





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. The 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.^[4] Symptoms of classical SS include the slow progression of cerebellar ataxia, hearing loss, myelopathy, and dementia.^[4] It is typically due to a spinal dural defect and subsequent fluid collection in the epidural space.^[8] More recently, duplicated dura matter and a collection of fluid between the two layers have been reported in some patients with SS.^[6] Repair of the dural defect often stabilizes or resolves symptoms.^[2,7] Constructive interference in steady-state (CISS)-magnetic resonance imaging (MRI) or fast imaging employing steady-state acquisition (FIESTA)-MRI has

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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],

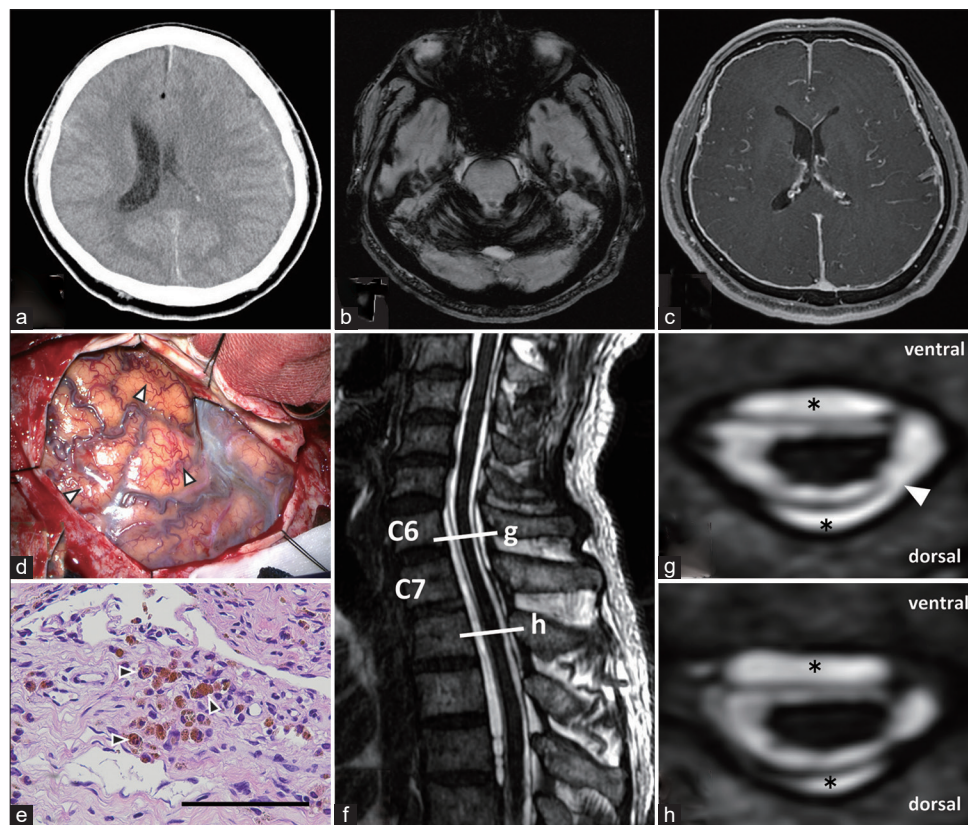


Figure 1: Key features and imaging findings before the dural defect closure operation. (a) Brain CT: chronic thin left subdural hematoma. (b) Brain T2*-MRI: thick symmetrical deposition of hemosiderin on surface of cerebellum/brainstem. (c) Contrast Brain MRI: marked dural thickening/enhancement, consistent 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 $\times 200$, black bar = $100\ \mu\text{m}$). (f) Sagittal reverse FIESTA-MRI: longitudinal ventral/dorsal fluid collection interdurally from C6 to Th4. (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/ μ L), 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 [Table 1].

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

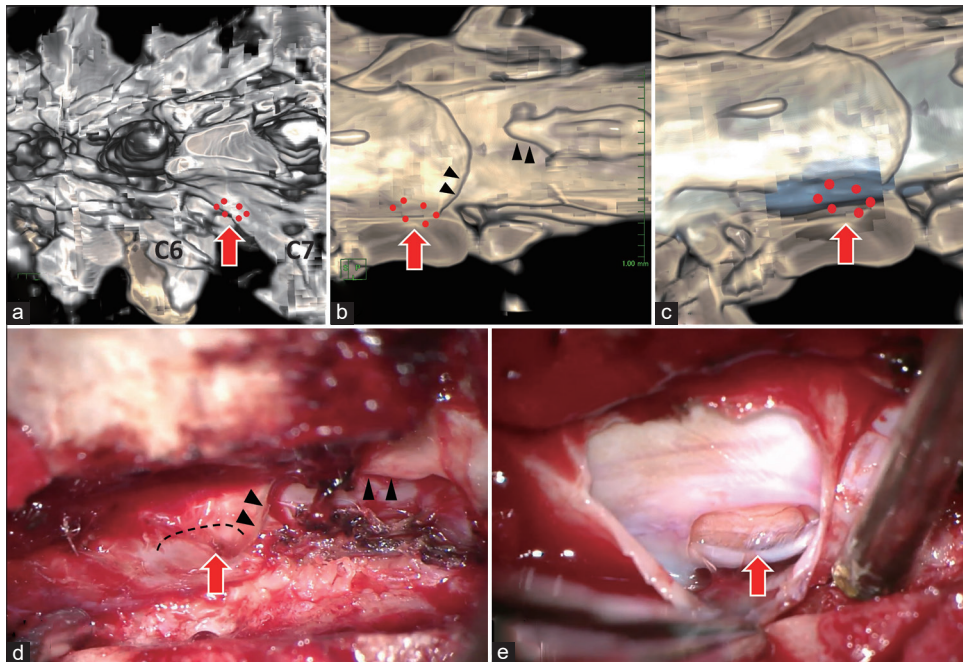


Figure 2: (a) A 3D-reconstruction of the FIESTA-MRI: preoperatively estimated location of the dural defect left side at C6/7. (b) A dorsal view of 3D-FIESTA-MRI showing the characteristic surface structure. (c) After opening the outer layer of the dura, a blue-colored spinal cord is shown through the opening. (d) Intraoperative photograph showing the identical duplicated dural folds around C6/7 (broken line: incision in outer dura). (e) Another intraoperative photograph showing the inner dural defect. All red arrows/red dots indicate the same dural defect, with black arrowheads in (b) and (d) indicate the same characteristic folds of the outer dural layer.

defect of inner dural layer cause dissection between two dural layers, and subsequent continuous leak of blood according to Monro–Kellie hypothesis.^[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,^[9] and Arishima *et al.* used coronary angioscope as a spinal endoscope along with CT myelography.^[11] Egawa *et al.* suggested

using CISS-MRI for detecting the defect,^[3] and Hosokawa *et al.* proposed the use of CISS reverse MRI.^[6] Hingwala *et al.* emphasized the usefulness of a 3D reconstruction image for visualizing the structure adjacent to CSF.^[5] 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.*^[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.

Table 1: Pre- and post-operative findings.

	Presurgery	12 months Postsurgery	3 years Postsurgery
MRI findings			
Interdural fluid collection	-	↓	→
Hemosiderin deposition	-	→	→
CSF findings			
Appearance	Xanthochromia	Clear	-
Protein (mg/dL)	146	54 ↓	-
Total bilirubin (mg/dL)	0.2	0 ↓	-
RBCs (number of cells/mm ³)	4200	000 ↓	-
Initial pressure (cmH ₂ O)	7	15.5	-
Symptoms			
Headache	Progressive	Improved	Improved
Sensorineural hearing loss	Progressive	Worsened	Worsened
Cerebellar ataxia	Progressive	Improved	Improved

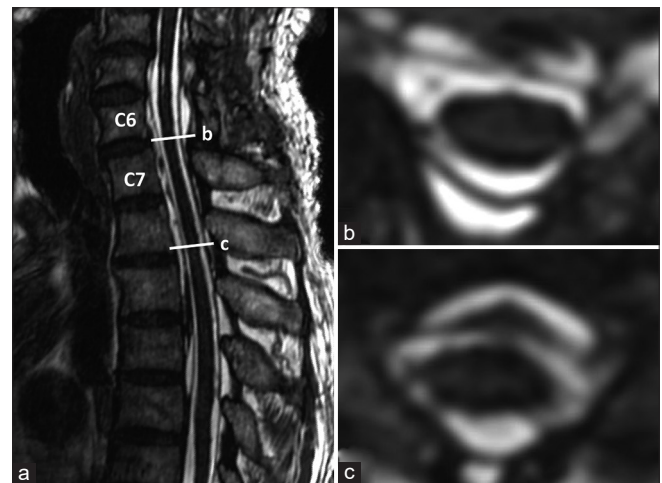


Figure 3: (a) Postoperative sagittal reverse FIESTA-MRI: interdural fluid collection was slightly decreased. (b and c) Axial reverse FIESTA-MRI showing a slight decrease of CSF in the interdural space. Both pictures are cross-sections corresponding to lines in Figure 3a.

Table 2: Review of the representative literature of SS treated by dural closure.

	Imaging modality	Cause of SS	Surgical treatment	Neurological outcome
Kumar <i>et al.</i> (2005) ^[9] (3 cases)	CT (dynamic) myelography	Spinal dural defect (3/3 cases)	Defect closure (muscle piece)	Stabilized (1/3) Not documented (2/3)
Egawa <i>et al.</i> (2013) ^[3] (2 cases)	CISS MRI	Spinal dural defect (2/2 cases)	Defect closure (suture)	Headache improved (1/2), stabilized (1/2) Ataxia and hearing loss worsened (1/2), stabilized (1/2)
Arishima <i>et al.</i> (2017) ^[11] (2 cases)	Spinal endoscopy CT myelography	Spinal dural defect (2/2 cases)	Defect closure (suture)	Headache improved (2/2) Other symptoms stabilized (2/2)
Hosokawa <i>et al.</i> (2018) ^[6] (3 cases)	CISS reverse MRI	Spinal inner dural defect (3/3 cases)	Defect closure (fat/fibrin glue)	Stabilized (3/3)
Present case (2022)	3D-FIESTA-MRI	Spinal inner dural defect	Defect closure (fat/fibrin glue)	Ataxia and headache improved Sensorineural hearing loss worsened

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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Conflicts of interest

There are no conflicts of interest.

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