Superficial siderosis of the central nervous system after brachial plexus injury

Case report

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超级侧壁硬膜下出血（CSF）是一种罕见的病变，由铁质色素沉着引起的硬膜外的贫血。该病变的临床表现包括颅内和脊髓的异常，通常表现为出血引起的疼痛、共济失调、感觉障碍和括约肌功能障碍。

**Case Report**

这51岁男性患者，从35岁起就因逐渐进展的共济失调和间歇性尿意困扰。他曾在车祸中受伤，导致左臂严重损伤，以及与之相关的姿势性共济失调，使他难以控制尿液的排尿。

在30岁时，患者在一家医院接受尿道镜检查，发现其左大腿皮肤出现深色斑点，提示可能为硬膜外出血。CT和MRI检查进一步证实了这一诊断，显示硬膜外存在较高信号的出血。

在38岁时，患者出现听力下降，尤其是高频听力，这可能与硬膜外出血有关。神经科医生进一步评估时发现患者双耳听力受损，以及共济失调和平衡障碍。

CT和MRI检查可帮助医生诊断脑内病灶，但在这病例中，出血区域未显示明显异常。

**Key Words** · hemosiderin · superficial siderosis · chronic subarachnoid hemorrhage · magnetic resonance imaging

**Superficial 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. 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. 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. 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.
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, T₂-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 T₁-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 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 four-vessel 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 arachnoid-medullary 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. In 1969, Hughes and Oppenheimer reviewed the literature and found about 30 cases, all of them diagnosed post mortem. In 1983, Pinkston, et al., 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-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.

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 subarachnoid 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.

References


Manuscript received April 29, 1993. Accepted in final form September 9, 1993. This paper was presented in part at the XIX Symposium on Brachial Plexus, Lausanne, Switzerland, in 1992. Address reprint requests to: Virginio Bonito, M.D., Department of Neurology I, Ospedali Riuniti, Largo Barozzi 1, 24100 Bergamo, Italy.