Superficial siderosis of the central nervous system after ventriculoperitoneal shunt

Case report

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The authors report a case of superficial siderosis of the CNS that developed after ventriculoperitoneal (VP) shunt placement for normal-pressure hydrocephalus.

A 65-year-old woman had undergone VP shunt insertion for normal-pressure hydrocephalus. Her gait disturbance, memory disturbance, and urinary incontinence all improved after the procedure. Two years later, however, her gait became ataxic and her appetite became poor. Brain MR imaging revealed a rim of hypointensity on T2-weighted sequences, enveloping the surface of the cortical fissure, cerebellum, and brainstem. Superficial siderosis of the CNS was diagnosed. Steroid administration improved her symptoms.

The authors know of only one case of superficial siderosis developing after VP shunt surgery in the English-language literature. Superficial siderosis should be acknowledged as a possible complication of VP shunt.

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KEY WORDS • superficial siderosis • ventriculoperitoneal shunt • complication

Superficial siderosis of the CNS is a rare disorder resulting from deposition of the iron-containing pigment hemosiderin in the leptomeninges and subpial layers of the CNS. Recurrent or continuous bleeding into the CSF has been implicated. A source of bleeding is found in more than half of all cases. Apart from idiopathic cases, the underlying causes of superficial siderosis have included intracranial brain tumor, spinal tumor, cerebral aneurysm, vascular malformation, and cervical root avulsion. Iatrogenic superficial siderosis has also been reported following hemispherectomy, spinal decompressive surgery, and posterior fossa surgery.

Ventriculoperitoneal shunt surgery is the most common procedure performed in most neurosurgical centers for the treatment of hydrocephalus. Complications are rare but include infection, hemorrhage, obstruction, and migration. Superficial siderosis has rarely been reported as a complication of VP shunt.

We report the case of a patient with superficial siderosis who presented with progressive cerebellar ataxia, hearing loss, and dementia after a placement of a VP shunt for the treatment of NPH. We urge the recognition of superficial siderosis as a possible complication of VP shunt therapy.

Case Report

History. This 65-year-old woman had a history that included, at age 48 years, surgical extirpation of a metastatic brain tumor (left breast cancer) in the right temporal lobe and subsequent whole-brain radiotherapy (total dose 50 Gy). She had been complaining of headache, memory disturbance, gait disturbance, and urinary incontinence. Brain CT scanning revealed a ventricular dilation (Fig. 1A), and a CSF tap test reduced her symptoms. Brain MR imaging demonstrated enlargement of the lateral ventricles, periventricular hyperintensity, and effacement of higher parietal cortical sulci. Her condition was therefore diagnosed as NPH. The patient underwent VP shunt placement after induction of general anesthesia. A Codman-Hakim programmable valve system (Medos SA) was introduced through a right occipital bur hole. At the time of ventricular puncture, and at confirmation of CSF flow from peritoneal tube, no blood was observed in the CSF. The initial pressure of the shunt valve was set at 17 cm H2O.

One day after the operation, brain CT scanning (Fig. 1B) revealed an accumulation of high density fluid in the bilateral posterior horn, suggesting a minor hemorrhagic...
complication. Intracerebral hemorrhage was not observed along the ventricular catheter. Postoperatively, she did not complain of headache or disturbance in consciousness. Her gait disturbance and urinary incontinence gradually improved after the pressure of the shunt valve was set at 12 cm H2O. She was subsequently discharged to home without any complication, and she was able to live independently. She was followed up in our outpatient clinic.

**Presentation and Examination.** Two years later, her gait became ataxic and she complained of appetite loss. She was alert and her orientation was maintained. Her Mini-Mental State Examination score was 17. With regard to her cranial nerves, she experienced sensorineural hearing loss and vestibular disturbance. All of her extremities were hypotonic and ataxic, and truncal ataxia was observed. Brain CT scanning demonstrated no recurrent ventricular dilation, but regions of slightly increased density remained in the left posterior horn (Fig. 1C). On T2-weighted MR imaging we observed a thin hypointense rim on the surface of the cerebral cortex, cerebellum, and brainstem (Fig. 2). On T1-weighted MR imaging, after Gd-DTPA injection, we did not note any enhanced lesion. Xanthochromic CSF was obtained through lumbar puncture, indicating 4000 red blood cells/μl and 24 white blood cells/μl (23 monocytes and 1 polycyte). The CSF protein was 280 mg/dl, CSF glucose was 57 mg/dl, and blood glucose was 98 mg/dl. Cytological analysis confirmed hemosiderin-laden macrophages. Malignant cells were not detected in the CSF.

**Diagnosis, Treatment, and Follow-Up.** Based on these findings, we diagnosed superficial siderosis of the CNS as a complication of the VP shunt procedure. Conversion of the VP shunt to a lumboperitoneal shunt was recommended to the patient's family but was declined. Oral betamethasone (1.5 mg/day) was commenced, and the patient's ataxia and appetite soon improved. She was able to walk without assistance for the following 6 months and was transferred to the rehabilitation hospital. Thereafter, her ataxia gradually deteriorated despite an increasing dosage of betamethasone. The last follow-up was made by the telephone interview with the patient's family at 3 years after the initial steroid treatment. The patient was bedridden and nutrition was given through gastrostomy tube.

**Discussion**

We report a case of superficial siderosis after VP shunt surgery for NPH. The patient in this case had a history of supratentorial craniotomy and radiotherapy for the treatment of a metastatic brain tumor. Examination of the CSF showed xanthochromia without malignant cells. The role of the previous craniotomy and radiotherapy in superficial siderosis in this patient is uncertain. However, CT scanning after VP shunt placement revealed a minor hemorrhage in the ventricle, which had not been present before the procedure. Based on these findings, superficial siderosis appears to have been a complication of VP shunt.

Superficial siderosis is caused by recurrent bleeding into the subarachnoid space, resulting in the deposition of hemosiderin in the leptomeninges and subpial layers of the brain and spinal cord. The syndrome is characterized by 3 clinical manifestations: 1) sensorineural deafness (95%), 2) cerebellar ataxia (88%), and 3) pyramidal signs (76%). Dementia is less common, but cognitive, social, and emotional functions are reported to be impaired. There have been several cases in which superficial siderosis was found at autopsy, although there had been no symptoms during life, and probably there is a presymptomatic phase to the illness, during which superficial siderosis is present but not sufficient enough to cause symptoms. Additionally, there was a clear delay in hemispherectomy cases between the procedure and the onset of symptoms, despite the presence of subarachnoid hemorrhage detected by lumbar puncture after surgery. As a result, diagnosis of superficial siderosis is difficult and may be delayed, unless the treating physician is aware of the possibility.

We know of only 2 cases of superficial siderosis after a CSF diversion procedure in the English-language literature. A 64-year-old man, according to the review by
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Kumar and associates, developed superficial siderosis after shunt placement for a fourth ventricle cyst at age 36 years, but clinical details were not described. McCarron et al. reported on a patient in whom superficial siderosis developed many years after multiple shunt revisions (ventriculoatrial and VP shunt) following posterior fossa surgery for Chiari malformation. They did not mention any relation between superficial siderosis and shunt surgery, however. In the present case, based on the finding during the operation that CSF was not xanthochromic, and that minor intraventricular hemorrhage was seen the day after, superficial siderosis probably developed as a complication of the VP shunt. How this happens is not known. It is possible that repeated impact of a ventricular catheter on the choroid plexus causes minor intraventricular bleeding, because intracerebral hemorrhage was not observed along the tract of ventricular catheter. Superficial siderosis in the present case, however, might be idiopathic because a number of idiopathic cases have been previously reported. According to the extensive review of superficial siderosis of CNS by Fearnley and colleagues, nearly half of cases (24 of 50 cases) were idiopathic.

It is important to be aware that superficial siderosis of CNS may occur as a possible late complication of VP shunt surgery, although it is very rare. Neurological deterioration after VP shunt insertion usually shows as a malfunction of the shunt system, which necessitates a change in the pressure settings or a revision of the shunt system. Use of a programmable shunt system may delay a diagnosis of superficial siderosis due to MR imaging (which is the gold standard for the diagnosis of superficial siderosis), because failure of the programmable valve has rarely been reported after magnetic exposure. It would have been better for us to obtain a CSF sample through the valve or lumbar puncture at the time the present patient exhibited clinical deterioration.

Treatment options for superficial siderosis are not established. Koeppen et al. have suggested that haem synthase antagonists could block the production of hemoglobin, which inhibits apoferritin production and thereby ferritin and hemosiderin, but as yet no such drug crosses the blood-brain barrier. Cochlear implantation has been attempted in selected patients as a treatment for hearing disturbance. Surgical intervention with ablation or removal of the bleeding source probably holds the greatest hope for normalizing, or at least stabilizing, the neurological symptoms. In the present case we recommended the conversion of VP shunt to lumboperitoneal shunt, but this was declined by the patient’s family. Steroid treatment transiently, for at least 6 months, improved the patient’s neurological symptoms, as was shown in a previous report.

Conclusions

Ventriculoperitoneal shunt placement can be a rare cause of superficial siderosis of the CNS. When we encounter neurological deterioration in a patient with a VP shunt, superficial siderosis should be included in the differential diagnosis.

Disclosure

The authors report no conflict of interest concerning materials and methods used in the present report and findings specified in this paper.

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References


Fig. 2. Brain T2-weighted MR images showing low-intensity deposition on the surface of the brainstem, cerebellum, and cerebral cortex. Isointensity deposition (arrow) remains in the right posterior horn, indicating the presence of intraventricular blood.


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