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Case Reports & Case Series

Superficial siderosis of the central nervous system despite successful treatment of cerebral arteriovenous malformation: A case report

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ABSTRACT

We report a case of superficial siderosis (SS) of the central nervous system that developed approximately 8 years after the treatment of cerebral arteriovenous malformation (AVM). A 17-year-old boy was diagnosed with cerebral AVM in the right frontal lobe, which was treated with embolization and radiation therapy. Radiation-induced encephalopathy occurred 5 years later, and the lesion was resected. Although the patient could walk with a cane following the surgery and rehabilitation, he exhibited complete paresis of the left upper extremity. In the course of the next 9 years, he showed cognitive function decline, paresis of the lower right extremity, and the inability to walk. He also exhibited dysarthria and paresis of the right arm. Respiratory failure developed at age 39 years, necessitating mechanical ventilation. Cerebral angiography revealed no recurrence of cerebral AVM; however, brain and spinal magnetic resonance imaging revealed broad hemosiderin deposition, which is characteristic of SS, and the presence of a postoperative cyst that was suspected to be the bleeding source. While eliminating the source of the bleeding is considered the best line of treatment for SS, this was not pursued due to the patient's irreversible neurological condition. Thus, clinicians should consider SS as a differential diagnosis for patients that exhibit gradual and progressive neurological defects several years after successful AVM treatment.

1. Introduction

Superficial siderosis (SS) of the central nervous system (CNS) is a gradual and progressive neurological disease characterised by cerebellar ataxia, sensorineural hearing loss, and myelopathy. It is caused by chronic or recurrent bleeding that results in hemosiderin deposition on the pial surface of the brain and/or spinal cord. The causes of bleeding include dural pathologies, vascular tumours, and vascular abnormalities [1]. Cerebral arteriovenous malformation (AVM) is considered a direct cause of SS. Elimination of the bleeding source is considered the best line of treatment. Here, we report a rare case involving a patient who developed SS of the CNS approximately 8 years after successful treatment for cerebral AVM.

2. Case report

A 17-year-old boy presented symptoms of headache and nausea, which were found to be associated with AVM in the right frontotemporal lobe. He received five sessions of embolization therapy, with brain haemorrhage occurring as a complication during the second session; this required emergency surgery for removal of the haematoma and resulted in incomplete left-sided hemiparesis. As his AVM was not completely treated, Gamma Knife surgery was performed. At age 22 years, he developed swelling in the right frontal lobe as a result of radiation-induced encephalopathy, and a portion of the affected lobe was surgically removed. Although the patient could walk using a cane following the surgery and rehabilitation, he exhibited complete paresis of the left upper extremity.

At age 30 years, the patient exhibited a decline in cognitive function, followed by paresis of the right lower extremity and inability to walk at age 37 years. The following year, he exhibited dysarthria and paresis of the right arm. Respiratory failure developed at age 39 years, necessitating mechanical ventilation.

Neurological examinations at this point revealed bilateral deafness, gaze nystagmus, spastic quadriparesis, and vesicorectal dysfunction. Cognition and speech could not be evaluated because he had undergone

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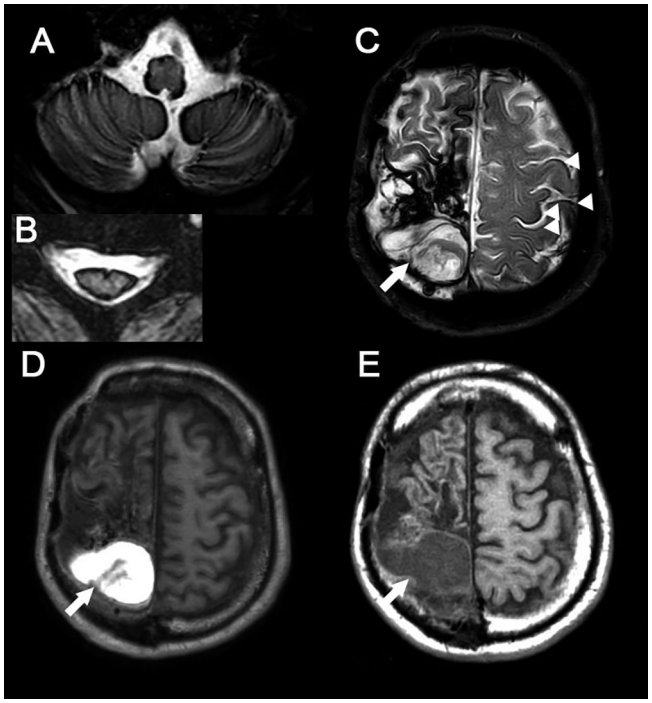

Fig. 1. Magnetic resonance imaging (MRI) findings for a male patient with superficial siderosis of the central nervous system that occurred as a late complication of neurosurgical procedures for cerebral arteriovenous malformation. (A, B, and C) Axial T2-weighted brain MRI shows hypointensity on the surface of the cerebellum, pons (A), and spinal cord (B) because of hemosiderin deposition. There is extensive hemosiderin deposition in the right hemisphere (C). Dense hemosiderin deposits outlining several gyri in the left hemisphere are represented by areas of marked hypointensity (arrowheads). A postoperative cystic lesion is also present in the right posterior lobe (arrow). (D) Axial T1-weighted brain MRI shows hyperintensity in the cystic lesion (arrow). (E) Axial T1-weighted brain MRI performed 8 months prior at another hospital shows isointensity in the cystic lesion (arrow). The signal changes between (D) and (E) suggest recent bleeding.

tracheotomy.

T2-weighted magnetic resonance imaging (MRI) of the brain and spinal cord showed hypointensity around the cerebellum, brain stem, and spinal cord (Fig. 1A and B), which are typical characteristics of SS. In addition, atrophy and encephalomalacia were observed in the right frontal lobe (Fig. 1C, D, and E). A wide hypointense lesion in the right hemisphere suggested hemosiderin deposition (Fig. 1C). The left hemisphere also showed areas of marked hypointensity outlining the gyri (Fig. 1C). A cystic lesion was present in the right posterior lobe (Fig. 1C, D, and E). T1-weighted MRI images of the cystic lesion revealed high signal intensity (Fig. 1D) compared to images acquired 8 months prior at another hospital, which exhibited isointensity (Fig. 1E). These findings suggest that bleeding had occurred in the cystic lesion more recently. Cerebral angiography detected no recurrence of AVM. On the basis of these clinical and imaging findings, the patient was diagnosed with SS of the CNS. Further treatment was not pursued, because his neurological condition was at an irreversible stage.

3. Discussion

SS is a progressive neurological disease resulting from the deposition of hemosiderin after chronic and repetitive bleeding in CNS [1]. AVM is considered a direct cause of SS [2]. However, in our case, cerebral angiography showed no recurrence of cerebral AVM. Moreover, the symptoms of SS manifested 8 years after the final neurosurgical procedure for radiation-induced encephalopathy. A previous report documented two cases of SS occurring as a delayed complication of neurosurgical procedures [3]; one patient exhibited a 37-year delay between the onset of symptoms and the prior neurosurgical interventions and radiotherapy, although the aetiology of SS remained unclear. In our case, a cystic lesion developed after treatment for radiation-induced encephalopathy, which may have caused the SS (Fig. 1C, 1D, and 1E). CSF-containing cysts, found in many SS patients, are considered to be the source of the bleeding [4]. In the present case, the cystic lesion showed signal changes in brain T1-weighted images, indicating recent bleeding (Fig. 1D and E).

4. Conclusion

This case study suggests that SS of the CNS is an uncommon and delayed complication of AVM treatment. Clinicians should consider this disease as a differential diagnosis when patients present with slow and progressive neurological defects several years after the complete elimination of AVM.

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Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.inat.2020.100671.

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