Clinical Case Reports International

Case Report
Published: 16 Nov, 2025

Superficial Siderosis of the Central Nervous System: A Case Report of the Sparsely Occurring Neurological Disorder Accompanied by Review of Literature

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Abstract

Background: Superficial Siderosis (SS) of the Central Nervous System (CNS) is a neurological dysfunction that occurs sparsely and is attributable to chronic and repetitive outpouring of blood into the cerebrospinal fluid. This results in accumulation of iron on the surface of the brain, and spinal cord that damage nerve tissues over time. Generally, these patients are presented with cerebellar ataxia, progressive and irreversible Sensorineural Hearing Loss (SNHL), and bilateral pyramidal impairment, but a very few of them manifest all three symptoms at the same time, which make diagnosis and treatment ardous.

Case Presentation: We report the case of a man in his late fifties who came to the hospital with complains of unsteadiness during walking, headache, decreased hearing, and tingling and numbness in his bilateral lower limbs for last 3-5 years. He had a medical history of hypertension and type II diabetes mellitus.

Results: A physical examination revealed cerebellar ataxia, gaze evoked nystagmus, broken pursuit and slow saccades, progressive bilateral SNHL, corticospinal & dorsal column involvement. Bilateral intention tremor was present. Bilateral finger nose test, Heel-knee-shin Test and tandem walking was impaired indicating cerebellar dysfunction. Cranial nerve VIII was impaired indicating Weber test lateralized to right side and Romberg sign positive.

Conclusion: This case indicates that patients presenting with progressive cerebellar ataxia, hearing loss, headache, bilateral Pyramidal & dorsal column signs, may have the possibility of developing superficial siderosis and the diagnosis should be further corroborated by Magnetic Resonance Imaging (MRI) along with angiography and venography.

Keywords: Superficial siderosis of the central nervous system; Cerebellar ataxia; Sensorineural deafness; Case report; Iron deposition

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Received Date: *07 Nov 2025*
Accepted Date: *12 Nov 2025*
Published Date: *16 Nov 2025*

Citation:
Mishra BK, Pradhan RK, Dash S, Mishra SV, Mathur D. Superficial Siderosis of the Central Nervous System: A Case Report of the Sparsely Occurring Neurological Disorder Accompanied by Review of Literature. Clin Case Rep Int. 2025; 9: 1738.

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Introduction

Superficial Siderosis (SS) of the central nervous system is a neurological disorder that occurs sparsely and is caused by excessive iron deposition on the brainstem, cerebellum, spinal cord, and even part of the cranial nerve surface caused by chronic outpouring of blood into the cerebrospinal fluid [1,2]. Despite being first identified more than a century ago, this condition is often underdiagnosed and leads to a slowly progressive but crippling neurological decline. The classical clinical triad of SS is comprised of progressive and irreversible sensorineural deafness, cerebellar ataxia, and pyramidal dysfunction, but a very few patients exhibit all three symptoms simultaneously, which makes diagnosis ardous [3]. Currently, there are many shortcomings of properly diagnosing the disease and providing timely treatment to the patient. This happens because of late diagnosis and dearth of long-term follow-up data which leads to distinct treatment to patients among different regions [4]. Susceptibility-Weighted Imaging (SWI) is an indispensable tool for the correct diagnosis of SS [5].

We report the case of a man in his late fifties who came to the hospital with complains of headache, unsteadiness during walking, decreased hearing, and tingling and numbness in bilateral lower limbs for last 3-5 years. His medical history included hypertension and type II diabetes mellitus. After thorough examination, we have come to a conclusion that this case with progressive

cerebellar ataxia, hearing loss, headache, bilateral pyramidal and dorsal column signs, may have SS of the CNS and the diagnosis is further corroborated by Magnetic Resonance Imaging (MRI) along with angiography and venography.

Case Presentation

We present a case report of a man in his late fifties who was hypertensive for last 5 years and type II diabetic for last 2 years. He had a five-year history of progressive truncal and limb ataxia, intention tremors and vertigo, holo-cranial headache for four years and bilateral progressive hearing loss for three years. Furthermore, he showed symptoms of tingling and numbness in bilateral lower limbs for four months. There was no bladder bowel involvement and no significant family history. He had complaints of unsteadiness during walking for last 5 years. He suffered from headache for last four years and tremulousness in both hands which was progressive. He suffered with impaired hearing for three years. He had no history of hypothyroidism, alcohol consumption and smoking.

The differential diagnosis suggested that there was a multi-neuraxial involvement. For gait ataxia and vertigo, flocculonodular lobe and connections were involved. For truncal ataxia, vermis was involved. For upper limb, neocerebellum was involved. For intention tremor, superior cerebellar peduncle (Dentate Nucleus Type) and for hearing loss -Bilateral VIII, the cranial Nerve was involved and for tingling sensation, large fiber sensory peripheral nerves were involved.

The patient was differentially diagnosed with superficial siderosis which was in favor of cerebellar ataxia, progressive b/l SNHL, corticospinal and dorsal column involvement and headache. Cranial nerves III, IV and VI pursuits, saccades were broken and slow. Bilateral horizontal gaze evoked nystagmus was present.

A physical examination of the nervous system revealed a Mini-Mental State Examination score of 26; higher mental function, conscious, oriented to time, place and person. Memory was immediate, recent, remote-normal. Speech and language were normal; Weber test results lateralized to right side; Bilateral plantar reflex was extensor; and a positive Romberg's sign.

Cerebellar signs showed nystagmus which was Bilateral Horizontal Gaze Evoked. Truncal Ataxia; bilateral Intention Tremor; impaired bilateral Finger Nose Test; a positive dysdiadochokinesia - Positive; impaired Heel-knee-shin Test; impaired tandem walking; wide based stance with ataxic Gait, swaying to either side, turning was slow.

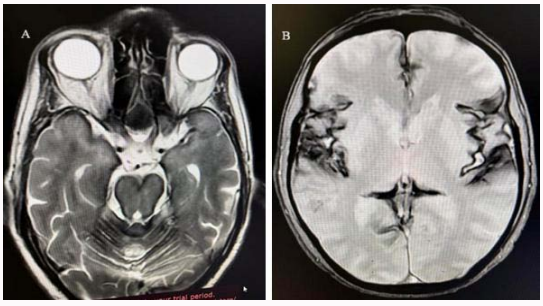

Figure 1: Diffuse sheet of T2 hypo intensity surrounding the cerebral parenchyma, brain stem and cerebellar parenchyma (A), with blooming on GRE (B), Disproportionate atrophy of the cerebellar parenchyma is seen in form of prominent sulci, fissures and ventricles.

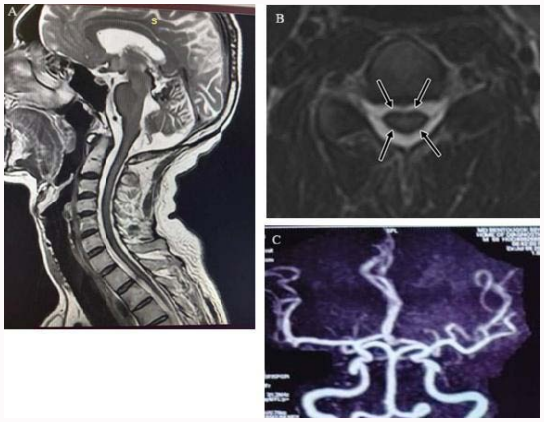

Figure 2: Cervico dorsal spine screening reveals T2 hypo intensity in the anterior and posterior aspect of the spinal cord (A, B). The spinal cord is mildly reduced in bulk with long segment of the T2 hyperintensity involving the central cord. Normal MR Angiography of the brain (C).

There were no extrapyramidal signs. There were no signs of meningeal irritation. Spine and cranium were normal. Autonomic nervous system showed no postural hypotension. He had no palpitation, and had resting tachycardia. He had no abnormal sweating.

Diffuse sheet of the T2 hypo intensity is seen surrounding the cerebral parenchyma, brain stem and cerebellar parenchyma with blooming on Gradient Recalled Echo (GRE) (Figure 1). Disproportionate atrophy of the cerebellar parenchyma and middle cerebellar peduncle is seen in form of the prominent sulci, fissures and ventricles. Cervicodorsal spine T2 hypo intensity was observed in the anterior and posterior aspect of the spinal cord (Figure 2). The spinal cord was mildly reduced in bulk with long segment of the T2 hyperintensity involving the central cord with thickening and clumping of the cauda equina nerve roots which is suggestive of chronic superficial siderosis of the brain and spinal cord with cerebellar atrophy. In addition, mild spinocerebellar atrophy was also observed with secondary myelopathy.

When the patient came to the hospital because of the clinical manifestation of walking instability, a physical examination indicated cerebellar ataxia. Diagnoses of genetic ataxia, multiple systems atrophy, mitochondrial disorders and other diseases were taken into consideration. According to the patient's history of slow progressive cerebellar ataxia, sensorineural hearing loss, and corticospinal tract symptoms, the finding of hemosiderin deposits mainly distributed along the meningeal lining on MRI, and the lack of a clear source of bleeding, demonstrated classical SS (idiopathic) of the CNS as the final diagnosis.

Discussion

The hallmark feature of Superficial Siderosis (SS) is the chronic proliferation of hemosiderin on the innermost meninges (pia mater and arachnoid mater) and space below pia mater of the CNS, including the brain, brainstem, cerebellum, cranial nerves, and spinal cord [6]. The cause of SS is elusive and the underlying pathology entails repeated leakage of blood into the Cerebrospinal Fluid (CSF) Fang, et al. showed that intraventricular cavernous hemangioma hemorrhage leads to SS of the CNS [7]. Interestingly, Orru, et al. [8] highlighted the significance of considering CSF-VF in the differential diagnosis of SS, especially when dural tears and epidural collections

are absent on imaging.

The continuous breakdown of blood products results in deposition of iron on brain surfaces eventually leading to toxicity in the brain [6]. Iron accumulation drives oxidative stress, gliosis, myelin damage, and neuronal degeneration in cerebellum, cranial nerve VIII, and spinal cord Auger, et al [9]. revealed that iron junks found in the cortical subtype of SS specifically when linked with Cerebral Amyloid Angiopathy (CAA) have been associated with reactive astrocytosis and microglial activation, indicating the involvement of a neuro-inflammatory component [10]. Clinically, SS presents three types of symptoms: progressive sensorineural hearing loss, cerebellar ataxia/gait disturbance, and myelopathic or pyramidal signs such as spasticity and bladder/bowel dysfunction when the spinal cord is affected [6]. In cases where SS manifests in the infratentorial area, hearing loss may be the first symptom. Ataxia is often observed as a neurological symptom because the surface of cerebellum is particularly vulnerable to developing this condition.

In the current case, the manifestation of cerebellar ataxia, gaze evoked nystagmus, tingling and numbness in bilateral lower limbs, decreased hearing, Progressive bilateral SNHL, orients with the classical description of SS, which strengthens the diagnosis. Current knowledge suggests that chronic and recurrent Subarachnoid Hemorrhage (SAH) of various causes is the main cause of SS of the CNS [11].

On the basis of distinct etiology in patients coming from different regions, pathogenesis and clinical features, SS can be classified into classical and secondary types. The classical form principally influences the sub tentorial region (cerebellum, brainstem, cranial nerves, and spinal cord) and is associated with meningeal bleeding. On the contrary, the secondary type emanates from recurrent or existing SAH [12]. A more recent classification categorizes SS on the basis of location of hemosiderin deposition: cortical superficial siderosis (deposits primarily on the cerebral cortex surface) or Infratentorial Superficial Siderosis (iSS) [13]. In iSS, hemosiderin is accumulated primarily in the brainstem, cerebellum, and spinal cord, potentially with supratentorial diffusion. The iSS type is further grouped into type 1 and type 2 subtypes. Type 1 iSS is characterized by the absence of cerebral hemorrhage as evidenced in imaging and the main feature of type 2 is the presence of clear spontaneous or traumatic intracranial hemorrhage as seen in imaging [14]. A recent cross-sectional and observational study conducted by Kharytaniuk, et al. determined the prevalence of iSS in the UK Biobank sample using pre-defined radiological criteria. Findings of this study revealed that five cases with radiological features of iSS were identified out of 10,305 SW MRIs reviewed, demonstrating cerebellar/superior vermis involvement [15].

The mechanisms underpinning the pathophysiology of SS of the CNS are poorly understood, but the prevailing theory describes a cascade initiated by continuous bleeding into the subarachnoid space. This sequence of events begins with the chronic or intermittent entry of blood into this space. Subsequently, red blood cells within the Cerebrospinal Fluid (CSF) disintegrates, releasing hemoglobin. This hemoglobin, in turn, releases heme, which then accumulates onto the surfaces of the brain, spinal cord, and cranial nerve tissues that are in close affinity with the CSF. Glial and microglial cells absorb the accumulated heme, inducing the synthesis of heme oxygenase-1 and ferritin. Heme oxygenase-1 breaks down heme into free iron and biliverdin, while ferritin binds the free iron to create hemosiderin, an iron-storage protein. The crucial final step involves the toxic accumulation of this hemosiderin within these tissues. This accumulation causes cellular apoptosis, oxidative stress, and generation of free radicals, ultimately culminating in neurodegenerative injury and damage within the CNS [16].

The clinical features of SS vary depending on the location, degree, severity, and duration of iron accumulation, as well as individual patient differences. The classic clinical trial of SS of the CNS comprises of progressive, irreversible sensorineural deafness (95%), progressive cerebellar ataxia (88%), and pyramidal dysfunction (76%) however, the simultaneous occurrence of all these symptoms occurs in less than half of patients [3]. It has been reported that cognitive defect influences nearly half of the patients. Moreover, the severity of defect significantly correlates with the disease's duration [3,17]. Other unusual symptoms that are observed include urinary problems, olfactory impairment [18], sexual dysfunction [19], and vestibular dysfunction [20]. Hearing loss, the most common and earliest symptom of SS, typically involves prodromal tinnitus followed by sensorineural hearing loss and eventual deafness within 15 years. This hearing loss is often more severe than expected for the patient's age. Unfortunately, clinicians often diagnose and treat only the hearing loss without considering issues in nervous system, leading to a missed diagnosis of the underlying cause. While cerebellar ataxia and progressive walking difficulties were the initial symptoms in the case discussed, the presence of hearing loss hints towards considering superficial siderosis as the diagnosis.

Before the development of MRI, the diagnosis of SS of the CNS was mainly dependent on pathological analysis, lumbar punctures, and surgical observations. Advances in imaging technology have since improved the detection rate of SS, with MRI becoming an essential diagnostic instrument. According to the MRI results of this patient, the hemositin deposits were mainly distributed on the brainstem, cerebellar surface, and spinal cord, which is consistent with the clinical signs of cerebellar damage (vague language and gait instability), brainstem damage (hearing loss and hoarse voice), and spinal cord damage (pyramidal tract sign) suggesting that the patient may have classical SS.

Conclusion

This case suggests that the possibility of SS should be considered in patients with progressive cerebellar ataxia, hearing loss, and the diagnosis should be further confirmed by MRI. For confirmed patients, it is important to find out the cause of the disease.

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