

Superficial Siderosis in Central Nervous System: A Case Report and Literature Review

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Superficial siderosis (SS) in central nervous system is a rare, slowly progressive disease and usually misdiagnosed or diagnosed too late when the patient is chronically devastated. A 55-year-old man with deafness and gait disturbance for ten years was referred from otorhinologist for evaluation of brain. Magnetic resonance image (MRI) showed symmetric hypointense rim partially delineated the bilateral hemisphere on gradient-recalled-echo T2-weighted image, and it was diagnosed as hemosiderin deposition in subarachnoid and subpial meningeal layer. The correct diagnosis of cerebral superficial siderosis can be achieved by careful neurological examination and MRI because computed tomography findings and symptoms are ambiguous. Serial follow-up of imaging study and education for patient are necessary to prevent progression of SS.

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KEY WORDS: Superficial siderosis · Sensorineural hearing loss · Hemosiderin · MRI.

Introduction

Superficial siderosis (SS) in central nervous system (CNS) is an uncommon, slowly progressive disease, which is characterized by hemosiderin deposition from repeated bleeding in subarachnoid space. The common clinical manifestations are ataxia, anosmia, bipyramidal signs and progressive bilateral symmetrical sensorineural hearing loss.^{1,5,6} Hemosiderin is continuously deposited due to recurrent subarachnoid bleeding, and that increases intracellular uptake of iron leading to damage of glial tissues.

The authors present a case of 55-year-old man with progressive bilateral hearing loss, anosmia, and ataxia, and report the cause of the first and eighth cranial nerve injury is developed from hemosiderin deposition and glial damage from recurrent trauma.

Case Report

The patient was a heavy alcoholic, and had a history of traumatic subarachnoid hemorrhage 10 years ago. He has been often slip or fall down in drunken state, and frequently transferred to emergency room due to headache or scalp laceration. The physical examination showed bilateral hearing difficulty, gait ataxia, and complete anosmia. The otorhinologist referred the patient after deciding there is no pathogenic lesion in the hearing or smelling organs. The authors reviewed serial follow-up computed tomography (CT) scans, but it gave no definite and reasonable finding for the neurologic deficits. Magnetic resonance image (MRI) revealed hypointense signal in the bilateral hemisphere on gradient-recalled-echo T2-weighted image, and it was concluded as hemosiderin deposition (Figure 1). These findings were those of SS in CNS.

Discussion

SS of CNS is an unrecognized disease in which hemosiderin is deposited in the leptomeninges, subpial layer, and ependymal surfaces. The clinical manifestations include slowly progressive hearing loss, ataxia, and anosmia.^{1,3} These

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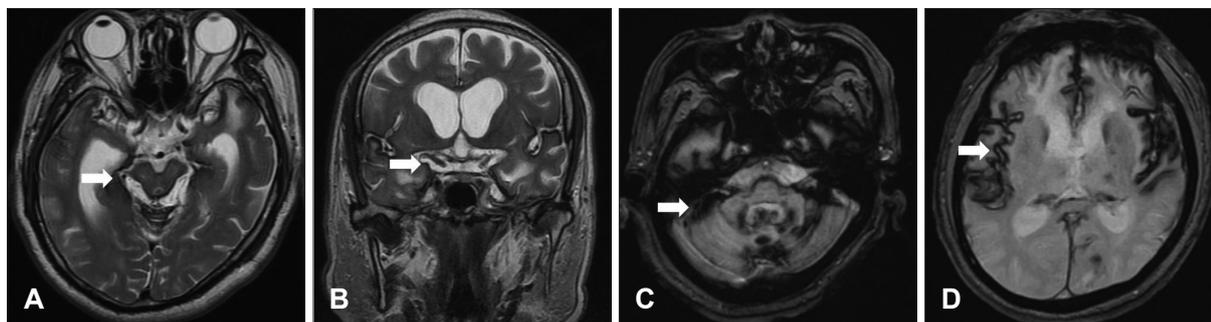


FIGURE 1. A, B: Axial and Coronal T2-weighted MR images show a variably thick hypointense rim outlined the brain stem and various cisterns. C, D: Axial gradient-recalled-echo T2-weighted images show a symmetric well-defined hypointense rim that delineated the cerebellum, some of the lower cranial nerves, bilateral sylvian fissures and other leptomeningeal layers.

features are related to dysfunction in cerebellum and cranial nerves in approximately 90% of the patients.⁵⁾ The hearing loss and anosmia are associated with injury of cranial nerves I, VII, and VIII, whereas ataxia results from atrophy of the cerebellum.

The most common cause of SS is the chronic bleeding into the subarachnoid space of the brain, which releases erythrocyte or blood cells, into the cerebrospinal fluid. The underlying causes have been reported in about 65% of the disease, which are as head trauma (13%), arteriovenous malformations (9%), postsurgical hematomas (7%), and chronic subdural hematomas (6%). The patient had multiple head traumas and history of traumatic subarachnoid hemorrhages. These multiple damages lead to blood breakdown products, heme depositing into the subpial layer of the brain. This toxic product might cause lipid peroxidation, membrane dysfunction and cell death. This process is a neuronal loss, a reactive gliosis and a demyelination which ultimately causes the SS.³⁾

SS is a radiological diagnosis and MRI is the only diagnostic method which shows characteristic features of hemosiderin deposit. The iron deposition that is a characteristic of superficial siderosis shows up as a hypointense rim in the affected areas, but hyperintense rim is known to be rarely seen.⁵⁾ The patient's MRI showed the typical findings such as hypointense rim around the cerebellum and the brain stem, and the cerebellar atrophy. Gradient-recalled-echo T2-weighted sequences are the most sensitive and specific, while spin-echo and fast spin-echo T2-weighted sequences reveals progressively decreased sensitivity to the susceptibility effects of hemosiderin deposit. Cerebrospinal fluid sampling may also reveal siderosis through xanthochromia, elevated presence of red blood cells, high iron and ferritin concentrations, and elevated levels of the proteins Tau, beta amyloid, neurofilament light protein, and glial fibrillary acidic protein. However, the cerebrospinal fluid findings might show no definite abnormal findings in some

cases.⁴⁾

There is no current treatment for SS, but only to help alleviate symptoms and to prevent progress of disease from causative factors. If a source of bleeding can be identified as tumor or vascular malformation, surgical removal of bleeding source can be considered, but has no effect on the pre-existing symptoms.⁷⁾ Some authors reported use of iron chelating drugs and steroids has been explored but there is little evidence to support their effectiveness.^{6,8)} The alleviation of most common symptom, the hearing loss, has been varyingly successful through the use of cochlear implants. Most patients do not notice a big improvement after a successful implantation which is most likely due to the damage of vestibulocochlea nerve, and no cochlea itself.⁹⁾ Some patients heal much faster and return to almost their normal hearing, but their efficacy remains limited.²⁾

Conclusion

The SS in CNS is a rare disease not easily diagnosed by clinical information alone, and it is required for the clinicians understand symptoms and diagnostic method of MRI to prevent progression of the disease.

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REFERENCES

- 1) Fearnley JM, Stevens JM, Rudge P. Superficial siderosis of the central nervous system. *Brain* 118(Pt 4):1051-1066, 1995
- 2) Hathaway B, Hirsch B, Branstetter B. Successful cochlear implantation in a patient with superficial siderosis. *Am J Otolaryngol* 27: 255-258, 2006
- 3) Koeppen AH, Michael SC, Li D, Chen Z, Cusack MJ, Gibson WM, et al. The pathology of superficial siderosis of the central nervous system. *Acta Neuropathol* 116:371-382, 2008
- 4) Kondziella D, Zetterberg H. Hyperphosphorylation of tau protein in superficial CNS siderosis. *J Neurol Sci* 273:130-132, 2008
- 5) Kumar N. Superficial siderosis: associations and therapeutic implications. *Arch Neurol* 64:491-496, 2007
- 6) Kumar N, Cohen-Gadol AA, Wright RA, Miller GM, Piepgras DG, Ahlskog JE. Superficial siderosis. *Neurology* 66:1144-1152, 2006

- 7) Levy M, Turtzo C, Llinas RH. Superficial siderosis: a case report and review of the literature. *Nat Clin Pract Neurol* 3:54-58; quiz 59, 2007
- 8) Scheid R, Frisch S, Schroeter ML. Superficial siderosis of the central nervous system - treatment with steroids? *J Clin Pharm Ther* 34:603-605, 2009
- 9) Sydlowski SA, Cevette MJ, Shallop J, Barrs DM. Cochlear implant patients with superficial siderosis. *J Am Acad Audiol* 20:348-352, 2009