Superficial siderosis

Superficial siderosis of the central nervous system is a rare intractable disease induced by chronic subarachnoid hemorrhage. Neurological deficits, such as cerebellar ataxia and hearing difficulties, gradually progress if left undiagnosed. Hemosiderin deposition is irreversible because standard medical treatment has not yet been established. Interventions at the source of bleeding may be the key to a preferable outcome of treatment for chronic subarachnoid hemorrhage; however, the source is not clear in many cases.

Case series

Among the consecutive cases diagnosed with a spontaneous cerebrospinal fluid (CSF) leak, cases of superficial siderosis associated with a Cerebrospinal fluid fistula due to a ventral dural defect were retrospectively analyzed.

Results: Among 77 cases of a Cerebrospinal fluid fistula, 7 cases (9%) of superficial siderosis were identified (median age of 59 years, male, 4 cases). Defects were diagnosed on 1-mm sliced fast imaging employing steady-state acquisition MRI (n = 5), conventional myelographic CT (n = 1), or dynamic myelographic CT (n = 1) at high thoracic levels (T1-T4). All defects were repaired by direct neurosurgery. During surgery, continuous bleeding from the epidural veins of the vertebral epidural venous plexus was identified as the source of subarachnoid hemorrhage. Epidural CSF pulsations through the defect prevented clot formation by the epidural veins. Dural repair stopped free communication between the subarachnoid and epidural spaces, leading to the disappearance of chronic subarachnoid hemorrhage.

Bleeding from the epidural venous plexus may be the cause of superficial siderosis associated with ventral dural defects. Neurosurgical repair may stop the progression of this condition [1)].

Case reports

A patient with superficial siderosis whose symptoms deteriorated after lumbar subarachnoid-peritoneal (L-P) shunt surgery. She received L-P shunt surgery based on the diagnosis of idiopathic normal pressure hydrocephalus at another hospital. The patient had a history of cervical surgery, and a dural defect was identified at the C4-5 level by a detailed magnetic resonance imaging study. We hypothesized that the L-P shunt reduced cerebrospinal pressure and increased bleeding from the fragile vessels in the dural defect, which might have increased hemosiderin deposition [2)]

A 61-year-old male with a 12-year history of limb weakness, muscle wasting, cramps, clumsiness, progressive unsteady gait, and fine motor impairments. His medical history included the resection of a left parietal meningioma and a myxopapillary ependymoma near the conus terminalis (L3/4) at the age of 51 years. The clinical examination revealed a motor neuron syndrome with a clear bilateral wasting of the hand muscles, a diffuse atrophy of the shoulder and calf muscles, and a weakness of the arms, fingers, hips, and feet. Deep tendon reflexes were symmetrically briskly hyperactive. Standing and walking were only possible with a support. Magnetic resonance imaging of the entire

neuroaxis showed progressive severe cerebral, brainstem, and spinal superficial siderosis in form of extensive hypointensities on T2-weighted gradient-echo images and susceptibility-weighted sequences. Despite a successful neurosurgical removal of the tumors and delaed medical treatment with an iron chelator for one year, they observed no clinical recovery or stability in the patient, making this case unique, and suggesting an irreversible neurodegenerative process. This case reinforces the need of including SS-CNS in the list of amyotrophic lateral sclerosis (ALS)-mimics and demonstrates the fundamental use of a complete neuraxial MRI investigation on evaluating possible ALS cases [3)]

1)
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2)
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3)
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