Superficial siderosis of the CNS is a rare disease characterized by the deposition of hemosiderin in the subpial layers of the CNS as a result of chronic subarachnoid bleeding. The arrest of bleeding is important for preventing the progression of this disease; however, the exact source of bleeding remains unknown in most cases because of a lack of objective surgical data. The authors of this report have described a unique case of superficial siderosis following cervical laminectomy and autograft fusion for the removal of a spinal schwannoma; the bleeding source was verified by intraoperative and histopathological findings. Bone marrow exposure to the intrathecal space may represent a chronic bleeding source in patients with superficial siderosis following CNS surgery including laminectomy or craniotomy. The following recommendations have been proposed for superficial siderosis of the CNS from both a preventative and a therapeutic perspective: 1) During CNS surgery, neurosurgeons should make every effort to prevent exposing bone marrow to the intrathecal space to avoid the risk of chronic subarachnoid bleeding. 2) In the case of a large dural defect and pseudomeningocele following CNS surgery, bone marrow around the dural defect should be considered as the bleeding source of superficial siderosis, and such cases should undergo revision surgery before the progression of this disease.

**Superficial siderosis: bleeding from the bone marrow after laminectomy for spinal tumor removal**

**Case report**

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Superficial siderosis of the CNS is a rare disease characterized by the deposition of hemosiderin in the subpial layers of the CNS as the result of chronic subarachnoid bleeding. On MRI, deposits of hemosiderin appear especially in the cerebellum and other areas including the cerebrum, brainstem, and spinal cord. Its main symptoms are slowly progressive sensorineural hearing loss and cerebellar ataxia. Several underlying conditions have been associated with siderosis: the presence of a CNS tumor or arteriovenous malformation, a history of CNS surgery with or without a CNS tumor, a history of head and neck trauma including nerve root avulsion injury, and intracranial hypotension associated with a spontaneous CSF leak.

Although no therapeutic drugs have been found to ameliorate superficial siderosis once hemosiderin is diffusely deposited and neurological deficits are irreversible, preliminary results have shown that deferiprone, an oral iron chelator, can reduce the deposition of hemosiderin in some patients. It is also known that the arrest of bleeding is important for preventing the progression of this disease; however, the exact source of bleeding remains unknown in most cases. To the best of our knowledge, previous studies have not substantiated the exact bleeding source with objective surgical data. We here describe a unique case of superficial siderosis following cervical laminectomy and autograft fusion for the removal of a spinal schwannoma; the bleeding source was verified by intraoperative and histopathological findings.

**Key Words**

- superficial siderosis
- bleeding source
- bone marrow
- spinal surgery
- neuroradiology
- oncology

**Case Report**

*History and Examination.* Twenty-six years earlier at another institution, a 53-year-old man had undergone cer-
vical laminectomy and posterior spinal fusion with an iliac crest bone graft for the removal of a cervical dumbbell schwannoma. The patient was not affected by any underlying factor associated with siderosis following surgery, including recurrence of the spinal tumor, repeat CNS surgery, new trauma, or intracranial hypotension; however, cerebral CT findings were nondiagnostic when he was later diagnosed with schizophrenia. He was referred to our hospital because screening cerebral MRI showed superficial siderosis of the CNS. No obvious neurological deficits, such as hearing loss and cerebellar ataxia, were observed on admission, although there were the social impairments associated with schizophrenia. He did not have orthostatic headaches, which are a common complication associated with intracranial hypotension. An examination of the CSF revealed that his opening pressure was within normal limits, and his red blood cell count ranged between 768 and 1034/μl.

Brain T2*-weighted MRI (Fig. 1) showed the low deposition of hemosiderin throughout the brainstem, cerebellum, and sylvian and interhemispheric cisterns. Spinal T2-weighted MRI detected a pseudomeningocele surrounded by the autograft bone (Fig. 2A). No obvious tumor recurrence was noted. Brain and spine MRI did not reveal abnormal anatomical findings except for a spinal dorsal pseudomeningocele, which was associated with the prior laminectomy; there were no traumatic lesions or collection of spinal ventral CSF, which is commonly associated with a spontaneous CSF leak.3 Computed tomography myelography showed communication between the subarachnoid space and the pseudomeningocele (Fig. 2D).

Operation. We originally suspected that the bleeding source was the fragile vessels at the dural defect or a residual tumor that had not been detected on preoperative imaging. We performed open surgery to identify the bleeding site and to stop the bleeding. We localized persistent bleeding from the bone marrow of the remaining vertebral arch, which was exposed to the intrathecal space at the rostral margin of the pseudomeningocele (Fig. 2B, C, and E). We achieved hemostasis by removing the bleeding bone en bloc. The dural defect was occluded with autologous fat secured to the dural edges. Histopathological examination revealed that the bone possessed bone marrow with microvessels (Fig. 2F) and that hemosiderin was weakly deposited on the inner wall surface of the pseudomeningocele (Fig. 2G). No other bleeding was observed intraoperatively, and we removed a small residual tumor that had not been detected on preoperative imaging.

Postoperative Course. The collection of subcutaneous CSF occurred following surgery. We reconfirmed complete hemostasis of the bone inside the pseudomeningocele in an additional repair surgery 3 weeks after the first surgery. Red blood cells had almost disappeared (<10/μl) on follow-up CSF examinations 2 months after the second surgery. The patient has remained clinically stable 12 months after the second surgery without apparent hearing loss or cerebellar ataxia. Chronic bleeding was stopped before the onset of irreversible neurological deficits.

Discussion

In the present case, the bleeding source of the superficial siderosis of the CNS was revealed as the bone marrow vasculature of the remaining vertebral arch, and not the fragile vessels at the dural defect or the residual tumor. The patient had no history of additional CNS surgery or any new trauma following spinal surgery; therefore, exposure of the bone marrow was thought to be associated with the laminectomy that had been performed for spinal tumor removal 26 years earlier. To the best of our knowledge, this is the first case in which the bleeding source was verified to be bone marrow exposed to the intrathecal space according to intraoperative and histopathological findings.

Patients with superficial siderosis frequently have a pseudomeningocele associated with a dural defect after CNS surgery or traumatic root avulsion injury.5 Another predisposing condition is a spontaneous spinal ventral dural defect with the collection of CSF, which is associated with spontaneous CSF leaks.7 The possible cause of bleeding is still debated, and two theories have been proposed. One theory is that fragile vessels at a dural defect or trauma sites may be the source of bleeding,8 while the other is that intracranial hypotension caused by spontaneous CSF leaks may result in chronic cerebellar hem-
Superficial siderosis: bleeding from the bone marrow

Repair surgery for a dural defect has recently been described as a treatment for superficial siderosis of the CNS; however, the exact source of bleeding remains unknown in most cases because it is difficult to find evidence of bleeding intraoperatively.

In the present case, bone marrow bleeding was confirmed as the only cause of the superficial siderosis based on the following findings: 1) intraoperative and histopathological confirmation of bone marrow bleeding inside the pseudomeningocele; 2) reconfirmation of hemostasis in the bone in the additional surgery to repair the CSF leak; 3) disappearance of red blood cells in the CSF after two surgeries; and 4) no obvious intracranial hypotension, which can result in cerebellar hemorrhage.

The findings in this case indicated that bleeding from bone marrow exposed to the intrathecal space does not always stop spontaneously, because CSF presumably continues to flow and remove blood clots over the bone marrow. Therefore, a small dural fistula due to incidental durotomy after laminectomy is not typically problematic; however, a large dural defect following osteotomy including laminectomy or craniotomy represents a potential issue in the long term.

We found only one similar case report on superficial siderosis after anterior cervical discectomy and fusion. In that case, the bone marrow of the autograft bone exposed to the intradural space was suggested as the bleeding source. However, the true bleeding source was unclear because surgery was not performed on the basis of a risk-benefit assessment. Chronic subarachnoid bleeding

Fig. 2. A: Sagittal T2-weighted spinal MR image showing a pseudomeningocele surrounded by the autograft bone. B and C: Coronal and axial CT myelograms showing the bone marrow of the remaining vertebral arch at the rostral margin of the pseudomeningocele (arrows). D: Axial CT myelogram showing the dural defect and communication between the subarachnoid space and a pseudomeningocele. E: Intraoperative photograph showing persistent bleeding from the bone marrow inside the pseudomeningocele. F: Photomicrograph showing bleeding bone involving bone marrow with microvessels. H & E, original magnification ×200. G: Photomicrograph showing the deposition of hemosiderin on the inner wall surface of the pseudomeningocele. H & E, original magnification ×200.
may have been arrested by hemostasis in the bleeding bone marrow.

A history of CNS surgery or traumatic root avulsion injury is a well-known predisposing factor in superficial siderosis. A large dural defect and surgical osteotomy or a traumatic bone fracture represents a common feature in each case. In comparing our case with other cases of superficial siderosis in general, we speculate that there may be more cases in previously published reports in which the bleeding source was the intrathecal bone marrow.\(^2,5,6\)

We propose the following recommendations for superficial siderosis of the CNS from both a preventative and a therapeutic perspective: 1) During CNS surgery, neurosurgeons should make every effort to prevent exposing bone marrow to the intrathecal space to avoid the risk of chronic subarachnoid bleeding. 2) In the case of a large dural defect and pseudomeningocele following CNS surgery, bone marrow around the dural defect should be considered as the bleeding source of superficial siderosis, and such cases should undergo revision surgery before the progression of this disease.

Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: Takai. Acquisition of data: Takai, Yokosuka. Analysis and interpretation of data: Takai, Yokosuka. Drafting the article: Takai, Yokosuka. Critically revising the article: Komori. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Takai. Study supervision: Takai, Taniguchi.

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Manuscript submitted April 4, 2013. Accepted August 18, 2014.

Please include this information when citing this paper: published online September 19, 2014; DOI: 10.3171/2014.8.SPINE13328.

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