

# CLINICAL CASE REPORT

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## Superficial Siderosis: A Case Report

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**Key words:** superficial siderosis; magnetic resonance imaging.

**Summary.** Superficial siderosis of the central nervous system is the result of chronic recurrent hemorrhages (e.g., arteriovenous malformations, tumors, or trauma), which leads to the accumulation of cytotoxic hemosiderin and presents with hearing loss, cerebellar dysfunction, and myelopathy. This article presents a clinical case of an 11-year-old boy in whom the diagnosis of medulloblastoma was established. He underwent surgery, and after a few years, he began to complain of hearing loss. Magnetic resonance imaging revealed the cause of the hearing disturbance. The aim of this article is to review the recent literature related to the etiology, clinical and radiologic features of superficial siderosis, emphasizing the role of magnetic resonance imaging.

### Introduction

Superficial siderosis (SS) of the central nervous system (CNS) is an uncommon and often unrecognized disorder in which hemosiderin – a product of the breakdown of blood – is deposited in the leptomeninges, subpial layer, and ependymal surface of the CNS. This leads to progressive and irreversible neurological dysfunction (1, 2). SS is caused by chronic, slow, or repeated bleeding into the subarachnoid space (1, 3, 4). The first pathologic clinical presentation of SS was a case presented by Hamill in 1908, but only approximately 40 cases appeared in the literature before the widespread use of magnetic resonance imaging (MRI) enabling physicians to make the diagnosis without a biopsy (1, 2). MRI is the only diagnostic method, which shows pathognomonic features of hemosiderin accumulation on CNS surfaces (3).

### Case Report

Seven years ago, a boy at the age of 11.5 years was diagnosed with a posterior fossa tumor. The tumor was radically resected and proved to be a medulloblastoma on pathologic examination. Subsequently, the boy was treated with heavy chemotherapy: for the first time, methotrexate, cyclophosphamide, and vincristine were administered; for the second and third, vincristine and double dose of methotrexate; and for the fourth time, methotrexate, carboplatin, and etoposide. The postoperative MRI revealed no evidence of tumor recurrence and no abnormalities in the labyrinths or inner auditory canals. After two years, the child developed severe bilateral hearing loss. For this, new MRI scans were

performed almost every year, and in 2010, SS was seen as a hypointense rim at the mesencephalic tectum and cerebellar folia (Fig. 1). Retrospectively, this finding could readily be seen on previous scans as far back as 2005 (Fig. 2), and since then, the process seemed to be stable.

### Discussion

Around 65% of cases of superficial siderosis have a detectable underlying cause (1). Most frequently, it is secondary to current or previous CNS tumors (21%), followed by head or back trauma (13%), arteriovenous malformations/aneurysms (9%), post-surgical changes related to neurosurgeries (7%), brachial plexus injury (6%), amyloid angiopathy (3%), and chronic subdural hematomas (6%) (1, 3). It is known that superficial siderosis is caused by chronic bleeding into the subarachnoid space. Blood breakdown products are deposited into the subpial layer of the brain and spinal cord. Free iron catalyzes the breakdown of hydrogen peroxide to superoxide. This toxic product can cause lipid peroxidation, membrane dysfunction, and ultimate cell death. The final result is neuronal loss, reactive gliosis, and demyelination (4).

Hemosiderin deposition prevails in those parts of the CNS that are more exposed to cerebrospinal fluid such as the superior vermis, crest of the cerebellar folia, basal frontal lobe, temporal cortex, brainstem, spinal cord, nerve roots, and cranial nerves I and VIII (1, 2, 4). This predilection is elucidated by the specific flow pattern of cerebrospinal fluid (CSF) and the microscopic anatomy of the nervous system (5). Some parts of the CNS also have a greater

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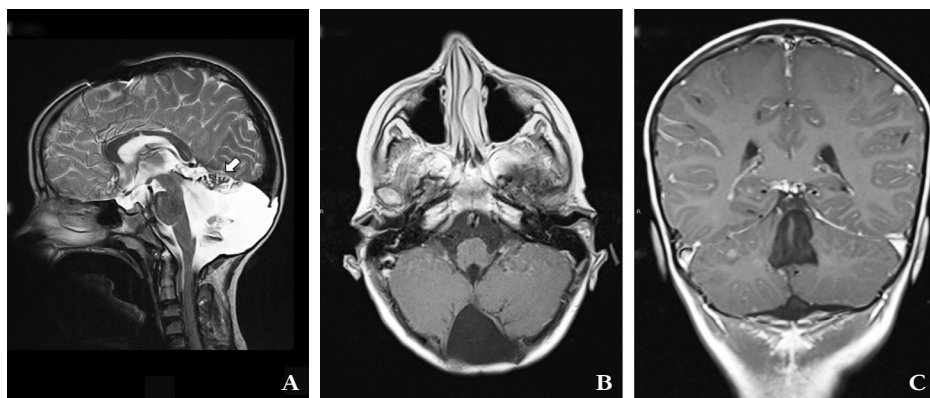


Fig. 1. MRI brain images of the patient

A, T2-weighted sagittal view; B, T1-weighted contrast-enhanced axial view; C, T1-weighted contrast-enhanced coronal view. In 2010, a relatively large postoperative parenchymal defect is seen at the site of previous tumor. There is no evidence of tumor recurrence and detectable dissemination of leptomeningeal tumor. Fig. 1A shows a hypointense rim at the mesencephalic tectum and cerebellar folia (arrow) – a pathognomonic feature of superficial siderosis.

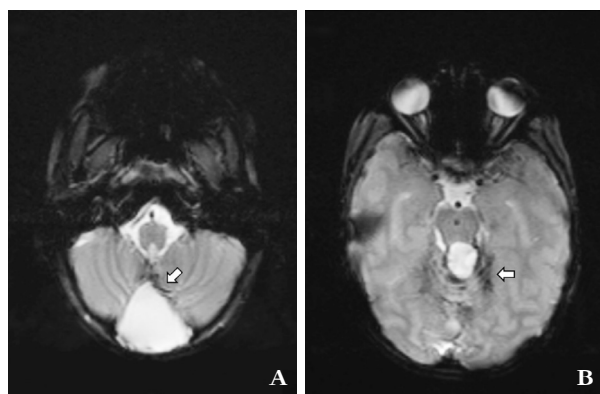


Fig. 2. T2-weighted axial views

A, T2-weighted axial view through the middle aspect of cerebellum; B, T2-weighted axial view through the superior aspect of cerebellum. The subtle marginal hypointensity was already seen in 2005 around the crest of the cerebellar folia (arrows).

number of specialized cells, such as Bergmann cells in the cerebellum, microglia, and superficial astrocytes, which are responsible for isolation of blood breakdown products. When these protective mechanisms are exhausted, toxic substances began to destroy the brain tissue. It should be noted that there is no correlation between the etiology of bleeding and specific sites of tissues pigmentation (1).

A latent period of the disease can last from 4 months to 30 years (2). Males are affected more frequently than females with a ratio of 2:1 (1). The classical clinical symptoms of SS are sensorineural hearing loss, cerebellar ataxia, and myelopathy; only 39% of patients have all three of these symptoms (1, 4). Other clinical findings such as dementia, urinary problems, headaches, anosmia, bowel problems, diplopia, ageusia are less common (1). Hearing loss, anosmia, and ageusia are associated with the damage of cranial nerves VIII, I, and VII, whereas cerebellar ataxia results from cerebellar at-

rophy. CSF in most cases appears normal, but sometimes may show elevated red blood cell count, high protein level, and xanthochromia.

MRI is the investigation modality of choice, which shows pathognomonic features of the disease. However, in the early period of SS, the findings are very subtle and can be easily overlooked (6). Gradient-echo T2-weighted images have a higher sensitivity for hemosiderin deposition (4). T2-weighted MR imaging (especially gradient-echo T2-weighted images) shows a characteristic marginal hypointensity around these parts of the CNS, which have a predilection for hemosiderin deposition (1, 2, 4, 6). T1-weighted imaging may show a less extensive, partially corresponding marginal hypointensity around the spinal cord, cerebellum – especially the vermis of the cerebellum – brainstem and lining of the lateral ventricles (4, 6). On fluid-attenuated inversion recovery, an MRI scan of the brain shows hypointensities throughout the leptomeninges (1). The sites of hemosiderin deposition typically do not enhance; however, if SS is associated with intracranial hypotension, there may also be evidence of meningeal enhancement (3, 6). The peripheral nervous system is not involved in SS.

Cerebellar (particularly the superior vermis and anterior cerebellar hemispheres) and spinal cord atrophy also can be seen on both MRI and computed tomography (CT).

When the diagnosis of SS is established, the second step should be identification of the bleeding source by performing additional imaging modalities (e.g., CT myelography, angiography, contrast material-enhanced brain MRI, total spine MRI).

Some conditions can mimic the features of SS on MRI. MR sequence artifacts and normal or abnormal brain surface vessels (veins) must be ruled out. Neurocutaneous melanosis can also show similar imaging findings, but the clinical picture is usu-

ally very different from SS as this entity most often manifests at the age of 2–3 years with characteristic skin pigmentation and high intracranial pressure. Leptomeningeal invasion may look similar to SS on MRI; however, if there is leptomeningeal melanoma, the meninges will enhance contrast material.

The neurological deficit made by hemosiderin is irreversible, and the only way to minimize poor outcomes is the early diagnosis of the disease and identification and ablation of the bleeding source. MRI enables the establishment of the correct diagnosis and thus allows avoiding complications leading to long-term debilitation.

## Conclusions

Superficial siderosis shows pathognomonic and unmistakable features on magnetic resonance imaging. However, these features are subtle in the early stage of the disease and can be easily overlooked. To avoid such a mistake, an accurate clinical assessment and evaluation of history and symptoms coupled with imaging studies should be carried out. Correct diagnosis will be confirmed by choosing special magnetic resonance imaging scans.

## Statement of Conflict of Interest

The authors state no conflict of interest.

## Paviršinė siderozė. Klinikinis atvejis

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**Raktažodžiai:** paviršinė siderozė, magnetinio rezonanso tomografija.

**Santrauka.** Centrinės nervų sistemos paviršinę siderozę, pasireiškiančią klausos netekimu, smegenėlių disfunkcija bei mielopatija, sukelia lėtinis pasikartojantis kraujavimas (pvz., arterioveninės malformacijos, navikinės arba trauminės kilmės) ir citotoksinis hemosiderino susikaupimas. Šiame straipsnyje apžvelgiamas 11 metų berniuko klinikinis atvejis. 11 metų berniukas, praėjus keleriems metams po meduloblastomos pašalinimo operacijos, apkurto. Klausos sutrikimo priežastis nustatyta tik atlikus magnetinio rezonanso tyrimą. Straipsnio tikslas – apžvelgti naujausią literatūrą apie paviršinės siderozės etiologiją, klinikinius ir radiologinius požymius, įvertinti magnetinio rezonanso svarbą diagnozuojant šią ligą.

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