Case Report

Diagnosis of spinal dural defect using three-dimensional fast steady-state MR in patient with superficial siderosis: A case report

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ABSTRACT

Background: Spinal dural defects can result in superficial siderosis (SS) of the central nervous system. Closure of the defect can stop or slow the progression of the disease. Here, we evaluated, whether preoperative three-dimensional fast steady-state acquisition MR could adequately detect these defects and, thus, facilitate their closure and resolution.

Case Description: A 65-year-old right-handed male presented with a 33-year history of the left C8 root avulsion and a 3-year history of slowly progressive gait difficulties and hearing loss. T2*-weighted imaging revealed symmetrical hemosiderin deposition throughout his central nervous system. A left C6-C7 dural defect involving only inner layer was identified using a three-dimensional MR (3D-FIESTA). It was treated through a left C6-7 hemilaminectomy and successfully sealed with adipose tissue and fibrin glue. Subsequently, the progression of cerebellar ataxia was halted, nevertheless the sensorineural hearing loss worsened even over the next 2 years.

Conclusion: 3D-FIESTA reconstruction was approved to be useful tool for identifying the tiny hole of the inner dural layer responsible for SS.

Keywords: Detection, Dural defect, Magnetic resonance imaging, Superficial siderosis

INTRODUCTION

Superficial siderosis (SS) of the central nervous system is a progressive disease caused by hemosiderin deposition due to repeated hemorrhage into the subarachnoid space. Symptoms of classical SS include the slow progression of cerebellar ataxia, hearing loss, myelopathy, and dementia. It is typically due to a spinal dural defect and subsequent fluid collection in the epiarachnoid space. More recently, duplicated dura matter and a collection of fluid between the two layers have been reported in some patients with SS. Repair of the dural defect often stabilizes or resolves symptoms. Constructive interference in steady-state (CISS)-magnetic resonance imaging (MRI) or fast imaging employing steady-state acquisition (FIESTA)-MRI has...
been reported to be useful for detecting the defect.[6] Here, we report a 65-year-old male with SS, whose spinal dural defect was identified by three-dimensional FIESTA-MRI and successfully closed by a minimally invasive surgery.

CASE DESCRIPTION

A 65-year-old man presented with a history of the left C8 root avulsion 33 years before, 3-year history of slowly progressive gait difficulties and hearing loss, and a 2-week history of a headache. A neurological examination revealed sensorineural deafness, mild truncal ataxia, and findings consistent with the long-standing left C8 root avulsion. The brain CT on arrival demonstrated a left chronic subdural hematoma, and following evacuation, the headache was promptly resolved [Figure 1a]. In addition, the patient underwent neck clipping of a right middle cerebral artery aneurysm, because a latent subarachnoid hemorrhage from the aneurysm was suspected on brain MRI [Figures 1b and c]. The CSF was xanthochromic and engorged pial vessels were prominent throughout the surface of the cerebrum [Figure 1d]. The whole brain surface was yellowish, and we confirmed microscopically the hemosiderin-laden macrophages on the surface of the arachnoid [Figure 1e]. However, no evidence of bleeding was observed around the aneurysm. Thereafter, a cervical spinal inner dural defect and fluid collection between the two dural layers around it was detected by CT myelography and spinal MRI, which facilitated accurate surgical closure of the defect.

MRI results and laboratory studies

T2*WI after the evacuation surgery revealed hemosiderin deposition throughout both cerebral hemispheres and cerebellar hemispheres as well as spinal cord [Figure 1b],

![Figure 1](image-url)

Figure 1: Key features and imaging findings before the dural defect closure operation. (a) Brain CT: chronic thin left subdural hematoma. (b) Brain T2*-MRI: thick symmetrical deposition of hemosiderin on surface of cerebellum/brainstem. (c) Contrast Brain MRI: marked dural thickening/enhancement, consistent with CSF hypovolemia. (d) Intraoperative view during aneurysmal clipping: diffuse hemosiderin deposition, and engorged pial vessels throughout the surface of the cerebrum (arrowheads). (e) Hematoxylin-eosin stain of arachnoid: hemosiderin-laden macrophages (arrowheads) is prominent on the surface (original magnification ×200, black bar = 100 µm). (f) Sagittal reverse FIESTA-MRI: longitudinal ventral/dorsal fluid collection interdurally from C6 to Tb4. (g and h) Axial reverse FIESTA-MRI: fluid collection (asterisk) adjacent to the dural sac, and dural defect at the left C6/7 level (arrowhead). Cross-sections as indicated by the lines in Figure 1f.
and the dura matter was thickened and remarkably enhanced by gadolinium consistent with intracranial hypovolemia [Figure 1c]. Reversed FIESTA-MRI (TR = 4.6 ms, TE = 1.9 ms, ST = 0.5 mm; Discovery MR750w 3.0Tesla, General Electric; Fairfield, CT) revealed an accumulation of CSF between two dural layers of C6-Th4, involving both the dorsal and ventral dural layers [Figures 1f-h]. A 3D reconstruction of FIESTA-MRI scan was used to visualize the range of duplicated dura matter and the characteristic shape of the outer layer [Figures 2a-c].

A lumbar puncture before the craniotomy revealed opening pressure of 7 cmH₂O, xanthochromia (4200 red blood cells/µL), and an elevated protein level (146 mg/dL) [Table 1].

Surgery and postoperative course

After completing a left C6-7 hemilaminectomy, no extradural fluid collection was identified, and a nearly identical dural structure to the preoperative 3D-FIESTA-MRI was confirmed [Figure 2d]. Once a 5-mm incision was made in the outer dural layer, CSF within the interdural space flowed out, and a fistula of inner dural layer was found exactly at the site of exposure which was closed completely with an adipose tissue and fibrin glue [Figure 2e].

Postoperatively, the reverse FIESTA-MRI revealed slight decrease of CSF in the interdural space [Figures 3a-c]. One year later, xanthochromia and high protein concentration in the CSF were resolved, and the cerebrospinal pressure was normalized [Table 1]. Over the next 2 postoperative years, cerebellar ataxia and headache have resolved. However, the deposition of hemosiderin on T2*WI did not change and sensorineural hearing loss has worsened even thereafter [Table1].

DISCUSSION

Etiology of SS

Here, we reported a case of SS related to duplicated dura matter caused by a defect of the inner dural layer, which is detected utilizing 3D-FIESTA-MRI and minimized the operative invasiveness. Kumar et al. first reported cases of spinal dural defect treated by surgical repair.[8] Cheng et al. considered that an increased epidural pressure may damage the fragile venous plexus, leading to recurrent microbleeding.[2] Recently, Hosokawa et al. suggested that the
defect of inner dural layer cause dissection between two dural layers, and subsequent continuous leak of blood according to Monro–Kellie hypothesis.\textsuperscript{[6,10]}

**Repair of spinal dural defects to treat SS**

Various imaging modalities have been utilized to identify the dural defects responsible for SS. Kumar et al. reported the usefulness of dynamic CT myelography,\textsuperscript{[9]} and Arishima et al. used coronary angioscope as a spinal endoscope along with CT myelography.\textsuperscript{[1]} Egawa et al. suggested using CISS-MRI for detecting the defect,\textsuperscript{[9]} and Hosokawa et al. proposed the use of CISS reverse MRI.\textsuperscript{[6]} Hingwala et al. emphasized the usefulness of a 3D reconstruction image for visualizing the structure adjacent to CSF.\textsuperscript{[3]} Representative literatures focusing on imaging modalities to find spinal dural defects are listed in Table 2. In this case, 3D-FIESTA-MRI showed that the fluid collection was isolated and surrounded by the duplicated dural layers, which was suggested by Hosokawa et al.\textsuperscript{[6]}. Although we could not see a spinal dural defect from the outside, we could confidently identify the location of the defect with only a 5-mm incision of the outer layer thanks to the nearly identical 3D dural image. Therefore, 3D-FIESTA-MRI might be useful for minimizing the surgical invasiveness, especially for cases with fluid collection in the interdural space.

![Figure 3: (a) Postoperative sagittal reverse FIESTA-MRI: interdural fluid collection was slightly decreased. (b and c) Axial reverse FIESTA-MRI showing a slight decrease of CSF in the interdural space. Both pictures are cross-sections corresponding to lines in Figure 3a.](image)

| Table 1: Pre- and post-operative findings. |
|------------------------------------------|
| MRI findings                             |
| Interdural fluid collection              |
| Hemosiderin deposition                   |
| CSF findings                             |
| Appearance                               |
| Protein (mg/dL)                          |
| Total bilirubin (mg/dL)                  |
| RBCs (number of cells/mm\(^3\))          |
| Initial pressure (cmH\(_2\)O)            |
| Symptoms                                 |
| Headache                                 |
| Sensorineural hearing loss               |
| Cerebellar ataxia                        |

| Presurgery | 12 months Postsurgery | 3 years Postsurgery |
|------------|-----------------------|---------------------|
| MRI findings                             |
| Interdural fluid collection              |
| Hemosiderin deposition                   |
| CSF findings                             |
| Appearance                               |
| Protein (mg/dL)                          |
| Total bilirubin (mg/dL)                  |
| RBCs (number of cells/mm\(^3\))          |
| Initial pressure (cmH\(_2\)O)            |
| Symptoms                                 |
| Headache                                 |
| Sensorineural hearing loss               |
| Cerebellar ataxia                        |

| Table 2: Review of the representative literature of SS treated by dural closure. |
|------------------------------------------|
| Imaging modality | Cause of SS | Surgical treatment | Neurological outcome |
|------------------|-------------|--------------------|----------------------|
| Kumar et al. (2005)\textsuperscript{[9]} (3 cases) | CT (dynamic myelography) | Spinal dural defect (3/3 cases) | Defect closure (muscle piece) | Stabilized (1/3) |
| Egawa et al. (2013)\textsuperscript{[9]} (2 cases) | CISS MRI | Spinal dural defect (2/2 cases) | Defect closure (suture) | Not documented (2/3) |
| Arishima et al. (2017)\textsuperscript{[1]} (2 cases) | Spinal endoscopy | Spinal dural defect (2/2 cases) | Defect closure (suture) | Headache improved (1/2), stabilized (1/2) |
| Hosokawa et al. (2018)\textsuperscript{[8]} (3 cases) | CISS reverse MRI | Spinal inner dural defect (3/3 cases) | Defect closure (fat/fibrin glue) | Ataxia and hearing loss worsened (1/2), stabilized (1/2) |
| Present case (2022) | 3D-FIESTA-MRI | Spinal inner dural defect | Defect closure (fat/fibrin glue) | Stabilized (3/3) |

![Figure 3: (a) Postoperative sagittal reverse FIESTA-MRI: interdural fluid collection was slightly decreased. (b and c) Axial reverse FIESTA-MRI showing a slight decrease of CSF in the interdural space. Both pictures are cross-sections corresponding to lines in Figure 3a.](image)
CONCLUSION

The 3D-FIESTA-MRI was an excellent tool for visualizing the dural defect responsible for SS of the central nervous system and was critical for preoperative planning to occlude the fistula with a minimal invasiveness.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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