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# An Unusual Case Report: Superficial Siderosis of Bilateral Internal Auditory Canal Presenting as Temporary Facial Palsy

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**Abstract:** ***A 33-year-old woman with a history of heart valve replacement on warfarin presented with brief history of facial weakness and hearing loss. MRI showed superficial siderosis (SS) affecting both internal auditory canals—an uncommon and localized finding, as SS usually causes permanent nerve damage. Her recovery with conservative treatment suggests such damage may be reversible. This case highlights the need for early MRI diagnosis.***

**Keywords:** Superficial siderosis, facial palsy, hearing loss

## 1. Introduction

Superficial siderosis (SS) is a rare neurological disorder characterized by the deposition of hemosiderin, an iron-containing pigment, on the surface of the central nervous system (CNS). This deposition results from recurrent or chronic bleeding into the subarachnoid space, where cerebrospinal fluid (CSF) circulates. The condition is most commonly diagnosed on magnetic resonance imaging (MRI), which demonstrates a characteristic hypointense rim on T2*-weighted images, corresponding to hemosiderin deposits on the pial surfaces of the brain and spinal cord [1].

During prolonged or recurrent bleeding episodes, ferritin synthesis may become insufficient. This impairment limits the sequestration of free iron, which accumulates in the subpial layers and catalyses the formation of highly reactive hydroxyl radicals via Fenton chemistry. These radicals contribute to oxidative damage and neurodegeneration, thereby exacerbating neuronal injury and cognitive dysfunction following subarachnoid haemorrhage [2].

Clinical manifestations vary depending on the site of hemosiderin deposition. Common neurological symptoms include:

- **Cranial nerve deficits:** Sensorineural hearing loss, facial weakness, or visual disturbances, depending on affected nerves.
- **Gait ataxia:** Broad-based, unsteady gait due to cerebellar vermis involvement.
- **Myelopathy:** Pyramidal signs, sensory disturbances, bladder dysfunction, and spasticity with spinal cord involvement [3].

Early recognition and diagnosis are crucial for managing the condition and addressing the underlying cause of bleeding. (3)
Peripheral facial palsy (PFP) involves lower motor neuron dysfunction of cranial nerve VII and may result from infections, trauma, tumours, autoimmune disorders, or pregnancy. While Bell's palsy is the most common cause, viral reactivation (HSV-1, VZV) is implicated in many cases [4]. Rarely, cranial nerve neuropathy may arise from microbleeds or intra-axial lesions affecting the root entry zone (REZ) or attached segment (AS) [5].

In this case report, an unusual case of temporary facial nerve palsy is seen caused by microbleeds located at the attached segment (AS) and root entry zone (REZ) of the facial nerve associated with bilateral internal auditory canal superficial siderosis.

## 2. Case Presentation

A 33-year-old female presented with **acute right-sided facial weakness** and **decreased hearing** for one day, associated with **giddiness**. She had a history of **mitral stenosis** for which pt had underwent **mitral valve replacement 20 years prior**, and also **left atrial clot removal in 2022**, and she has been put on **long-term warfarin therapy**. She also had **hypertension on treatment**.

On **clinical examination**, she had **House-Brackmann Grade IV right facial palsy**, bilateral **intact tympanic membrane**, and a **positive Romberg's test**. **Pure tone audiometry** revealed **profound sensorineural hearing loss (SNHL)** in the right ear.
**MRI brain** performed one month later revealed:

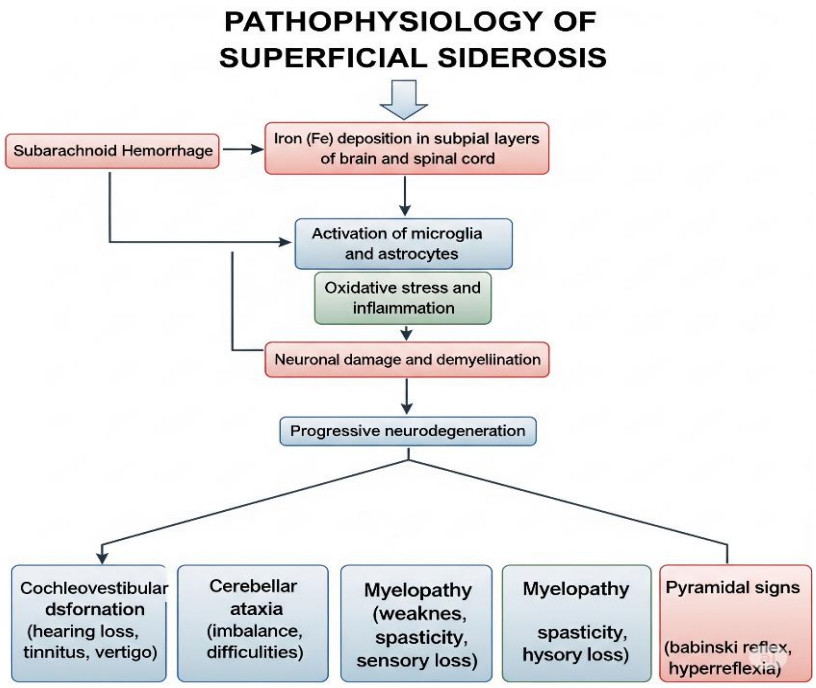
- **FLAIR hyperintense and T2-weighted hypointense signals** in **bilateral internal auditory canals (IACs)**.
- **Indistinct intracanalicular segments of the 7th and 8th nerve complexes**.
- **Blooming artifact** consistent with **hemosiderin deposition**, suggestive of **superficial siderosis**, though evaluation was limited by the adjacent **petrous bone**.

No active ENT intervention was required. **Serial follow-up** demonstrated **gradual recovery of facial weakness** with **conservative management**.

## 3. Discussion

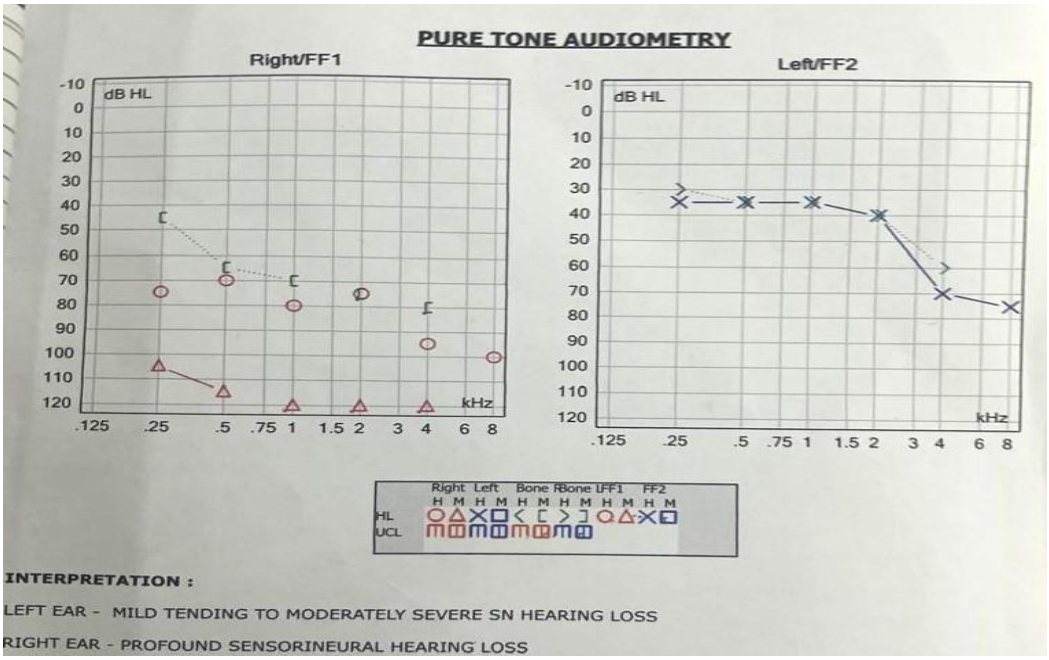
Superficial siderosis (SS) is a **rare neurodegenerative disorder** caused by **chronic subarachnoid bleeding** with subsequent **hemosiderin deposition**. This iron-laden pigment preferentially accumulates along the cerebellum,

brainstem, cranial nerves, and spinal cord, triggering neurodegenerative changes over time [6]




The classical triad of SS consists of **sensorineural hearing loss (**due to cranial nerve VIII involvement**), cerebellar ataxia (**due to cerebellar hemosiderin deposition**), and pyramidal tract signs** (from involvement of corticospinal tracts) [3]. This atypical presentation emphasizes that SS can occasionally manifest with isolated or transient cranial neuropathies before the full spectrum of symptoms develops.

In our case report, patient had complaints of decreased hearing in right ear since 2 days on performing audiometry report as below.

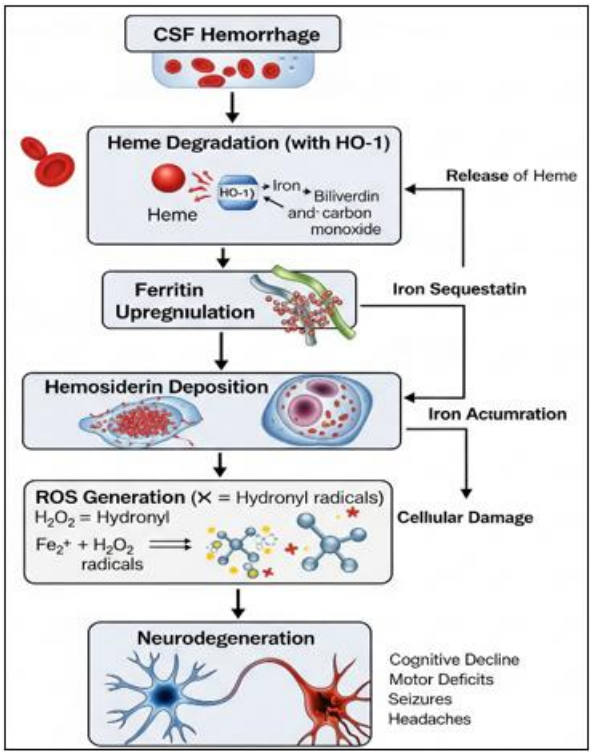



Our case distinguishes itself in two main unusual respects:

1) **Bilateral internal auditory canal (IAC) siderosis**, a relatively uncommon distribution in the absence of extensive cerebellar or spinal findings, is rarely described. Most reported cases of SS demonstrate widespread CNS siderosis rather than a focal IAC-predominant distribution.
2) **Transient facial palsy** as the initial manifestation is exceedingly unusual. While cranial nerve VIII involvement is common due to its long cisternal course and susceptibility to iron deposition [7], cranial nerve VII involvement is infrequent. When facial palsy occurs, it is usually permanent and associated with advanced disease [7,8]. Only isolated case reports describe **temporary or**

**reversible facial nerve palsy**, suggesting partial demyelination or conduction block rather than full axonal loss, explaining the favourable recovery with conservative management.

The **pathophysiology of SS** is linked to a cascade of iron-mediated neurotoxicity. Chronic bleeding into the subarachnoid space releases red blood cells, which break down and liberate heme. This heme is metabolized by heme oxygenase-1 (HO-1) in glial cells to free iron ($Fe^{2+}$), which is normally sequestered by ferritin. However, with persistent haemorrhage, ferritin storage becomes saturated, resulting in iron accumulation and formation of hemosiderin along CNS surfaces. Excess free iron catalyses the formation of reactive oxygen species via Fenton reactions, driving lipid peroxidation, protein and DNA damage, gliosis, and eventual neuronal loss. Purkinje cells and the vestibulocochlear nerve (cranial nerve VIII) are particularly susceptible to this oxidative injury. (6)




Cranial nerves VII and VIII are more prone to early involvement because of their **long intracranial course** and **prolonged exposure to CSF**, which leaves them vulnerable to iron-mediated toxicity and its extensive central myelin lining, which is vulnerable to damage from siderosis-susceptible microglia. [5,8].

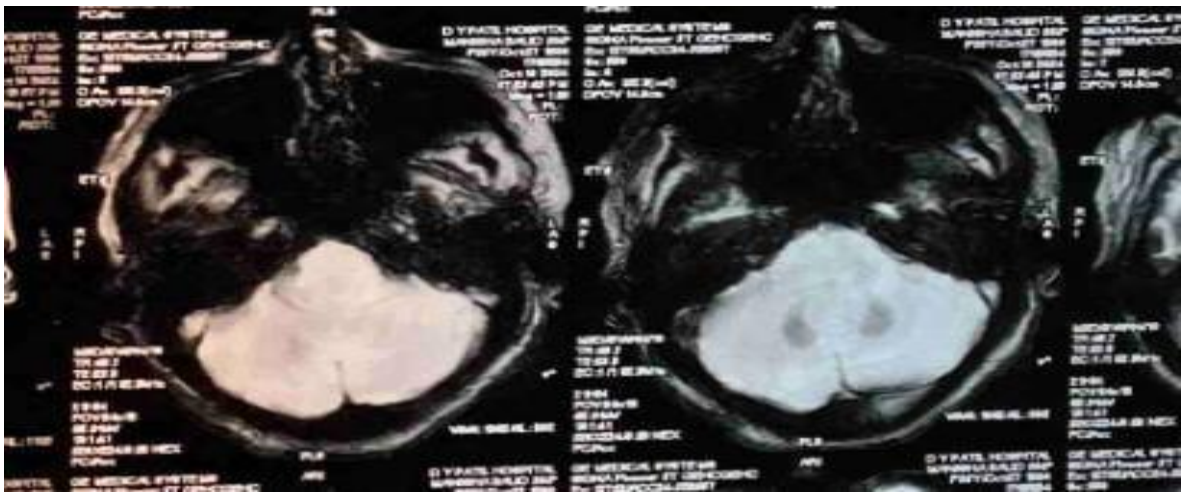
House-Brackmann facial nerve grading system is concise, rapid, subjective and discontinuous scale which classifies the facial weakness as below. [7]

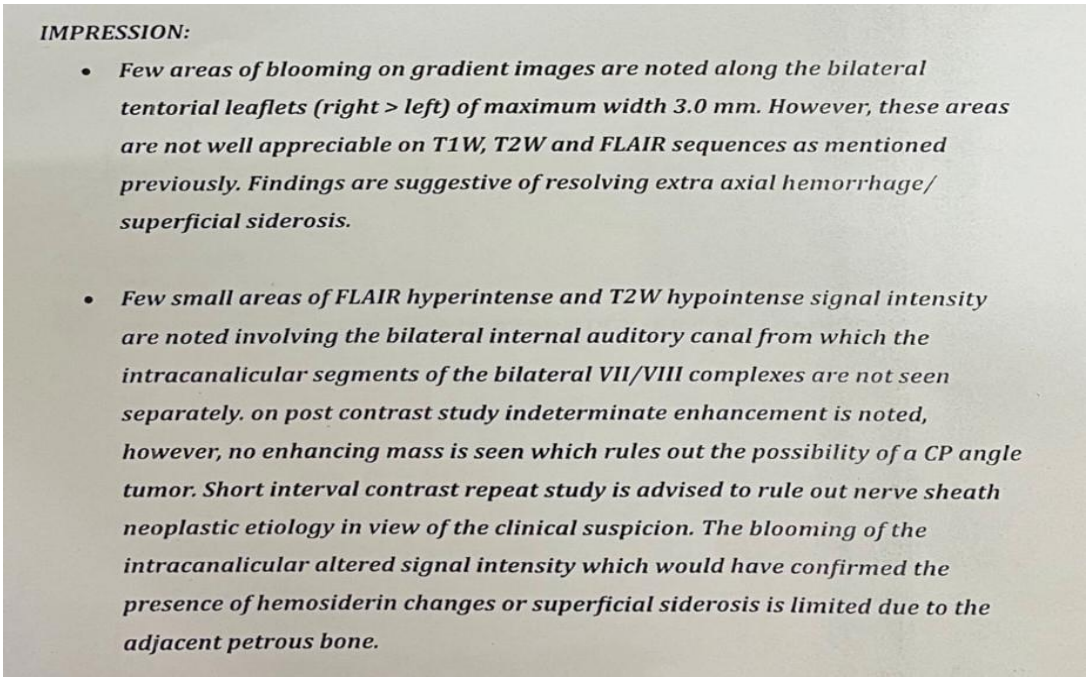
| Grade | Description | Characteristics |
|---|---|---|
| I | Normal | Normal facial function in all areas |
| II | Mild dysfunction | Gross: slight weakness noticeable on close inspection; may have very slight synkinesis<br>At rest: normal symmetry and tone<br>Motion: Forehead—moderate-to-good function<br>Eye—complete closure with minimum effort<br>Mouth—slight asymmetry |
| III | Moderate dysfunction | Gross: obvious but not disfiguring difference between two sides; noticeable but not severe synkinesis, contracture, and/or hemifacial spasm<br>At rest: normal symmetry and tone<br>Motion: Forehead—slight-to-moderate movement<br>Eye—complete closure with effort<br>Mouth—slightly weak with maximum effort |
| IV | Moderately severe dysfunction | Gross: obvious weakness and/or disfiguring asymmetry<br>At rest: normal symmetry and tone<br>Motion: Forehead—none<br>Eye—incomplete closure<br>Mouth—asymmetric with maximum effort |
| V | Severe dysfunction | Gross: only barely perceptible motion<br>At rest: asymmetry<br>Motion: Forehead—none<br>Eye—incomplete closure<br>Mouth—slight movement |
| VI | Total paralysis | No movement |

According to this standard grading scale we examined the facial nerve weakness symptoms and documented as below.

Facial nerve examination when patient presented and follow up when improvement was seen in facial weakness. **MRI**, particularly **susceptibility-weighted imaging (SWI)** or **gradient-echo (GRE) sequences**, remains the **diagnostic gold standard**. These sequences demonstrate the characteristic hypointense rim caused by hemosiderin deposition. Notably, the **extent of radiological siderosis does not always correlate with clinical severity** , especially in early or atypical presentations [6].

In this patient MRI Scan and report suggestive of enhancement in B/L internal auditory canal as below.




IMPRESSION:

- Few areas of blooming on gradient images are noted along the bilateral tentorial leaflets (right > left) of maximum width 3.0 mm. However, these areas are not well appreciable on T1W, T2W and FLAIR sequences as mentioned previously. Findings are suggestive of resolving extra axial hemorrhage/ superficial siderosis.

- Few small areas of FLAIR hyperintense and T2W hypointense signal intensity are noted involving the bilateral internal auditory canal from which the intracanalicular segments of the bilateral VII/VIII complexes are not seen separately. on post contrast study indeterminate enhancement is noted, however, no enhancing mass is seen which rules out the possibility of a CP angle tumor. Short interval contrast repeat study is advised to rule out nerve sheath neoplastic etiology in view of the clinical suspicion. The blooming of the intracanalicular altered signal intensity which would have confirmed the presence of hemosiderin changes or superficial siderosis is limited due to the adjacent petrous bone.

**Management** of SS is primarily directed at **identifying and controlling the bleeding source**, which may involve surgical repair of dural defects or endovascular intervention.

**Iron chelation therapy**, such as **oral deferiprone (15–30 mg/kg/day with CBC monitoring)**, has been explored to reduce CNS iron burden and stabilize neurological function [8]. Supportive measures, including **facial physiotherapy, audiology evaluation, and rehabilitation**, play an important role in symptom management.

In our patient, **conservative therapy** led to **gradual improvement in facial weakness** and **partial hearing recovery**. This highlights that, in select cases with minimal or reversible neuronal injury, non-surgical management can yield functional improvement.

## 4. Conclusion

Superficial siderosis of the central nervous system is an uncommon but clinically significant cause of progressive cranial neuropathies. This case highlights an **atypical presentation with bilateral internal auditory canal involvement and transient facial palsy**, rather than the classical triad of sensorineural hearing loss, ataxia, and pyramidal signs. Early recognition with **MRI**, coupled with **timely evaluation for potential bleeding sources**, is crucial to prevent irreversible neurological damage. Even in rare presentations such as ours, **conservative management can lead to meaningful symptomatic improvement** when neuronal injury is incomplete.

**Declaration of Patient Consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for including images and other clinical information to be reported in the journal. The patient understands that her names and initial's will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

**Conflicts of Interest**
There are no conflicts of interest.

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