Intraspinal hemorrhage in spontaneous intracranial hypotension: link to superficial siderosis? Report of 2 cases

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Spontaneous intracranial hypotension due to a spinal CSF leak has become a well-recognized cause of headaches, but such spinal CSF leaks also are found in approximately half of patients with superficial siderosis of the CNS. It has been hypothesized that friable vessels at the site of the spinal CSF leak are the likely source of chronic bleeding in these patients, but such an intraspinal hemorrhage has never been visualized. The authors report on 2 patients with spontaneous intracranial hypotension and intraspinal hemorrhage, offering support for this hypothesis. A 33-year-old man and a 62-year-old woman with spontaneous intracranial hypotension were found to have a hemorrhage within the ventral spinal CSF collection and within the thecal sac, respectively. Treatment consisted of microsurgical repair of a ventral dural tear in the first patient and epidural blood patching in the second patient. The authors suggest that spontaneous intracranial hypotension should be included in the differential diagnosis of spontaneous intraspinal hemorrhage, and that the intraspinal hemorrhage can account for the finding of superficial siderosis when the CSF leak remains untreated.

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Case Reports

Case 1

This 33-year-old man noted the gradual onset of suboccipital orthostatic headaches on April 15, 2014, associated with nausea, emesis, tinnitus, and low-back pain. Results of brain MRI were normal, but a spinal MRI study performed 2 days after the onset of symptoms showed an extensive ventral thoracolumbar extradural CSF collection and a hematoma within the lumbar ventral CSF collection. Symptoms persisted in spite of bed rest, and MRI performed 1 week later showed a persistent CSF leak and evolution of the hematoma. Two weeks later MRI showed resolution of the hematoma with a persistent CSF leak. The patient underwent lumbar and thoracic epidural blood patching with excellent but only temporary relief of symptoms (4–5 days). Digital subtraction myelography showed a CSF leak originating at the T9–10 level, and the patient underwent uneventful surgical repair of the ventral dural tear, using a posterior transdural approach.
lated epidural veins could be visualized through the dural defect. The patient has remained asymptomatic during 12 months of follow-up, and postoperative spine MRI confirmed resolution of the CSF leak (Fig. 1A–H).

Case 2

This 62-year-old woman suffered an acute bifrontal orthostatic orthostatic headache on July 24, 2014, associated with blurred vision, aural fullness, and neck pain. Brain MRI showed subdural fluid collections, brain sagging, meningeal enhancement, and pituitary enlargement. Symptoms persisted in spite of bed rest. Spinal MRI performed 58 days after the onset of symptoms showed intrathecal hemorrhage and an extensive circumferential spinal extradural CSF collection, confirmed by CT myelography. Following placement of a single lumbar epidural blood patch, symptoms resolved completely and follow-up brain and spine MRI obtained 1 month later showed normal results (Fig. 1I–M). The patient has remained asymptomatic during 8 months of follow-up.

Discussion

Spontaneous intraspinal hemorrhage can be due to vascular malformations, tumors, or coagulopathy, and also can be observed following intracranial hemorrhage. Often no cause of the hemorrhage can be identified. None of these factors were present in the patients reported here. We suggest that spontaneous spinal CSF leaks and intracranial hypotension should be included in the differential diagnosis of spontaneous intraspinal hemorrhages.

Superficial siderosis is characterized by hemosiderin deposits in the subpial layers of the brain and spinal cord and is due to chronic bleeding within the subarachnoid space. A ventral spinal CSF leak can be demonstrated in approximately half of patients with superficial siderosis. A history of orthostatic headaches with brain MRI findings of spontaneous intracranial hypotension is present in many of these patients, whereas in others no history of orthostatic headaches can be elicited or the brain MRI does not show findings of spontaneous intracranial hypotension.
In our study of cerebellar hemosiderin deposits in spontaneous intracranial hypotension, all patients had a ventral spinal CSF leak, almost always with symptoms of very long duration, measured in years or decades.

A unique feature of the 2 presently described patients with spontaneous intracranial hypotension is the early spinal imaging that showed intraspinal hemorrhage. In the patient with a ventral CSF leak (Case 1), symptoms persisted in spite of multiple epidural blood patches. The hematoma, which was confined to the ventral extradural CSF collection, persisted for at least 1 week on imaging. This patient eventually underwent surgery and a ventral dural defect with underlying dilated epidural veins was confirmed intraoperatively. In the patient with a circumferential spinal CSF leak (Case 2), however, the leak and the intrathecal hemorrhage resolved after a single epidural blood patch. Symptoms of spontaneous intracranial hypotension due to a ventral CSF leak often persist, not only with conservative treatment but also with epidural blood patching.

Conclusions

These 2 cases support the hypothesis that superficial siderosis is related to hemorrhage from the spinal CSF leak site. It is of interest to note that none of the reported patients with superficial siderosis and a ventral CSF leak had any treatment for their CSF leak until superficial siderosis was diagnosed.

Whether it is the chronicity of the CSF leak, the unique anatomical location of the dural defect, or a combination of both that predisposes patients with a ventral spinal CSF leak to superficial siderosis remains to be established.

References


Disclosures

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

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