

CASE REPORT

Malpositioned spinal instrument as a possible cause of superficial siderosis

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Study design: Case report.

Objective: To report a patient with superficial siderosis as a complication after posterior fixation surgery for odontoid fracture.

Setting: Department of Neurosurgery, Hokkaido University, Japan.

Methods: A 36-year-old man had undergone C1–C2 posterior fixation using lamina hooks for an odontoid fracture in 1997. In 2003, he presented with hearing loss and ataxia; and in 2006, a diagnosis of superficial siderosis was made and spinal instrument malpositioning was detected.

Results: The malpositioned instrument, suspected as the cause of superficial siderosis, was removed.

Conclusions: Superficial siderosis of the central nervous system is rare; it results in progressive hearing loss, cerebellar ataxia and pyramidal sign. Chronic hemorrhage in the subarachnoid space precipitates hemosiderin around the cerebellum and brainstem resulting in neurological symptoms. Recurrent hemorrhage and cervical root pathology, for example, root avulsion, are factors; the symptoms worsen gradually and result in hemostasis. Superficial siderosis because of complications from spinal instrumentation surgery is extremely rare. If the instrument is malpositioned in the subarachnoid space, we suggest its removal.

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Keywords: superficial siderosis; spinal surgery; complication; spinal instrumentation

Introduction

Superficial siderosis of the central nervous system is characterized by progressive hearing loss, cerebellar ataxia and pyramidal sign.^{1,2} Recurrent subarachnoid hemorrhage deposits hemosiderin on the surface of the nerve tissue and produces symptoms. Cervical nerve root pathology, for example, avulsion injury, is one cause of recurrent hemorrhage.^{1,2} We report superficial siderosis because of malpositioning of a spinal instrument as a complication of spinal surgery.

Case presentation

A 36-year-old man with odontoid fracture had undergone posterior fixation between C1 and C2 with lamina hooks in 1997. In 2003, he developed headache and ataxia; in 2006, he developed hydrocephalus and xanthochromia of the cerebrospinal fluid (CSF) of unknown origin. He was referred

with a diagnosis of superficial siderosis. We detected a lower hook protruding into the spinal canal; but he refused its removal. He again presented in 2008 with left hearing loss, mild monoparesis of the left lower extremity, spasticity of all extremities, positive left Babinski and Chaddock reflex, and loss of pain and temperature sensation below C3. T2-weighted spin-echo magnetic resonance imaging revealed a hypointense rim around the brainstem, cerebellum, vermis, the cortex adjacent to the interhemispheric and Sylvian fissure and an insular region (Figure 1); it was more pronounced on gradient-echo T2* sequences. A hypointense rim on the spinal cord was recognized on spinal T2-weighted magnetic resonance imaging. On computed tomographs (CTs), the lower portion of the left lamina hook extruded into the spinal canal (Figure 2); on CT myelographs, it was seen in the subarachnoid space. CSF was harvested with lumbar puncture three times, 3 days and 3 months after the first investigation. On one CSF study, his erythrocyte count was 1226/ μ l; on two others, the CSF was clear; in all studies the protein level was 73–124 mg per 100 ml.

The instrument was removed with a surgical procedure. The position was prone with a 3-point head pin fixation, and a posterior midline skin incision was made. After the

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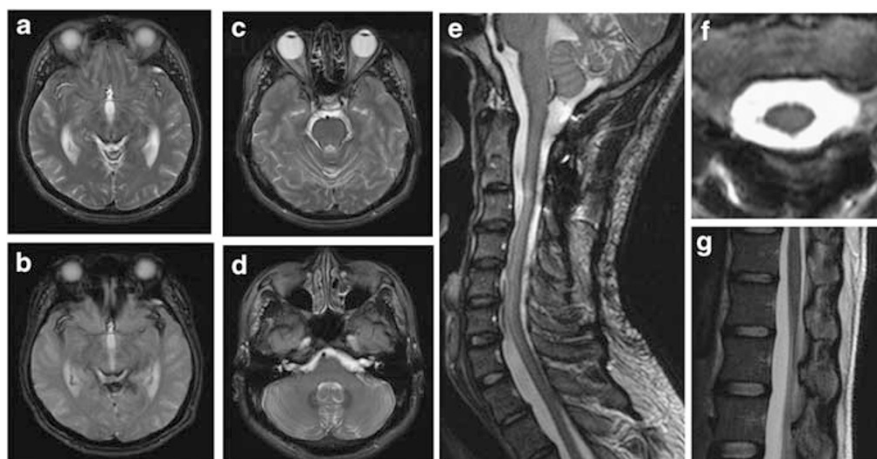


Figure 1 Cerebral T2-weighted magnetic resonance imaging (MRI)- (a, c and d) and T2*-weighted images (b). A marginal hypointense rim is seen at the cortex adjacent to the insular and peri-Sylvian region (a), quadrigeminal plate (a), brain stem (c), upper vermis (c) and eighth nerve (d). Comparison of (a) and (b) at the same height shows that the hypointense rim is more pronounced on image (b). Cervical T2-weighted sagittal, axial and lumbar T2-weighted sagittal images are shown in (e), (f) and (g), respectively. A hypointense rim is also seen at the spinal cord. (Spin-echo T2-weighted images (TR 4540.00 and TE 96.00) are shown in (a), (b) and (d). A gradient echo T2*-weighted image (TR 800.00 and TE 26.00) is shown in (b). Spin-echo T2-weighted images (TR 3500.00 and TE 100.00; TR 5230.00, and TE 98.00; and TR 4000.00 and TE 115.00, respectively) are shown in (e), (f) and (g).

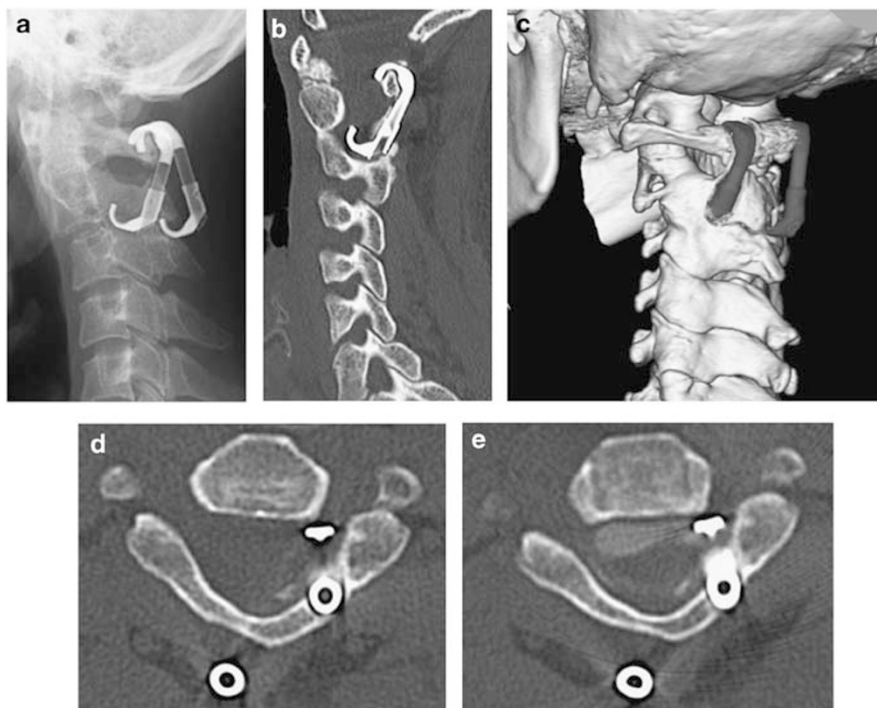


Figure 2 (a): Plain x-ray film. The right lamina hook is in an appropriate position, whereas the left lamina hook is malpositioned. (b): Multiplanar reformatted CT image (sagittal view). The lower hook protrudes into the intervertebral foramen between C2 and C3. (c) Volume-rendering CT image. The lower portion of the hook is embedded in the lamina of C2. (d): Bone window CT image at the C2/3 level (axial view). The tip of the lower hook protrudes into the intervertebral foramen. (e) CT myelogram at the C2/3 level raising the suspicion that the tip of the lower hook is located at the nerve root in the subarachnoid space. CT, computed tomograph.

dissection of the connective tissue from the instrument, the CSF began to leak from the side of the left hook, indicating that it had penetrated the dura mater and extruded into the subarachnoid space. As it was buried in the bone, bone shaving was necessary. Despite laminectomy, the hook could

not be removed; to determine whether it adhered to the nerve tissue and to avoid bleeding, we placed a midline dural incision. The dura adhered tightly to the subarachnoid membrane; the subarachnoid space was pale, indicating chronic bleeding. The hook was adjacent to the origin of the

nerve root. The hook was dissected from adjacent connective tissue; only then could it be removed without difficulty under direct vision. The dural defect was patched with fascia and fibrin glue, and lumbar drainage was placed to prevent CSF leakage.

At 3 months post-removal, his CSF has remained normal without symptom recurrence.

Discussion

Superficial siderosis is a rare condition and is because of chronic subarachnoid hemorrhage. It is characterized by hearing loss, cerebellar ataxia and pyramidal sign.^{1,2} Dementia, hydrocephalus, anosmia, lumbar backache, bilateral sciatica, neck pain and lower motor neuron signs with muscle wasting have been reported.² A source of bleeding has been reported in 54% of the cases either because of dural pathology including a CSF cavity lesion or cervical root lesion, a vascular tumor or a vascular abnormality.² If chronic excess of intrathecal iron overloads the ability of microglial cells for ferritin biosynthesis, ferric ions, toxic and potent generators of oxygen radicals may induce lipid peroxidation.³ The cardinal CSF signs of superficial siderosis are recurrent hemorrhage and xanthochromia; elevated iron, ferritin and protein levels have been reported.² As bleeding may be recurrent rather than continuous, repeated CSF studies are necessary to confirm hemorrhage.²

Magnetic resonance imaging is the most sensitive diagnostic examination. A marginal hypointensity rim, reflecting hemosiderin, is a characteristic of T2-weighted images. In all reported patients, this finding was made around the brainstem and cerebellum;² in many, the rim surrounded the spinal cord and cranial nerves. The cerebellum, basal frontal lobe, cortex adjacent to the interhemispheric fissure, insular and peri-Sylvian regions and the mesial temporal lobe around the hippocampal and choroidal fissures may be involved.^{1,2,4} This hypointensity is more pronounced on gradient-echo T2* sequences. Angiography and technetium-labeled red blood cell scans seldom show the bleeding source.⁴

As there are few treatments to improve existing symptoms, the identification and surgical ablation of underlying causes should be attempted early.^{1,2} The chelating agents, trientine

dihydrochloride and desferrioxamine, the free radical scavengers vitamins C and E or the monoamine-oxidase-B (MAO-B) inhibitor selegiline^{1,2,4} appear ineffective.

Cohen-Gadol *et al.*³ reported superficial siderosis as a complication of spinal surgery. In their patient, a dural tear occurred during anterior decompression and fixation for cervical ossification of the longitudinal ligament, a pseudo-meningocele formed, and superficial siderosis was diagnosed 2 years later. They suggested that the cause was pseudomeningocele. The venous anatomy of the spinal cord and roots may play a crucial role in chronic hemorrhage. As the veins converge and anastomose at the origin of the spinal nerves, they are easily damaged by minor trauma. Consequently, movement of the spinal cord and roots during ordinary neck motion may result in further trauma and bleeding.² In our case, it is supposed that the tip of the hook existed at the origin of nerve root, and easily caused bleeding by usual neck movement. This was thought as pathogenesis of superficial siderosis.

Conclusion

It is not clear whether superficial siderosis as a complication of spinal surgery is an irreversible condition or not. However, superficial siderosis is thought as irreversible condition, so early surgical intervention is recommended.^{1,2} It could be the same in a complication of spinal surgery. Consequently, malpositioned spinal instruments, especially in the intrathecal space, should be removed to prevent the development and progression of superficial siderosis.

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