Superficial Siderosis Associated with a Spinal Dural Defect

Toshitada Hiraka1*, Masafumi Kanoto1, Yuki Toyoguchi1, Ryousuke Igari2, Takeo Kato2, and Takaaki Hosoya1

Keywords: superficial siderosis, dural defect, spine, normal cerebral spinal fluid pressure

Superficial siderosis of the central nervous system is a rare chronic progressive disease caused by continuous repeated hemorrhage in the subarachnoid space. The ability of the brain to biosynthesize ferritin in response to prolonged contact with hemoglobin iron is important in the pathogenesis of superficial siderosis. This frequently results in cerebellar ataxia and sensorineural deafness. Causes of superficial siderosis include head or back injury, brachial plexus or nerve root injury, dural defect, tumors, vascular disease such as anteriovenous malformation/fistula, and prior intradural surgery.1 Some reports have described superficial siderosis due to spinal dural defect with fluid collection in the spinal canal, and also the surgical closure of dural defects for treatment of superficial siderosis associated with abnormal communication between the subarachnoid space and epidural fluid collection. Thus, it is important to detect dural defects by imaging examination.

A 58-year-old man was admitted to our hospital for further examination of an unsteady gait. Screening brain magnetic resonance imaging (MRI) had revealed superficial siderosis (Fig. 1A). Audiometry revealed bilateral sensorineural hearing impairment that the patient was not aware of in daily life. Lumbar puncture indicated an opening pressure of 130 mm H2O. Cerebral spinal fluid (CSF) was clear and colorless, and CSF examination revealed elevated protein (72 mg/dl) but no other abnormalities.

The patient underwent brain and spinal MR imaging using a 3 Tesla unit (MRI Signa HDxt 3.0T; General Electric medical Systems, USA), which showed no tumor or vascular disease. Axial T2-weighted spinal imaging showed epidural fluid collection from C3 to Th10 (Fig. 1B), and axial Cube T2 of the spine revealed a dural defect at the ventral dural sac at the Th1 level (Fig. 1C). Sagittal T1-weighted images with fat suppression after gadolinium administration (T1WI-Gd) showed dilation of the anterior spinal vein (Fig. 1D). Computed Tomography (CT) myelography was performed and confirmed a CSF leak around the dural defect (Fig. 1E, F). The patient was diagnosed with superficial siderosis due to spinal dural defect. He decided against surgery and was kept under observation.

In cases of superficial siderosis with dural defect, it is often difficult to detect the location of the dural defect. High-resolution constructive interference in steady-state (CISS) MR images were reported to be useful in detecting the dural defect preoperatively.2 We also successfully identified dural defect sites in a superficial siderosis case by Cube T2 and CT myelography. Cube is a relatively new three dimensional (3D) fast spin-echo pulse sequence with parallel imaging and extended echo train acquisition that can be reformatted into any plane to visualize even small and low-contrast lesions without a partial-volume effect. Imaging of the cervical spine using the 3D T2-weighted technique reportedly conferred superior delineation of anatomical structure compared to two dimensional (2D) T2-weighted sequences.3 Although CT myelography is needed to confirm the site of dural defect in preoperative or follow-up cases, Cube T2 is very useful in detecting the suspected site of dural defect compared with other 2D sequences prior to CT myelography.

Conflicts of Interest

The authors declare that they have no conflict of interest in this manuscript.
Fig 1. (A) Axial gradient echo image (repetition time [TR] / effective echo time [TE], 660.00 ms / 20.00 ms) shows superficial siderosis. (B) Sagittal T2-weighted spinal image (TR / TE, 3320.00 ms / 124.70 ms) shows epidural fluid collection from C3 to Th10. (C) Axial three dimensional (3D) T2-weighted images obtained with Cube (Cube T2, 1.4 mm, TR / TE, 1820 ms / 92.87 ms) of the spine demonstrates a dural defect (arrow) at the ventral dural sac at the Th1 level. (D) Sagittal T1-weighted images with fat suppression after gadolinium administration (T1WI-Gd), TR / TE, 540.00 ms / 7.96 ms) show dilation of the anterior spinal vein. (E–F) Computed Tomography (CT) myelography confirms cerebral spinal fluid (CSF) leak around the dural defect.

References