of the brain documented a slight hyperdensity around the pons and mesencephalon (Fig. 4) and confirmed the presence of



FIG. 2. Left: Coronal T_2 -weighted magnetic resonance (MR) image (spin-echo sequence: TR 2200 msec, TE 80 msec, excitations 1) showing a rim of hypointensity consistent with ferromagnetic pigment on the surface of the brain stem, mesial temporal lobes, and sylvian fissures. The proximal segments of the cighth cranial nerves are also abnormally hypointense. The convexity is spared. *Right:* Axial T_2 -weighted MR image (spin-echo sequence: TR 2000 msec, TE 80 msec, excitations 1). The hypointense rim covers the ventral surface of the pons, the proximal portion of the fifth cranial nerve, the superior aspect of the cerebellar hemispheres, and the superior vermis, which appears atrophic. The ventricular surface of the brain stem is spared.

infratentorial atrophy. Brain-stem auditory evoked potentials and somatosensory evoked potentials of the lower limbs were unremarkable, and visual potentials were normal.

During the following 10 months, four separate lumbar punctures disclosed a xanthochromic CSF, with red blood cell sedimentation; this was not related to the original trauma. Cytological and biochemical examinations of the CSF and the Link index were normal. Oligoclonal immunoglobulin G bands were absent. A scintigram with labeled red blood cells and a fourvessel angiogram were then performed to identify the source of bleeding, with negative results.

Operation and Postoperative Course. In May, 1992, the patient underwent left hemilaminectomy at the C-7 and T-1 levels, reduction and repair of the meningocele, and electrocoagulation of a hyperemic arachnoidmedullary scar pad. After treatment, he exhibited left Horner's syndrome and transient worsening of the sensorimotor left leg deficit. This bladder dysfunction also worsened, with urinary retention requiring frequent catheterization. Two lumbar punctures, performed 2 and 4 weeks after surgery, demonstrated complete normalization of the CSF, which was clear and colorless.

Discussion

The first case of superficial siderosis of the CNS was described in 1940 by Noetzel.⁹ In 1969, Hughes and Oppenheimer⁵ reviewed the literature and found about 30 cases, all of them diagnosed post mortem. In 1983, Pinkston, *et al.*,¹⁰ reported the first case studied by CT, with suggestive but not specific findings of superficial siderosis. In the following years, MR imaging specifi-

Superficial siderosis after cervical root avulsion



FIG. 3. Sagittal T_2 -weighted magnetic resonance image (TR 2000 msec, TE 80 msec, excitations 1). The cervical cord is outlined by abnormal superficial hypointensity and appears hypotrophic. The dorsal region and the conus showed the same appearance.

cally revealed the deposition of ferromagnetic pigment, allowing the diagnosis in patients while they were still alive.^{2,3}

The clinical symptoms of superficial siderosis of the CNS are variable and present a slowly progressive evolution; hearing loss and ataxia predominate, often accompanied by sphincter disturbances, spastic paraparesis, and sensory deficit. Mental deterioration sometimes occurs. Almost all cases reported in the literature have exhibited CSF abnormalities consistent with bleeding; subclinical recurrent subarachnoid hemorrhages or chronic bleeding was suggested in patients with known neoplasms or vascular malformations.⁷ A history of brachial plexus injury, as in our case, seems to be an exceedingly unusual finding in cases of superficial siderosis.

In 1991, Koeppen and Borke⁶ succeeded in inducing experimental superficial siderosis in rabbits treated with intrathecal injections of red blood cells. After 3 to 6 months, deposits of ferritin and hemosiderin were found on the surface and in the superficial layers of the brain stem, cerebellum, and piriform cortex. In these regions, microglial cells were able to catabolize hemoglobin; they contained ferritin granules, subsequently transformed into hemosiderin.

With regard to the mechanism of the neurotoxic effect, it has been hypothesized that, when the local iron excess overloads the ferritin biosynthesis capacity of the microglial cells, then ionic iron leads to lipid peroxidation and tissue necrosis.⁶ The peculiar vulnerability of the acoustic nerve possibly depends on its long course within the cerebellopontine angle cistern and on the presence of central glia in a long segment inside the internal auditory canal. Similarly, infratentorial structures and the spinal cord could be selectively involved, being extensively exposed to CSF.



FIG. 4. Plain computerized tomography scan showing marked atrophy of the upper cerebellar vermis. Note the slight hyperdensity surrounding the pons.

In our case, the diagnosis of superficial siderosis was made many years after the initial onset of symptoms, when MR imaging showed the typical findings and four separate lumbar punctures showed CSF abnormalities consistent with chronic persistent bleeding. In an attempt to identify the source of bleeding, which we suppose to be at the sight of the traumatic meningocele, a scintigram with labeled red blood cells was performed; however, this technique was not sensitive because of the paucity of bleeding and its location (close to subaortic trunks). Angiography was also unable to give specific information and excluded only major vascular malformations. Only exploratory hemilaminectomy demonstrated the scarring responsible for the subarachnoid bleeding. Surgical treatment succeeded in stopping further bleeding, as shown by subsequent lumbar punctures. An arrest in the evolution of symptoms and signs in this patient is conceivable, whereas improvement of already established deficits seems improbable.

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