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# A Case of Idiopathic Infratentorial Superficial Siderosis

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Superficial siderosis is attributed to hemosiderin deposition in the subpial layers of the nervous system. The clinical features of infratentorial superficial siderosis (ISS) are hearing loss, cerebellar ataxia, and corticospinal tract signs and the most common cause of idiopathic ISS is a dural defect. As magnetic resonance imaging (MRI) has advanced, the diagnosis of infratentorial superficial siderosis can be confirmed by unique radiological findings in MRI. Here, we report on a female patient diagnosed with idiopathic ISS by means of clinical symptoms and radiological findings.

**Keywords:** Superficial siderosis; Hearing loss; Cerebellar ataxia

Case Report

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## INTRODUCTION

Superficial siderosis is attributed to hemosiderin deposition in the subpial layers of the nervous system following recurrent hemorrhages in subarachnoid spaces (1). Infratentorial superficial siderosis (ISS) presents with distinct clinical characteristics, including progressive hearing loss, cerebellar ataxia, and corticospinal tract signs (2). As magnetic resonance imaging (MRI) has advanced, the diagnosis of ISS can now be confirmed by unique radiological findings on MRI (3). Here, we report on a 64-year-old woman who presented with progressive cerebellar dysfunction and was finally diagnosed with idiopathic ISS by MRI.

## CASE REPORT

A 64-year-old woman visited the Department of Neurology presenting with progressive gait disturbance with postural instability for six months. She had no trauma, smoking or alcohol drinking history. She had no infectious symptoms, such as fever and chilling sensations. She denied any exposure to toxic materials associated with the symptoms. On neurological examination at admission, hearing difficulty, dysarthria, limb, and gait ataxia were identified. Tests of muscle power, sensory, and deep tendon reflex were normal.

Complete blood count, liver function tests, random blood sugar, renal function tests, thyroid function tests, antinuclear antibody, serum ferritin, serum ceruloplasmin, VDRL (venereal disease research laboratory) test for syphilis, HIV test, chest X-ray, and electrocardiography were normal.

Interestingly, unique radiological findings were demonstrated on the brain and spine MRI with a 3-Tesla scanner (Fig. 1). The T2-weighted axial brain and spine MRI

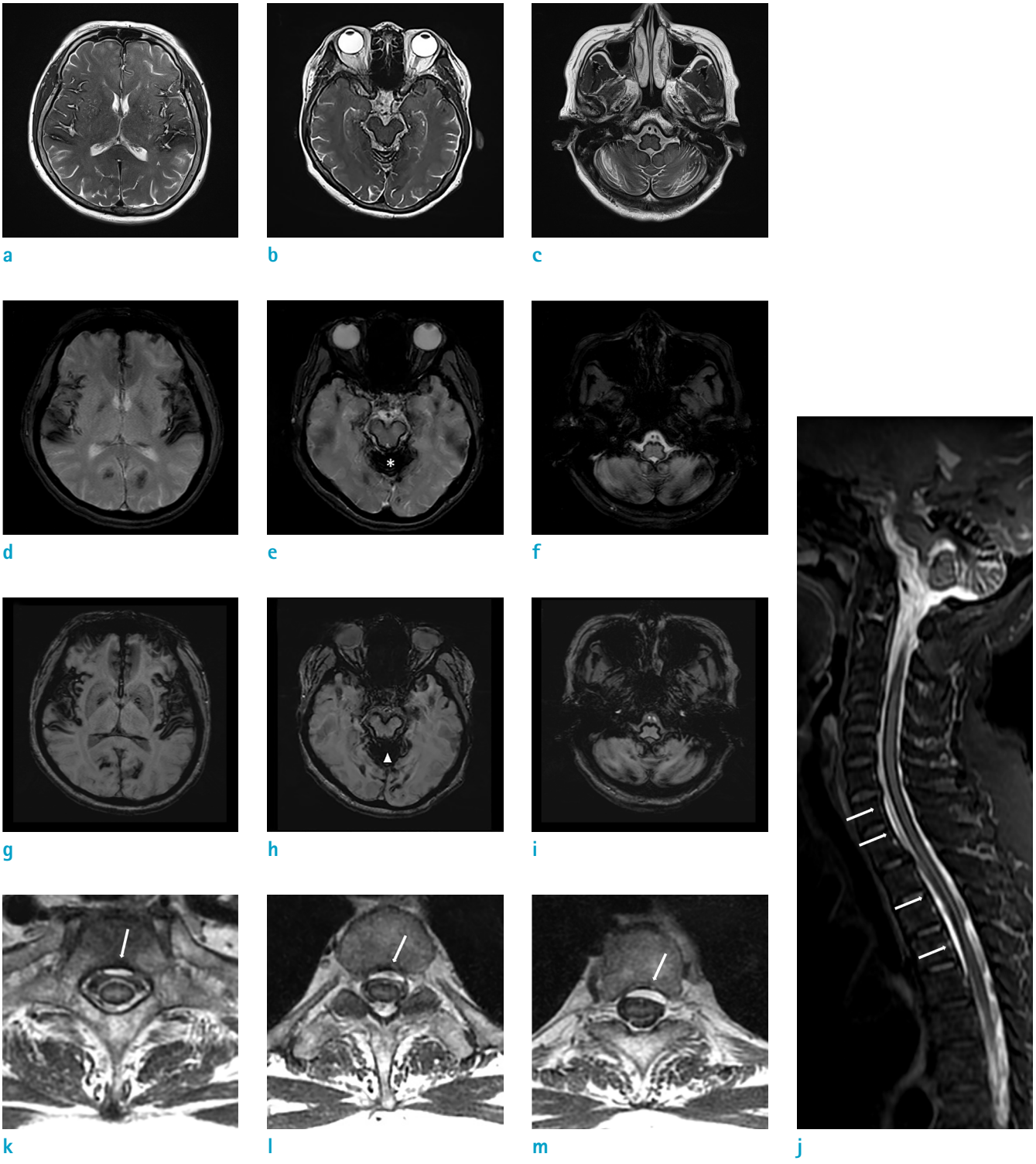


**Fig. 1.** Idiopathic infratentorial superficial siderosis. Brain magnetic resonance imaging (MRI) (a-c) and spine MRI (k-m) show low signal intensities in the cerebrum, cerebellum, brain stem, and spinal cord in T2-weighted images and the corresponding T2*-weighted gradient-recalled echo (GRE) images (d-f) or susceptibility-weighted images (SWI) (g-i) show low signal intensities in the same regions, which suggest a typical hemosiderin deposition pattern. The "blooming" effect on the SWI (h, arrowhead) is more remarkable compared to that on the T2-weighted GRE images (e, asterisk). Spine MRI (j-m) shows cerebrospinal fluid collection (arrows).

images showed low signal intensities along the outlining surfaces of the cerebrum, cerebellum, brain stem, and spinal cord (Fig. 1a-c, 1k-m). Low signal intensities in the corresponding regions were remarkably noted on the T2*-weighted gradient-recalled echo (GRE) and susceptibility-weighted axial images on the brain MRI (Fig. 1d-f, 1g-i). The "blooming" effect was noted on the GRE images (Fig. 1e) and susceptibility-weighted images (SWI) (Fig. 1h). The "blooming" effect on the SWI was more prominent than that on the GRE image. The fat-suppressed T2-weighted sagittal image of the cervical and thoracic spine (Fig. 1j) showed extradural cerebrospinal fluid (CSF) collection at the ventral portion probably attributed to the dural defect. We diagnosed this patient with idiopathic ISS successfully by means of distinct clinical features and radiological findings.

## DISCUSSION

Superficial siderosis is a rare neurological disorder that is caused by the accumulation of hemosiderin in the subpial layers of the nervous system, including the brain, spinal cord, and cranial nerves (1). It was first described in 1908 (4) and hypothesized to result from an insidious, low-volume, and repetitive leakage of red blood cells into the subarachnoid space (5).

The clinical features of ISS are sensorineural hearing loss, cerebellar ataxia, corticospinal tract signs, cognitive impairment, urinary bladder dysfunction, and hyposmia (2). In the past, superficial siderosis could only be suggested from clinical findings and CSF studies. Thus, diagnostic confirmation was only possible by autopsy.

With the advent of MRI, superficial siderosis is now a common radiological diagnosis. Especially, the blood-sensitive sequence of the MRI is extremely helpful in diagnosing superficial siderosis (3). On T2-weighted images, a low signal-intense rim, indicating hemosiderin deposition, is seen in those parts of the nervous system that are adjacent to CSF (6). On T1-weighted images, a less extensive, partially corresponding high signal-intense rim can be seen, which has been proposed to be secondary to the presence of blood breakdown products at different stages of evolution (7). Atrophy and signal intensity abnormality of the involved regions suggests tissue damage combined with superficial siderosis (8). T2*-weighted GRE images are exquisitely sensitive for this condition and thus, hemosiderin deposition appears thicker than on T2-weighted MRI (3). There is also a blooming dark signal along the subpial regions of the nervous system. Susceptibility-weighted imaging is also widely accepted for detecting blood-degradation products in the brain with high sensitivity (9). SWI is known to be more sensitive to paramagnetic material than T2*-weighted GRE images. Thus, SWI showed higher sensitivity than T2*-weighted GRE images in detecting superficial siderosis (9).

Numerous secondary causes, including cerebral amyloid angiopathy, subarachnoid hemorrhage, intracranial hemorrhage, and reversible cerebral vasoconstriction syndrome, are presumed based on the spatial distribution and timing of siderosis (2). Superficial siderosis is classified into cortical and infratentorial superficial siderosis according to the distribution by MRI. Cortical superficial siderosis confined to the supratentorial region of the brain is often associated with cerebral amyloid angiopathy (10). However, infratentorial superficial siderosis involved in the infartentorial region of the brain, with or without supratentorial involvement, is associated with different suggested causes (2). Our case could be classified as idiopathic infratentorial superficial siderosis. The most common cause of idiopathic ISS is dural abnormalities, including pseudo-meningocele, extra-arachnoid CSF collection, and dural ectasia. The pathophysiology of the superficial siderosis due to dural defects is explained by the persistent or repetitive red blood cell leakage from the dural venous and capillary network to the subarachnoid space (2). CT myelography may help localize dural defects (2). Although there is no established therapy with evidence of efficacy and safety, surgical repair or blood patch of the dural defects and iron-chelating agents can be considered (11). In the future, CT myelography will be considered before repairing dural defects in these patients.

In conclusion, we reported a rare neurological disorder, idiopathic ISS. We demonstrated the unique MRI findings that could provide useful information for understanding patients with idiopathic ISS.

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