Case Report

From Spinal Ependymoma to Superficial Siderosis: A Bottom to Top Cause of Progressive Neurological Deterioration

The Neurohospitalist
2024, Vol. 14(4) 454–456

DOI: 10.1177/19418744241273217
journals.sagepub.com/home/nho
Sage

Diana Valente, MD[1], Inês Gil, MD[2], Hipólito Nzwalo, MD, PhD[3], and Catarina Felix, MD[1]

Abstract

Superficial siderosis (SS) is an infrequent condition characterized by hemosiderin deposition in the central nervous system, resulting from chronic subarachnoid hemorrhage, often linked to dural mater diseases. Through a case report of a 50-year-old male with severe sensorineural hearing loss and newly diagnosed epilepsy, we explore SS triggered by a spinal ependymoma, diagnosed via resonance magnetic imaging (MRI). This case highlights the necessity of comprehensive neuroaxis imaging to identify treatable etiologies. The complexity of SS, with its varied clinical presentations, necessitates early detection and a multidisciplinary treatment approach. Despite limited treatment options and the uncertain efficacy of therapies like deferiprone, early intervention is crucial for mitigating irreversible neurological damage and enhancing patient prognosis.

Keywords

sensorineural hearing loss, ependymoma, superficial siderosis

Introduction

Superficial siderosis (SS) is a rare cause of progressive neurological impairment, due to hemosiderin in the subpial layers of the brain and spinal cord.[1,2] Hemosiderin, an iron storage complex, generated by blood breakdown, accumulates due to chronic or repeated low-grade bleeding into the subarachnoid space.[1-4] Red cells leaking into the CSF break down into heme, prompting microglia and Bergmann glia to release haemoxygenase-1 and apoferritin, respectively.[1,2,5] Haemoxygenase-1 breaks down heme into biliverdin and free iron, which is bound by apoferritin to form ferritin and then hemosiderin.[1,2,5] Excess unbound iron becomes neurotoxic once synthesis capacity is exceeded, causing tissue damage.[3,5]

The most common cause of SS is related to diseases of the dura mater.[1,2,4] This condition is best diagnosed using iron-sensitive magnetic resonance imaging (MRI) sequences, which reveal the distinctive subpial deposition of hemosiderin.[1,5] Typical symptoms include bilateral symmetrical sensorineural hearing loss, cerebellar ataxia, and signs of corticospinal tract dysfunction.[1-5]

The case presented here involves SS caused by a spinal ependymoma, highlighting 1 of the possible etiologies of this complex condition.

Case Description

We report the case of a 50-year-old man who was assessed at an epilepsy clinic due to new onset focal seizures. Past medical history was remarkable for meningitis in childhood, followed by slight intellectual disability. The patient, a pneumatic hammer operator, had developed severe sensorineural hearing loss over the years, believed to be occupational in origin. Upon evaluation, hearing loss, executive dysfunction, anterograde memory impairment and a cerebellar syndrome with dysmetric saccades, nystagmus, axial and appendicular ataxia and intentional tremor were demonstrated at examination. At this point, the patient and his family did not report any evolution of symptoms over time. He was treated with eslicarbazepine 800 mg daily with remission of seizures. Routine electroencephalogram was normal. Brain MRI revealed no findings of vascular malformations, aneurysm, amyloid angiopathy, intracranial tumor, or posterior fossa pathologies but a prominent hemosiderin deposition, represented by leptomeningeal hypointense signal intensity in gradient echo sequence (Figure 1A–D), as well as marked cerebellar atrophy (Figure 1C) were present. A diagnosis of SS was established on imaging findings. Upon re-evaluation

[1]Neurology Department, Algarve University Hospital Center, Faro, Portugal
[2]Radiology Department, Algarve University Hospital Center, Faro, Portugal
[3]Algarve Biomedical Research Institute, University of Algarve, Faro, Portugal

Corresponding Author:
Diana Valente, MD, Neurology Department, Algarve University Hospital Center, Rua Leão Penedo, Faro 8000-386, Portugal.
Email: dianavalente@live.com.pt

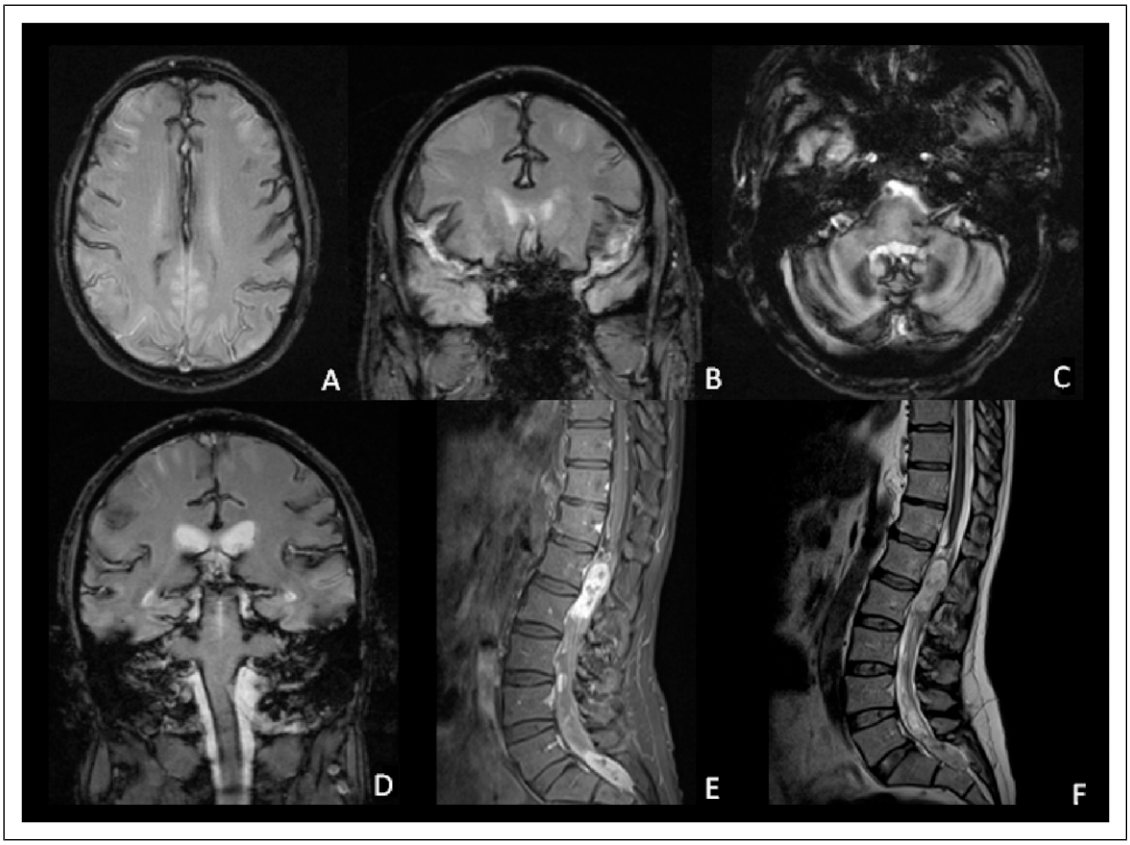

Figure 1. Axial gradient echo images showing superficial siderosis from the level of centrum semiovale (A), temporal lobes (B) and cerebellum (C), which is atrophic. Image C shows deposition of hemosiderin in the internal auditory meatus and auditory-vestibular nerves. The coronal view depicts deposition of hemosiderin in cervical medulla (D). The sagital lumbar and sacrum images (E - T1 weighted with gadolinium; F – T2 weighted image) shows multiple intradural masses of cauda equina suggestive of ependymomas and also a peripheral T2 hypointensity of the lower segments of medulla and conus medullaris, which also reflects superficial siderosis.

and interview, the motor and cognitive symptoms had in fact been worsening over the previous year.

Laboratory evaluation, including platelet count, partial thromboplastin time, prothrombin time, and liver function tests, excluded coagulation disorders as the cause of the chronic hemorrhage. The spinal MRI performed as part of the etiological workout depicted the presence of a cauda equina lesion suggestive of ependymoma (Figure 1E and F).

A neurosurgical evaluation was obtained and surgery was proposed, however, the patient declined it. He had progressive cerebellar signs and hearing loss in follow-up.

Discussion

SS commonly manifests as a slowly progressive cerebellar ataxia with hearing loss, typically appearing in the fourth to sixth decades of life after years of chronic, asymptomatic, low-grade subarachnoid bleeding.[1,3,6] SS manifestations vary widely, mimicking otorhinolaryngological and neurological diseases, including degenerative cerebellar ataxia.[1,3,6] Other symptoms encompass myelopathy-related disturbances, cognitive dysfunction, visual disturbances, seizures and cranial nerve palsies.[1-3,5,7,8]

The most common etiology for SS is spinal dural abnormalities, often dural tears, occurring in the spine or the posterior fossa.[1,2,4] A dural tear can result from disc herniation, intrinsic dural disease due to a connective tissue disease, cranio-spinal traumatic injury, including nerve root avulsions, or neurosurgery.[1,2,5,7-9]

Other reported etiologies that can generate subarachnoid haemorrhage (SAH) and thus siderosis, include tumors, arteriovenous malformations (e.g., cavernous haemangioma), ruptured aneurysms and systemic coagulopathy.[1-5,8-9] In the elderly, cerebral amyloid angiopathy should also be considered.[4]

Tumors are seldom found as a cause of SS, but several types have been implicated, both pre- and postoperatively, most commonly haemorrhagic ones.[1-3,9,10] Reported intracranial tumors include ependymoma, meningioma, oligodendroglioma, pineocytoma, pituitary adenoma, melanocytoma, astrocytoma, paraganglioma, craniopharyngioma and metastases. Reported spinal tumors include

ependymoma, melanocytoma, paraganglioma astrocytoma and teratoma.[8,10]

Despite extensive investigations, the cause of SS is often not apparent.[1] Our case demonstrates that if a full neuroaxis imaging examination is not performed, a spinal lesion as a cause of SS can be missed. Therefore, conducting an MRI of the neuroaxis is essential for determining the source of SS.[3] If the source remains unidentified additional tests like computed tomography (CT) or MRI myelography, CT angiography (CTA), MR angiography (MRA), and digital subtraction angiography may be necessary.[1,5]

Effective management relies on the early identification and surgical correction of the bleeding source.[1-3,5,6] Therapy is advisable even in asymptomatic SS cases with active leaks, due to the risk of progressive neuronal damage.[1] Decision-making for surgery should be personalized, considering the underlying cause, active bleeding, time of evolution, neurological damage and patients' comorbidities.[1,2]

In our case, neurosurgical removal of the spinal ependymoma might have served as an effective treatment to halt the progression of SS, as cessation of symptoms post-excision has been documented.[6,9]

Deferiprone, an iron chelator that crosses the blood-brain barrier, has been used sporadically, showing some clinical and radiological improvement, but its efficacy remains uncertain due to limited evidence.[1,2,5]

The often-delayed diagnosis of this condition can lead to significant morbidity, limiting treatment effectiveness.[2] Due to its complexity and broad range of symptoms, SS requires multidisciplinary clinical input.[2]

In conclusion, we report a case of SS caused by a spinal ependymoma. In SS neurologic damage, which is often irreversible, is related to the time of exposure to the deleterious effect hemosiderin.[1,3] There is no specific available treatment. Hence, recognition of any treatable etiology is essential to halt disease progression and improve prognosis.

Authors' Note

All authors take full responsibility for the data collected, analysis, interpretation, and conduct of this report.

Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

ORCID iDs

Diana Valente https://orcid.org/0000-0003-1049-6719
Hipólito Nzwalo https://orcid.org/0000-0002-1502-3534

References

1. Kumar N. Superficial siderosis: a clinical review. *Ann Neurol*. 2021;89(6):1068-1079. doi:10.1002/ana.26083
2. Kharytaniuk N, Cowley P, Sayal P, et al. Classical infratentorial superficial siderosis of the central nervous system: pathophysiology, clinical features and management. *Practical Neurol* 2022;11. doi:10.1136/practneurol-2021-003324
3. Pikis S, Cohen JE, Vargas AA, Gomori JM, Harnof S, Itshayek E. Superficial siderosis of the central nervous system secondary to spinal ependymoma. *J Clin Neurosci*. 2014;21(11): 2017-2019. doi:10.1016/j.jocn.2014.05.020
4. Weidauer S, Neuhaus E, Hattingen E. Cerebral superficial siderosis: etiology, neuroradiological features and clinical findings. *Clin Neuroradiol*. 2023;33:293-306. doi:10.1007/s00062-022-01231-5
5. Wilson D, Chatterjee F, Farmer SF, et al. Infratentorial superficial siderosis: classification, diagnostic criteria, and rational investigation pathway. *Ann Neurol*, 2017;81(3):333-343. doi:10.1002/ana.24850
6. Spengos K, Vassilopoulou S, Tsivgoulis G, Karachalios G, Vassilopoulos D. Superficial siderosis due to a lumbar ependymoma mimicking adult-onset spinocerebellar ataxia. *Clin Neurol Neurosurg*. 2007;109(8):705-707. doi:10.1016/j.clineuro.2007.04.015
7. Kumar N, Cohen-Gadol AA, Wright RA, Miller GM, Piepgras DG, Ahlskog JE. Superficial siderosis. *Neurology*. 2006;66(8): 1144-1152. doi:10.1212/01.wnl.0000208510.76323.5b
8. Kumar N. Superficial siderosis associations and therapeutic implications. *Arch Neurol*. 2007;64(4):491-496. doi:10.1001/archneur.64.4.491
9. Choi KE, Na SH, Jeong HS, Im JJ, Kim Y. Superficial siderosis of the central nervous system due to spinal ependymoma. *Ann Geriatr Med Res*. 2018;22(1):43-45. doi:10.4235/agmr.2018.22.1.43
10. Tosaka M, Sato K, Amanuma M, et al. Superficial siderosis of the central nervous system caused by hemorrhagic intraventricular craniopharyngioma: case report and literature review. *Neurol Med -Chir*. 2015;55(1):89-94. doi:10.2176/nmc.cr.2012-0362