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Surgical Neurology International

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SNI: Epilepsy

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Case Report

Classical type of superficial hemosiderosis presenting with temporal lobe epilepsy

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Received: 13 February 2025
Accepted: 08 May 2025
Published: 06 June 2025

DOI
10.25259/SNI_153_2025

Quick Response Code:

ABSTRACT

Background: Classical type of superficial hemosiderosis (SH) is subpial hemosiderin deposition mainly affecting the cerebellum, brainstem, and spinal cord, which generally presents with cerebellar ataxia and sensorineural hearing disturbance. We here report a rare case of the classical type of SH presenting with temporal lobe epilepsy and perform a literature review on similar cases.

Case Description: A 63-year-old man with four episodes of impaired awareness and confusion lasting for around 5 minutes after feeling vague uneasiness, suggesting focal impaired awareness seizure, visited a neurosurgical clinic. T2*-weighted magnetic resonance imaging (MRI) showed hemosiderin deposition on the surface of the cerebellum, brainstem, upper spinal cord, and bases of bilateral frontal and temporal lobes. Neurological examination found mild gait ataxia and anosmia. Audiogram showed sensorineural high-frequency hearing loss. Electroencephalogram showed rhythmic theta activities accompanied by intermittent sharp waves over the right fronto-temporal region during a subclinical seizure episode, which led to the diagnosis of temporal lobe epilepsy. Up-dosing of levetiracetam to 1,500 mg/day brought about a seizure-free status. Gait disturbance, however, gradually deteriorated over the following 6 months. Spinal MRI and myelogram found a dural defect at the T3 level. The 4 mm long defect was surgically closed, which led to the gradual improvement of the gait ataxia.

Conclusion: In this case of the classical type of SH due to a dural defect, temporal lobe epilepsy is presumably caused by the neurotoxicity of decomposed products of hemoglobin impregnated in the temporal lobes.

Keywords: Anosmia, Ataxia, Hearing disturbance, Superficial hemosiderosis, Temporal lobe epilepsy

INTRODUCTION

Superficial hemosiderosis (SH) is a central nervous system (CNS) disease caused by subpial hemosiderin deposition secondary to chronic hemorrhage in the subarachnoid space.[4,10-12,14,25] Secondary or type 2 SH shows local cerebral hemosiderin deposition around the bleeding site due to various pathologies, including cerebral amyloid angiopathy and arteriovenous malformation.[14,25] Whereas classical or type 1 SH shows diffuse hemosiderin deposition mainly

on the infratentorial CNS structures, such as the cerebellum and brainstem, and also on the upper spinal cord.[12,14,25] Recently, spinal dural diseases, especially dural defects, have emerged as the primary etiology of the classical SH.[12-14,25] Classical SH generally presents with slowly progressive cerebellar ataxia, hearing impairment, spinal cord signs, and cognitive impairments.[4,14,16,19,25] However, secondary epilepsy due to classical SH is very rarely reported. It is not referred to as one of the typical clinical features of classical SH in recent reviews or large case series.[4,14,16,19,25] We herewith describe a case of classical SH presenting with local epilepsy presumably resulting from the deposition of decomposed hemoglobin products over the temporal lobes. We also perform a literature review on this rare condition.

CASE DESCRIPTION

An otherwise healthy 63-year-old man, a farmer, visited a neurosurgical clinic with four episodes of impaired awareness and confusion lasting for around 5 minutes after feeling vague uneasiness. He looked around during these episodes. He also had some 10-minute-long memory disturbances after the episodes. No apparent automatic movements during the episodes were reported. The episodes suggested that he had focal impaired awareness seizures (FIAS) attacks. Abnormalities found on magnetic resonance imaging (MRI) at the clinic urged further examination at our neurosurgical center. Neurological examination showed mild cerebellar ataxia, hearing disturbance, anosmia, and taste impairment. Audiogram showed bilateral sensorineural high-frequency hearing loss. The Mini-Mental State Examination score was 28/30. T2*-weighted MRI found thick deposition of hemosiderin on the surface of the cerebellum, brainstem, upper spinal cord, and bases of the bilateral frontal and temporal lobes [Figure 1]. Hippocampal sclerosis was not observed. Three-dimensional computed tomography (CT) angiography found no vascular abnormalities causative of intracranial bleeding. Electroencephalography (EEG) showed background activity consisting of a posterior dominant 8–9 Hz alpha rhythm, reactive to eye opening. There were occasional low-amplitude spike-like discharges over the right and left temporal regions during the drowsy state. During the light sleep state, 8 Hz alpha-like rhythmic discharges appeared predominantly over the right fronto-temporal region at F8 and F4. These discharges built up to rhythmic theta activity spreading to the right hemisphere, which was accompanied by intermittent sharp waves maximal at F8 [Figure 2]. This EEG activity suggested a subclinical seizure that may be originating from the right temporal region.

He was given an increasing dose of levetiracetam up to 1,500 mg/day, which prevented further epilepsy attacks. However, gait disturbance slowly progressed, eventually disturbing even daily activities. Sagittal T2*-weighted spinal MRI showed the deposition of hemosiderin over the upper thoracic spinal cord [Figure 3a] and osteophytes at the T3–

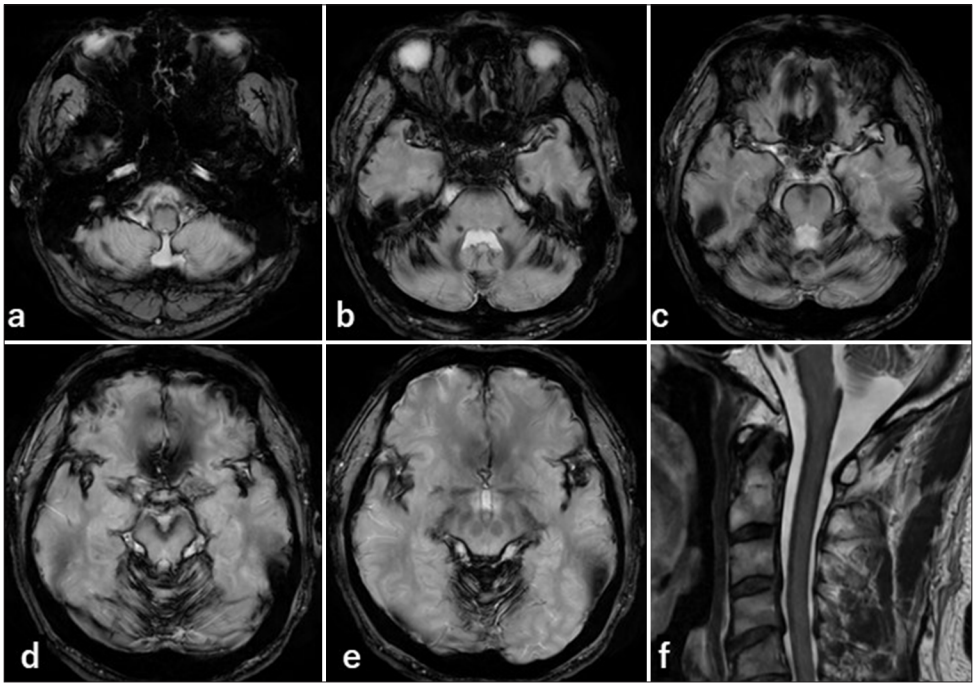

Figure 1: T2*-weighted magnetic resonance imaging of the brain and the upper cervical cord at the presentation. (a-e) It showed low-intensity rims, hemosiderin deposition, on the infratentorial structures including cerebellum and the base of supratentorial structures including the temporal lobes. (f) It also showed hemosiderin deposition on the surface of the upper cervical cord.

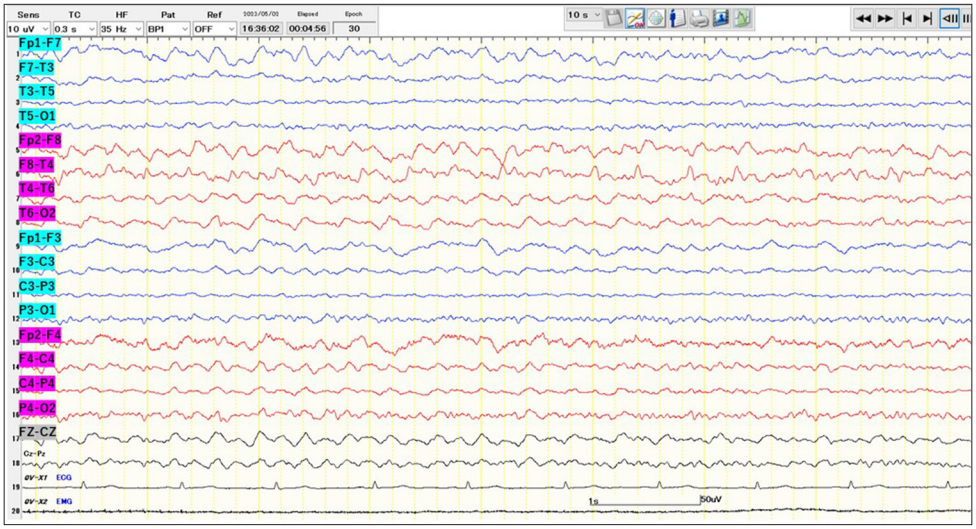

Figure 2: Longitudinal bipolar electroencephalography during light sleep showed high-amplitude rhythmic theta activity in the right fronto-temporal region, accompanied by intermittent sharp waves maximal at F8, suggesting a subclinical seizure. Blue lines are from the left-side electrodes. Red lines are from the right-side electrodes.

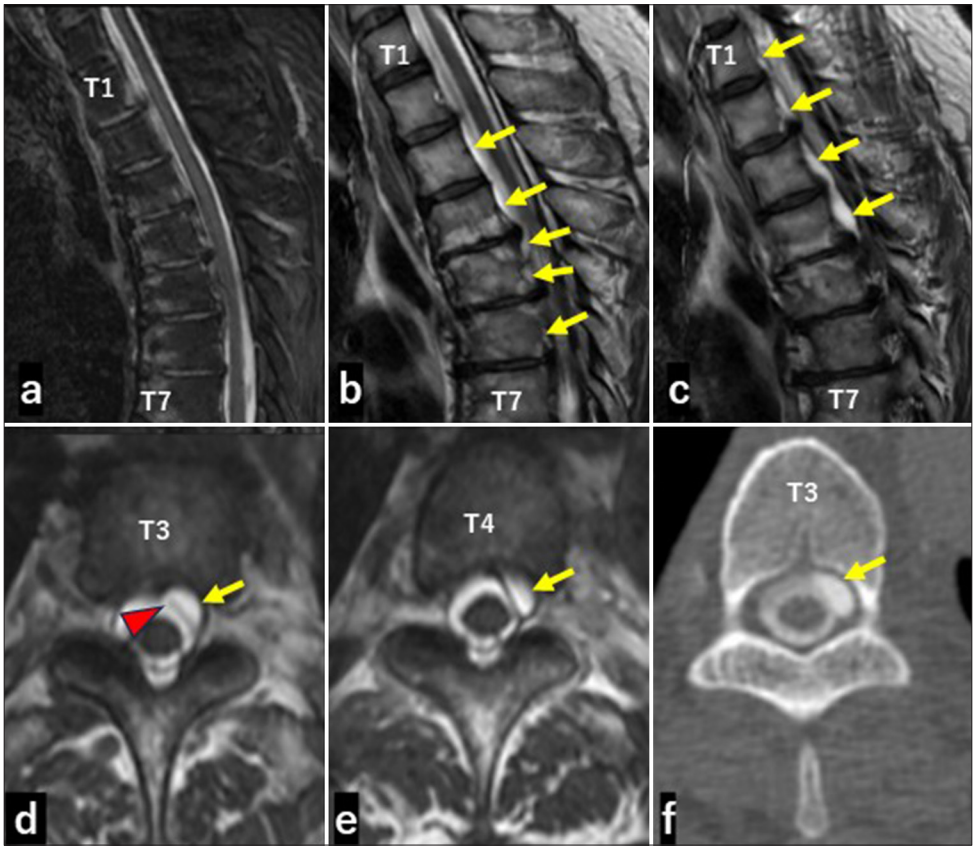

Figure 3: Neuroimaging of the thoracic spine. (a) T2*-weighted midline sagittal magnetic resonance imaging (MRI) of the thoracic spine showed hemosiderin deposition on the upper thoracic spinal cord and osteophytes at the T3-T4 and T4-T5 disc levels. (b, c) T2*-weighted off-midline sagittal MRI showed fluid collection outside the dura mater at the T1-T6 level (arrows). (d, e) Axial constructive interference in steady state-weighted MRI showed a left antero-lateral extradural fluid collection at the T3 and T4 levels (arrow). It also showed a dural defect at the T3 level (arrowhead in d). (f) A computer tomographic myelogram showed that the contrast material in the cerebrospinal fluid leaked into the extradural space (arrow).

T4 and T4–T5 disc levels. Left para-midline sagittal scans showed an extradural fluid collection at the T1–T6 level [Figure 3b and c]. Axial constructive interference in steady state-MRI showed a left antero-lateral extradural fluid collection at the T3 [Figure 3d] and T4 [Figure 3e] levels. It also suggested a dural defect at the lower T3 vertebral body level [Figure 3d, arrowhead]. A CT myelogram confirmed that the extradural fluid space communicated with the subarachnoid space through the defect [Figure 3f]. He underwent T2–T4 level laminectomy. Microscopic and endoscopic exploration found a 4 mm long dural defect [Figure 4a and b]. A piece of collagen matrix graft (DuraGen®, Codman, Raynham, Massachusetts) was inserted into the extradural space through the defect. A 9–0 stitch approximated the edges of the dural defect. The defect was then covered by another piece of DuraGen [Figure 4c]. The postoperative course was uneventful, and MRI showed the disappearance of the extradural CSF space [Figure 4d].

The unsteadiness of the gait gradually improved since the surgery. He returned to rice farming 4 months after the surgery. FIAS attacks have not recurred for the past 19 months.

Literature review

In almost all large case series and past literature reviews, seizures were not mentioned as a symptom of SH.[4,14,16,19,25] Only a report published in 2006 on 30 case series treated at the Mayo Clinic described "Other symptoms that were felt to be possibly related to the SH included seizures in four."[11] However, details of these four cases were not given.

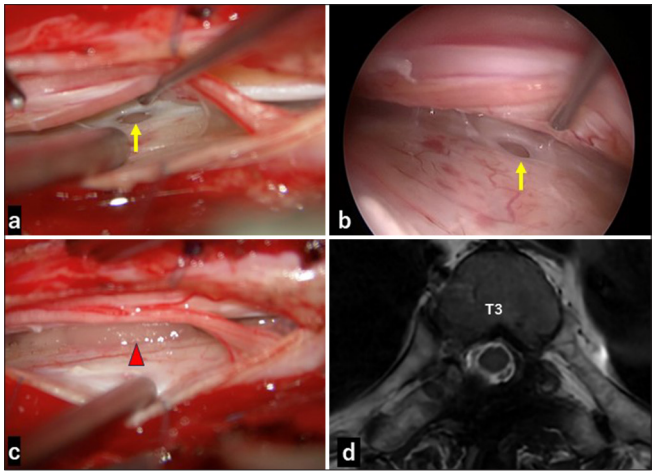

Figure 4: Intraoperative exploration and postoperative magnetic resonance imaging (MRI). (a) Microscopic exposure at the lower T3 level showed the dural defect (arrow). (b) Endoscopic observation confirmed the defect (arrow). (c)The defect was sealed using collagen matrix grafts and a stitch (arrowhead). (d) Postoperative axial constructive interference in steady state-weighted MRI revealed the disappearance of the extradural cerebrospinal fluid collection.

Our literature retrieval, however, found 11 reported cases (ten males and one female), including ours, in which seizures are considered to be due to SH [Table 1].[2,3,8,9,18,20,21,24,26] Two other case reports, including one in which the seizure could be attributable to multiple brain cavernomas rather than SH and one from an otolaryngology journal in which no details of the seizure, EEG, or MRI findings were given, were excluded from this review.[5,15]

The median age of these 11 patients was 55 years old (range: 12–69). Partial seizures, described as simple partial seizures, FIAS, headache, visual hallucination, and blanking spells, were seen in six. Convulsive seizures, described as generalized tonic-clonic seizures, loss of consciousness with convulsion, limb rigidity, and secondary generalization, were seen in six. The epileptic focus on EEG was in the temporal leads in six patients, and not described in five. SH on the temporal lobes on MRI was observed in all patients. At least one classical symptom of SH, such as hearing impairment and gait ataxia, was recorded in all patients. Past medical history regarding CNS disease was recorded in six patients, including surgeries on CNS tumors, head injuries, cervical injury, cerebellar stroke, and radiation. The cause of SH was reported to be a spinal dural defect in two patients, past spinal tumor surgery in one, past treatment for cerebellar medulloblastoma in one, and unknown in the other seven.

DISCUSSION

This is a case of SH who initially presented with FIAS or complex partial seizure. EEG showed a right temporal focus. Spinal MRI found a T3 level dural defect, the surgical closure of which led to a gradual improvement of ataxia.

SH is a rare disorder characterized by subpial diffuse hemosiderin deposition caused by prolonged, slow subarachnoid hemorrhage on the surface of the brain and spinal cord.[4,11,12,14,25] It can be detected as a hypointensity rim by high-resolution MRI machines using T2- or T2*-weighted sequences. Infratentorial structures, such as the cerebellum, brainstem, upper spinal cord, and cranial nerves, are mainly affected, but supratentorial structures, such as the basal cortices of temporal and frontal lobes, are also involved.[11,14,25] The prolonged supply of neurotoxic heme, a breakdown product of hemoglobin, on the surface of the neuronal structures causes various neuronal damages.[10,14,25] The most common symptoms of SH are cerebellar ataxia, sensorineural hearing disturbance, and myelopathy, reflecting the prevalent parts of hemosiderin deposition on the CNS.[4,14,16,19] Anosmia and cognitive impairment may also be present, reflecting its deposition on the medial and inferior surfaces of the frontal lobes.[4,14,16,19,25] While seizures are not referred to as a presentation of SH in major past reviews and large case series, only one case series mentioned that seizures were seen in four out of their 30 patients, without details.[11]

Table 1: Reported cases of classical superficial siderosis (SS) presenting with epilepsy

Author	Ref. No.	Year	Country	Age / Sex	Epilepsy type	Epileptic focus on EEG	Other symptoms or signs due to SH	Temporal lobe SS	Past CNS medical history	Etiology of SS
O'Riordan JI	20	1996	Ireland	34/M	Simple partial seizure	ND	Hearing impairment, anosmia	yes	Head injury	Unknown
Iannaccone S	8	1999	Italy	33/M	Headache, LOC, right limbs rigidity	Left temporal	Deafness, gait ataxia	yes	Severe head injury	Unknown
Wang K	24	2010	China 18	27/M	LOC with convulsions	Right temporal	Hearing loss, myelopathy	yes	no	Unknown
Machino Y	18	2010	Japan	62/M	FIAS GTCS	Left temporal	Ataxia, anosmia, hearing loss	yes	Spinal tumor	Past spinal tumor surgery
Jadhav TM	9	2012	India	12/M	Blanking spells	ND	Dysarthria, hearing loss, ataxia, memory disturbance	yes	Cerebellar medulloblastoma	Surgery and radiation
Chen CY	2	2015	China	58/M	GTCS	left temporal	Slight gait ataxia	yes	Radiation for pharyngeal ca.	Unknown
Petelin Gadze Z	21	2018	Croatia	69/M	Visual hallucination secondary generalization	Right temporo-occipital	Hearing loss, hyposmia, ataxia, myelopathy	yes	Head injury cerebellar stroke	Unknown
Xu L	26	2021	China	29/M	Headache, GTCS	ND	Hearing loss, ataxia, memory disturbance	yes	Cervical injury	Spinal dural defect
Deng L	3	2024	China	67/F	ND	ND	Hearing impairment, ataxia, hyposmia, cognitive impairment	yes *	ND	Unknown
Deng L	3	2024	China	55/M	ND	ND	Hearing impairment, ataxia, hyposmia	yes *	ND	Unknown
Our Case		2025	Japan	63/M	FIAS	Right temporal	Ataxia, anosmia, hearing impairment	yes	no	Spinal dural defect

ca.: Carcinoma, CNS: Central nervous system, EEG: Electroencephalogram, F: Female, FIAS: Focal impaired awareness seizure, GTCS: Generalized tonic clonic seizure, LOC: Loss of consciousness, M: Male, ND: Not described, Ref.: Reference, SS: Superficial siderosis, *: Deposition of hemosiderin on the supra- and infratentorial structures

However, our literature survey found 11 reported cases, including ours, of SH presenting with seizures.[2,3,8,9,18,20,21,24,26] Among these 11, seizure semiology was described in eight patients: simple partial seizures, such as FIAS in six patients, and convulsive seizures in six patients. Hemosiderin deposition was seen on the surface of the temporal lobe in all patients. Among the six patients with a described epileptic focus on EEG, all were on the temporal lobe.

A recent report showed that post-stroke cerebral SH is far more common in patients with post-stroke epilepsy (PSE) than in patients without PSE.[23] Post-stroke cerebral SH is independently related to PSE.[23] Another study showed that hemosiderosis was significantly correlated with epilepsy following subarachnoid hemorrhage.[7] Thus, it is plausible that the temporal lobe structure surrounded by the thick hemosiderin rim can become epileptogenic due to the neurotoxicity and irritancy of decomposition products of hemoglobin,[10,14,25] even though apparent hippocampal sclerosis on MRI was not recorded in any past reports, including ours. Meticulous history taking may find more epilepsy cases in patients with SH presenting with other more common symptoms. Furthermore, it may be an acceptable idea to include T2*-weighted sequences in routine MRI surveillance to find cortical hemosiderosis, including SH, in patients with focal epilepsy.

Although secondary or type 2 SH shows local hemosiderin deposition around the bleeding site due to various pathologies,[17,19,25] classical or type 1 SH shows diffuse hemosiderin deposition mainly on the infratentorial CNS structures, as in our case. According to a literature review by Levy *et al*. published in 2007, CNS tumors were the most common etiology (15%), followed by head trauma (13%) and vascular malformations (9%), but 35% were designated as "idiopathic."[16] Meanwhile, Kumar reported fluid-filled collections in the spinal canal found in 14 out of 30 patients of SH, some of which were due to dural tears.[11] Later, he introduced a concept of "duropathy" as an etiological background of SH.[13] A recent series of 48 cases of classical SH from the National Hospital for Neurology and Neurosurgery in the United Kingdom reported dural abnormalities, including dural tears, traumatic nerve root avulsion, or postoperative pseudomeningocele as an etiology in 40 patients (83%).[25] A multi-institutional study in Japan, including 56 classical type SH patients, found that 50% of the patients had cystic lesions or dural abnormalities in the spinal canal.[19] A four-dimensional dynamic CT myelography study on 20 SH patients with ventral fluid-filled collection in the spinal canal demonstrated dural defects in all, mainly at the upper thoracic spine.[6] Takai and Taniguchi reported the repair of dural defects in seven patients presenting with spontaneous cerebrospinal fluid leaks accompanied by SH.[22] They found continuous bleeding from the epidural veins. These recent pieces of evidence suggest that the majority of "idiopathic" classical SH patients have an extradural fluid-filled collection in the spinal canal, which is caused by dural tears. Moreover, slow continuous bleeding from the epidural space results in hemosiderin deposition on the upper spinal cord, infratentorial CNS structures, and the base of the cerebrum. Closure of the defect may deter the aggravation and even bring about improvement of SH symptoms,[1] as was achieved in our case.

CONCLUSION

We reported a rare case of SH presenting with a simple partial seizure with a right temporal focus. An anti-epileptic drug has well-controlled the seizures. He also showed common symptoms of SH, such as hearing disturbance and gait ataxia. The progression of gait ataxia ceased after the repair of a dural tear at the T3 level. This report stresses the importance of meticulous history taking about symptoms suggesting partial seizures in SH patients and searching for the dural tear in the spinal canal for the quick and proper treatment of SH. In addition, this report suggests the routine check-up of brain hemosiderosis, including SH, using T2*-weighted MRI to find the etiology in patients with simple partial seizures.

Acknowledgment: The authors would like to thank Professor Koji Iida, Department of Epileptology, Hiroshima University Hospital, for the EEG reading and Haibunsha Kagoshima for performing a critical review of this manuscript and English language editing.

Ethical approval: The Institutional Review Board approval is not required.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship: Nil.

Conflicts of interest: There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation: The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Arita K, Yokota K, Nagano Y, Yamahata H, Higa N, Yamamoto M, *et al*. Classical type of superficial hemosiderosis presenting with temporal lobe epilepsy. Surg Neurol Int. 2025;16:225. doi: 10.25259/SNI_153_2025