

Epilepsy as an Initial Presentation of Superficial Siderosis of the Central Nervous System:A Case Report

Ruohan Ma

The Third Hospital of Hebei Medical University

Jialei Sun

The Third Hospital of Hebei Medical University

Weigang Luo

The Third Hospital of Hebei Medical University

Guisong Zhang

The Third Hospital of Hebei Medical University

Huiling Ren

renhui ling@hebmh.edu.cn

The Third Hospital of Hebei Medical University

Case Report

Keywords: Superficial Siderosis of the Central Nervous System, Epilepsy, Brachial Plexus Injury

Posted Date: October 18th, 2026

DOI: <https://doi.org/10.21203/rs.3.rs-11018629/v1>

License:   This work is licensed under a Creative Commons Attribution 4.0 International License.

[Read Full License](#)

Additional Declarations: No competing interests reported.

Abstract

Background

Superficial siderosis of the central nervous system (SSCNS) is a rare neurological disorder resulting from chronic subarachnoid haemorrhage. Its classical clinical features comprise sensorineural hearing loss, cerebellar ataxia and pyramidal tract dysfunction. However, epilepsy as an initial presentation remains uncommon and insufficiently characterized.

Case Presentation:

We describe a 34-year-old male with a remote history of right brachial plexus injury, who presented with recurrent epileptic seizures as the predominant early symptom, followed by progressive hearing loss and gait instability. Neuroimaging demonstrated characteristic hemosiderin deposition on T2*-weighted and susceptibility-weighted imaging (SWI) sequences. Transient, seizure-related subcortical and periventricular restricted diffusion on diffusion-weighted imaging resolved completely within one month. Anti-epileptic medication achieved partial seizure control, but the patient's neurological deficits exhibited gradual progression.

Conclusion

Epilepsy may represent an uncommon initial presentation of SSCNS. Targeted MRI sequences facilitate diagnosis. SSCNS should be considered in the differential diagnosis of patients with unexplained epilepsy, particularly those with a history of craniospinal trauma.

Background

Superficial siderosis of the central nervous system (SSCNS) is a rare, chronic neurological disorder caused by recurrent or persistent subarachnoid haemorrhage, leading to progressive deposition of hemosiderin along the leptomeninges, brainstem, cerebellum, spinal cord and cranial nerves^[1]. The classical clinical triad comprises progressive sensorineural hearing loss, cerebellar ataxia and pyramidal tract dysfunction, which is present in approximately 80% of affected individuals^[2]. Epilepsy is a rare manifestation, reported in fewer than 10% of cases, and its role as an initial presenting feature remains under-recognised in clinical practice^[3]. Brachial plexus injury is a well-established cause of secondary SSCNS, accounting for 10–15% of all cases^[4]. The underlying mechanism involves chronic, low-grade micro-haemorrhage from post-traumatic dural defects, pseudomeningoceles or traumatic diverticula, rather than a single acute haemorrhage^[5]. Over years or decades, repeated erythrocyte degradation results in excessive hemosiderin accumulation, leading to oxidative stress, gliosis, axonal damage and progressive neurodegeneration^[6].

Here, we report a rare case of SSCNS in which epilepsy preceded the development of classical neurological features. This case highlights an clinical presentation and provides further clinical observations regarding the possible relationship between chronic haemosiderin deposition and epileptic seizures. [7].

Case Presentation

A 34-year-old right-handed male manual worker was admitted with recurrent generalised epileptic seizures and persistent neck and lumbar pain. Eighteen years prior to admission, he had sustained a right brachial plexus injury in a road traffic accident. Four years before presentation, he underwent two cervical spine procedures for persistent pain. No formal diagnosis of epilepsy had been established before admission, and he received no standardized anti-seizure therapy; there was no history of toxic exposure. Fourteen years before admission, the patient began to experience recurrent episodes of neck and lumbar pain associated with headache, nausea and vomiting. Severe attacks were accompanied by generalised convulsions and impaired consciousness. Seizure frequency increased gradually despite treatment with analgesics and various antiepileptic medications. In the two months preceding admission, seizures occurred daily, accompanied by severe headache, nausea, vomiting and diaphoresis. Symptoms were partially relieved by lying in the lateral decubitus or kneeling-crawling position. During admission, two episodes of status epilepticus occurred, each lasting more than five minutes, successfully terminated with intravenous diazepam (10 mg) and intramuscular phenobarbitone sodium. Post-ictally, the patient had a transient reduced conscious level.

DWI obtained immediately after seizures showed bilateral symmetrical periventricular and subcortical restricted diffusion, which resolved completely within one month, consistent with seizure-related cytotoxic oedema [8](Fig. 1). Brain MRI revealed marked hypointensities along the surfaces of the brainstem, pons and cerebellum on T2*-weighted and SWI sequences [9](Fig. 2). Cervical MRI demonstrated linear hypointensities surrounding the spinal cord, in keeping with extensive superficial haemosiderin deposition [10](Fig. 3).

(A) 3 d pre-seizure: No restricted diffusion

(B) Seizure onset day: Bilateral symmetrical restricted diffusion

(C) 3 d post-seizure: Expanded restricted diffusion

(D) 10 d post-seizure: Gradual signal resolution

(E) 17 d post-seizure: Further signal resolution

(F) 1 mo post-seizure: Complete resolution of abnormal signals

(A-F) Upper row: DWI; lower row: corresponding ADC maps. Corresponding to the temporal evolution of seizure-associated imaging abnormalities.

EEG showed slow background activity with continuous, diffuse, regular symmetric or asymmetric mixed slow waves of 2–4 Hz with moderate to high amplitude over bilateral anterior head regions, consistent with nonspecific encephalopathic changes.

Cerebrospinal fluid (CSF) examination showed normal opening pressure (130 mmH₂O). Routine biochemistry, microbiological studies, autoimmune encephalitis antibodies and next-generation sequencing for pathogens were all unremarkable. Repeated electroencephalography (EEG) demonstrated diffuse slow wave activity but no definite epileptiform discharges.

The patient was commenced on a combination of levetiracetam, sodium valproate and topiramate for seizure control. Concomitant treatment included agents to reduce intracranial pressure, relieve cerebral vasospasm, and provide neuroprotection, antioxidant support and low-dose corticosteroids. Despite comprehensive medical management, his neurological deficits progressed slowly over time.

Discussion and Conclusions

This case illustrates an atypical presentation of SSCNS, in which recurrent epilepsy preceded the development of classical neurological features. Such presentations are rare and may lead to delayed diagnosis unless clinicians maintain a high index of suspicion ^[2, 3]. The remote brachial plexus injury was considered a possible contributing factor for chronic subarachnoid micro-haemorrhage, consistent with previous reports of traumatic dural defects as a precursor to SSCNS ^[4, 11].

The transient bilateral symmetrical periventricular and subcortical DWI abnormalities following seizures represents an uncommon imaging observation in this patient with SSCNS.. These changes are believed to reflect seizure-induced cytotoxic oedema, exacerbated by chronic iron-mediated neurotoxicity and increased subcortical white matter vulnerability. This pattern differs from typical ischaemic lesions, which follow vascular territories and do not resolve rapidly, and helps to distinguish seizure-related changes from structural or inflammatory pathology ^[7, 8].

The pathophysiology of epilepsy in SSCNS is multifactorial. Chronic haemosiderin deposition causes direct meningeal and cortical irritation. Free iron released via the Fenton reaction generates reactive oxygen species, inducing neuronal membrane instability, lipid peroxidation and DNA damage, thereby lowering the seizure threshold. Reactive gliosis further disrupts synaptic transmission and promotes neuronal hyperexcitability ^[1, 6].

Early diagnosis of SSCNS may help identify potential bleeding sources and allow consideration of appropriate management strategies. ^[5] SWI and T2*-weighted MRI are the gold-standard imaging modalities for detecting superficial haemosiderin deposition, even before classical clinical features develop ^[7]. Antioxidant and iron-chelating treatments, such as deferiprone, represent promising adjuvant approaches in the management of SSCNS and require further formal clinical evaluation ^[10].

Key differential diagnoses included ischaemic stroke, posterior reversible encephalopathy syndrome (PRES), mitochondrial encephalopathy and autoimmune encephalitis. The absence of vascular territory lesions, normal inflammatory markers, negative CSF studies and resolution of DWI changes effectively excluded these conditions ^[8]. The characteristic imaging appearance on SWI and T2*-weighted sequences confirmed the diagnosis of SSCNS ^[9].

This case emphasises that SSCNS should be included in the differential diagnosis of patients with unexplained recurrent epilepsy, particularly those with a history of remote craniospinal trauma, brachial plexus injury or spinal surgery. Recognition of epilepsy as an early manifestation, supported by targeted MRI sequences including SWI and T2*-weighted imaging, may facilitate earlier recognition and appropriate clinical management. ^[1, 7, 9].

Limitations

This report has several limitations. First, conclusions are based on a single patient, and therefore the relationship between epilepsy and SSCNS cannot be established as causal. Second, larger studies are required to determine the frequency and clinical significance of epilepsy as a presentation of SSCNS. Finally, the optimal management strategy for epilepsy associated with SSCNS remains uncertain.

Learning Points

- Epilepsy may occur as an uncommon initial presentation of superficial siderosis of the central nervous system.
- Remote brachial plexus injury and craniospinal trauma may represent risk factors for secondary SSCNS.
- T2*-weighted and SWI MRI sequences are the gold standard for diagnosing superficial siderosis.
- Transient reversible subcortical and periventricular restricted diffusion on DWI may occur after seizures in SSCNS patients.
- Early diagnosis is critical to identify the bleeding source and consider interventions to slow disease progression.

Abbreviations

ADC Apparent Diffusion Coefficient

CSF Cerebrospinal Fluid

DWI Diffusion-Weighted Imaging

EEG Electroencephalography

PRES Posterior Reversible Encephalopathy Syndrome

SSCNS Superficial Siderosis of the Central Nervous System

SWI Susceptibility-Weighted Imaging

T2-WI T2-weighted Imaging

T2*-WI T2*-weighted Imaging

Declarations

Ethics approval and consent to participate

Ethical approval was not required for this case report in accordance with local and national guidelines, as it describes a single patient and involves no experimental intervention. Written and oral informed consent for participation and publication was obtained from the patient.

Consent to publish

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare no competing interests.

Author contributions.

RHM and HLR contributed to the study design. RHM conducted the literature search. RHM and JLS acquired the data. RHM and HLR wrote the article. All authors contributed to revising the manuscript critically for important intellectual content. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work.

Funding

This research received no external funding.

Author Contribution

Author contributions. RHM and HLR contributed to the study design. RHM conducted the literature search. RHM and JLS acquired the data. RHM and HLR wrote the article. All authors contributed to revising the manuscript critically for important intellectual content. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work.

Acknowledgement

Not applicable.

Availability of Data and Materials

All data included in this study are available from the corresponding author upon reasonable request.

References

1. Fearnley JM, Stevens JM, Rudge P. Superficial siderosis of the central nervous system. *Brain*. 1995;118 (Pt 4):1051–66.
2. Kumar N. Superficial Siderosis: A Clinical Review. *Ann Neurol*. 2021;89(6):1068–79.
3. Pichler M, Vemuri P, Rabinstein AA, Aakre J, Flemming KD, Brown RD, Jr., et al. Prevalence and Natural History of Superficial Siderosis: A Population-Based Study. *Stroke*. 2017;48(12):3210–4.
4. Bonito V, Agostinis C, Ferraresi S, Defanti CA. Superficial siderosis of the central nervous system after brachial plexus injury. Case report. *J Neurosurg*. 1994;80(5):931–4.
5. Aquilina K, Kumar R, Lu J, Rawluk D. Superficial siderosis of the central nervous system following cervical nerve root avulsion: the importance of early diagnosis and surgery. *Acta Neurochir (Wien)*. 2005;147(3):291–7; discussion 7.
6. Xu Q, Wen S, Li H, Yang G, Di M. Central nervous system superficial siderosis: A case report and literature review. *Medicine (Baltimore)*. 2025;104(28):e43312.
7. Hoskin J, Kaneko K, Munakomi S, Fife T. Superficial Siderosis. *StatPearls*. Treasure Island (FL): StatPearls Publishing Copyright © 2026, StatPearls Publishing LLC.; 2026.
8. Mariajoseph FP, Sagar P, Muthusamy S, Amukotuwa S, Seneviratne U. Seizure-induced reversible MRI abnormalities in status epilepticus: A systematic review. *Seizure*. 2021;92:166–73.
9. Kharytaniuk N, Cowley P, Sayal P, Eleftheriou P, Farmer SF, Chan E, et al. Classical infratentorial superficial siderosis of the central nervous system: pathophysiology, clinical features and management. *Pract Neurol*. 2022.
10. Rouault TA. Iron metabolism in the CNS: implications for neurodegenerative diseases. *Nat Rev Neurosci*. 2013;14(8):551–64.

11. Nasri A, Kacem I, Sidhom Y, Djebara MB, Gargouri A, Gouider R. Isolated spinal cord compression syndrome revealing delayed extensive superficial siderosis of the central nervous system secondary to cervical root avulsion. J Spinal Cord Med. 2018;41(4):490–5.

Figures

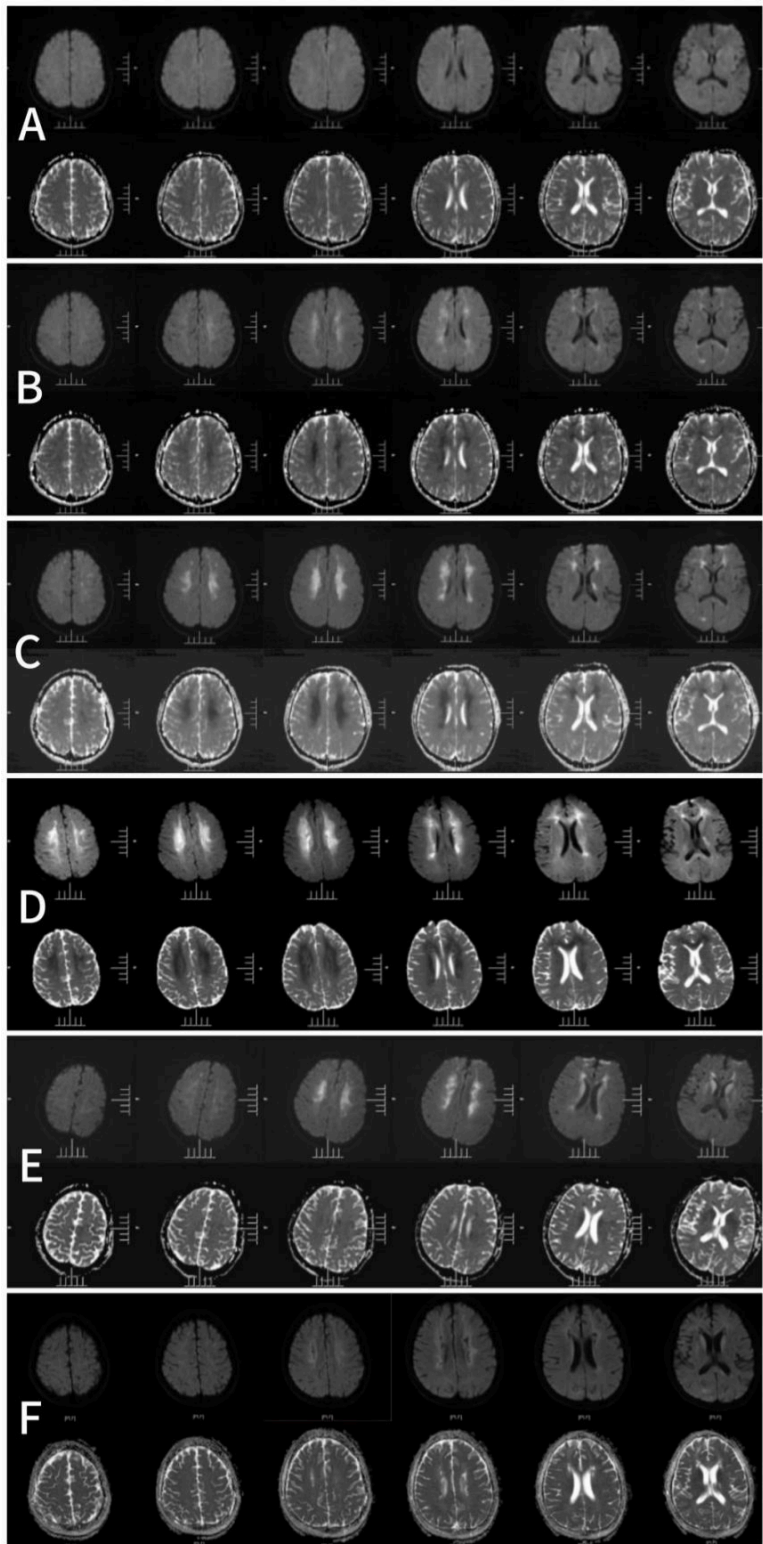


Figure 1

Serial diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) maps during and after epileptic seizures.

(A) 3 d pre-seizure: No restricted diffusion

(B) Seizure onset day: Bilateral symmetrical restricted diffusion

(C) 3 d post-seizure: Expanded restricted diffusion

(D) 10 d post-seizure: Gradual signal resolution

(E) 17 d post-seizure: Further signal resolution

(F) 1 mo post-seizure: Complete resolution of abnormal signals

(A-F) Upper row: DWI; lower row: corresponding ADC maps. Corresponding to the temporal evolution of seizure-associated imaging abnormalities.

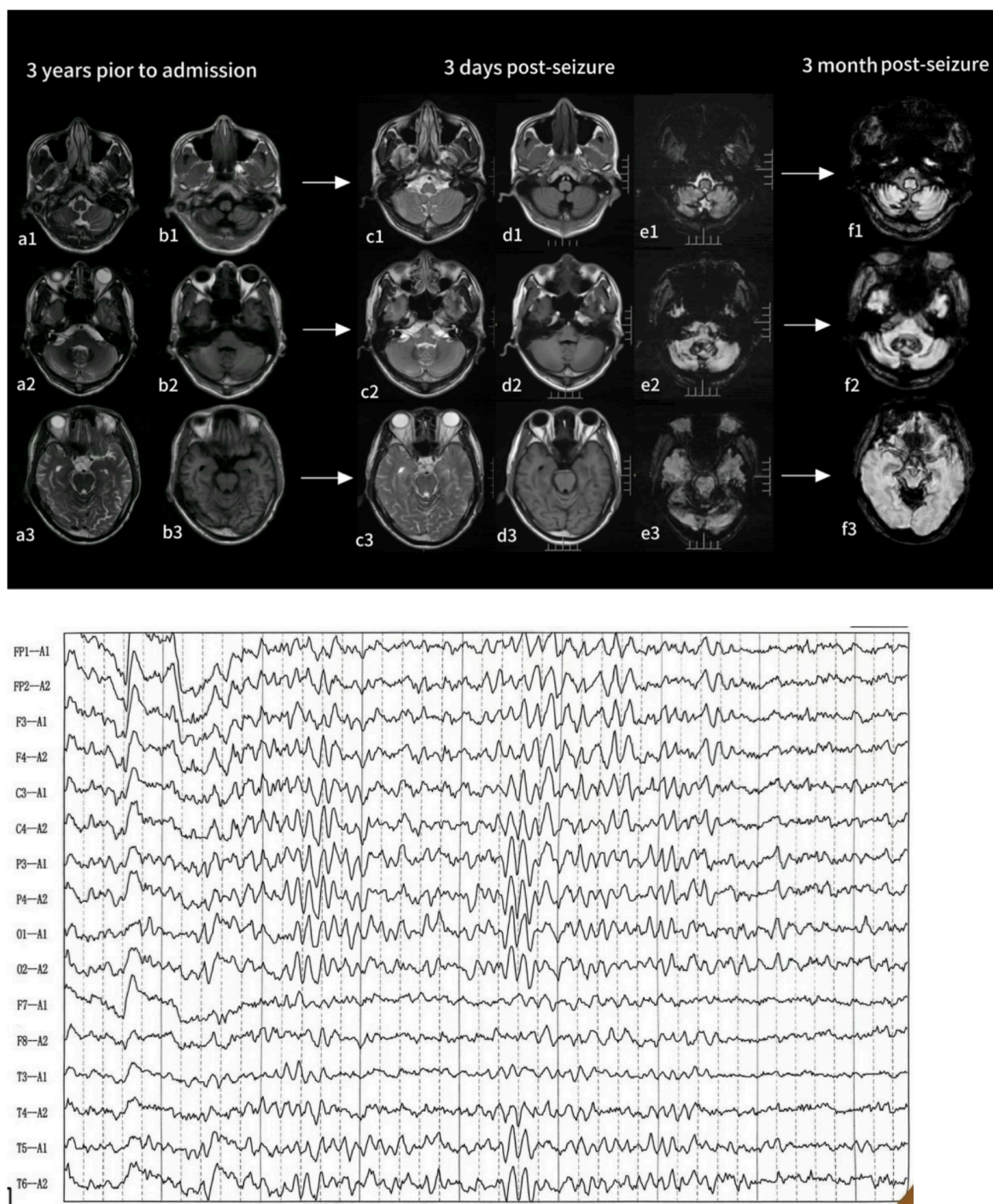


Figure 2

Magnetic resonance imaging (MRI) demonstrating superficial siderosis of the central nervous system (SSCNS). Serial brain MRI in a patient with superficial siderosis of the central nervous system (SSCNS) presenting with epilepsy. a1–a3: T2-weighted imaging 3 years before seizure onset, showing no acute parenchymal lesions. b1–b3: Corresponding T1-weighted imaging demonstrating subtle hypointensities along cerebral surfaces, cisterns, fissures and sulci in keeping with early haemosiderin deposition. c1–

c3: T2-weighted imaging at seizure onset, with no significant interval changes. d1–d3: Corresponding T1-weighted imaging showing persistent superficial hypointensities. e1–e3: Susceptibility-weighted imaging (SWI) at seizure onset revealing marked hypointensities along supratentorial/infratentorial surfaces, cisterns, fissures and sulci, typical of superficial siderosis. f1–f3: Follow-up SWI 1 month after seizure onset showing persistent chronic haemosiderin deposition.

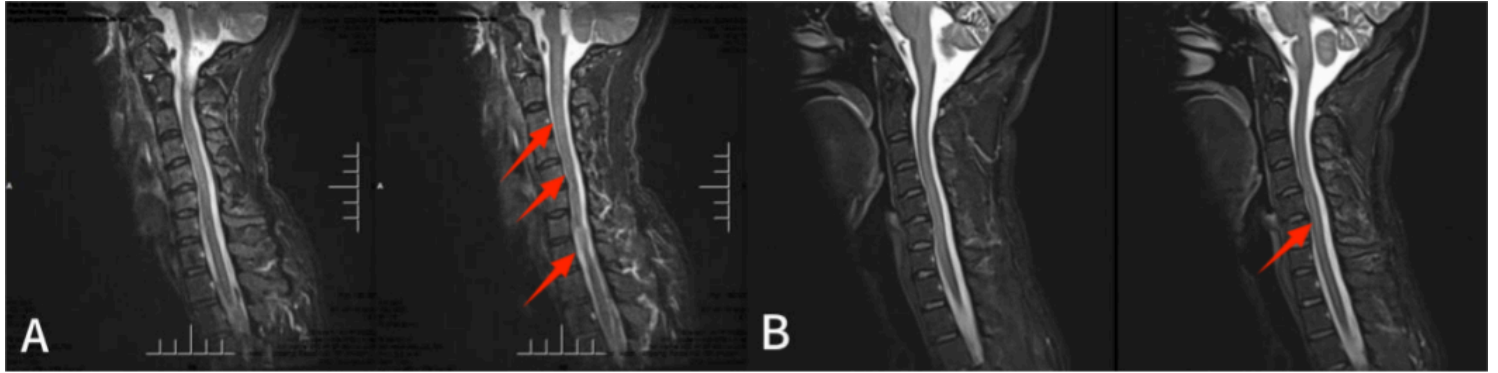


Figure 3

Sagittal T2-weighted cervical spine MRI. T2WI, 3 days post-seizure(A): Hypointensities along the spinal cord surface. 7 months prior(B):Cervical spine MRI, external hospital

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- [CARE.pdf](#)