RESIDENT & FELLOW SECTION

Pearls & Oy-sters: Dural defect repair as treatment for refractory headache from cerebrospinal fluid leak

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Pearls

- CSF leak should be considered in patients with suspected low-pressure, orthostatic headaches and a sacral canal fluid-intensity lesion demonstrated on imaging.
- Advanced imaging modalities, such as CT/magnetic resonance myelogram, and/or surgical exploration may be required to confirm a CSF leak.
- If epidural blood patch is ineffective, surgical repair may be necessary to manage refractory CSF leak.

Oy-sters

- A sacral fluid-intensity lesion on MRI may be mistaken as a perineural (Tarlov) cyst unassociated with CSF leak.
- Patients with CSF leak and suspected low-pressure orthostatic headaches may have a normal MRI of the head in up to 20% of cases.
- MRI spine may not be effective in localizing a CSF leak.

A 19-year-old woman presented to the headache clinic with a 5-year history of daily headaches and lower back pain after falling off a horse and landing on her buttocks. Her medical history was unremarkable. She was diagnosed with migraine and nonspecific musculoskeletal back pain. Ibuprofen provided no relief. She could not attend school due to her symptoms. Her neurologic examination was unremarkable. MRI of the head with and without gadolinium was initially interpreted as normal. MRI of the full spine with and without gadolinium demonstrated a large (85.1 mm × 23.4 mm × 52.9 mm) nonenhancing contained fluid-intensity lesion in the sacral canal space (from the mid-L5 level to S4 segment) (figure, A and B) reported as a Tarlov or arachnoid cyst. No dural ectasia was reported. CT spine demonstrated scalloping and distortion of the sacrum consistent with bone remodeling (figure, C and D). No lumbar puncture or CT myelogram was performed. It was later revealed in history that she had a profound orthostatic (worse upright) component to her headaches.

Based on the clinical–radiologic correlation, a CSF leak was suspected and the patient was treated with targeted epidural blood patch (30 mL) around the site of fluid-intensity lesion (figure, E) with immediate improvement in symptoms. However, symptoms slowly returned to pre-patch baseline over the next 2 years and an exploratory surgery was performed. A nickel-sized dural tear and communication defect (between the large fluid-intensity lesion and the subarachnoid space) were confirmed at the S1 level during the surgery. A diagnosis of a spinal extradural arachnoid cyst was made. Muscle grafts and fibrin glue (3 mL) were used to seal the dural defect plus partial drainage of the cyst. She had complete symptom resolution at her 1-year follow-up and had returned to full-time school. A follow-up MRI spine was declined due to symptoms resolution.

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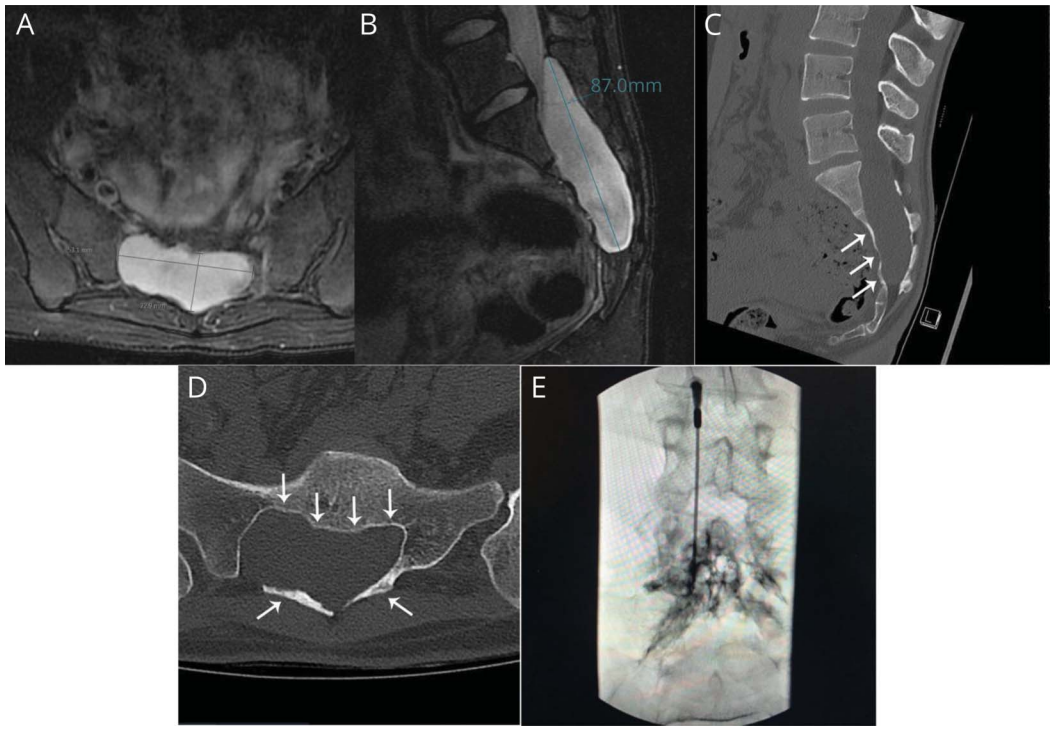
Figure MRI and CT of the spine and epidural blood patch



(A, B) MRI of the spine. (C, D) CT of the spine. (E) Epidural blood patch.

Discussion

This was a suspected case of CSF leak secondary to a traumatic fall. CSF leak can lead to low CSF pressure and debilitating headaches.[1,2] MRI head with and without contrast may demonstrate features suggestive of low CSF pressure such as subdural hygromas, enhancement of the pachymeninges, engorgement of venous plexuses, pituitary hyperemia, and brain sagging. However, up to 20% of patients do not demonstrate the classic MRI abnormalities.[1]

Numerous spinal manifestations of suspected low CSF pressure can be seen such as meningeal diverticula, extrathecal CSF collections, syringomyelia, and retrospinal C1-C2 fluid collections.[1] In our case, the radiographic features and location of the sacral canal fluid-intensity lesion could easily be mistaken as a Tarlov cyst, which was initially reported on the MRI. A Tarlov cyst is a CSF-filled nerve root cyst most commonly found at the sacral level and the exact cause of a Tarlov cyst is unknown. One theory is leakage of CSF into the nerve root sheath after a traumatic injury through an abnormal congenital connection between the subarachnoid space and the area surrounding the affected nerves (perineural region).[3] A Tarlov cyst can become symptomatic and grow due to buildup of fluid from points of minoris resistentiae leading to compression to adjacent structures such as nerve roots. It can rarely be the cause of CSF leak if it ruptures into the extradural space.[3] A spinal extradural arachnoid cyst can be caused by a rare small defect of the dura that leads to CSF accumulation and communication defects between the collection and the subarachnoid space.[4] This is similar to meningeal diverticula in the thoracic spine as a result of protrusion of the arachnoid membrane through spontaneous dural tears and it can be associated with CSF leak.[1,4]

It is difficult to distinguish between the 2 types of cysts or determine the location (intradural or extradural) through MRI alone. A CT or magnetic resonance (MR) myelogram could have provided more information. In our case, a dural defect was confirmed during surgery. It was causing leakage of CSF, which formed a collection in the sacral canal. The size of the collection was substantial and bone remodeling was demonstrated (figure, C and D), which may have accounted for our patient's back pain. This discovery has treatment implications as symptomatic sacral collections or cysts are sometimes managed through drainage. In our case, this could have resulted in worsening symptoms from lowering the CSF pressure further and recurrence of fluid collection in the context of an untreated dural defect.[3,5]

This case illustrates the importance of making a diagnosis based on clinical–radiologic correlation. A CSF leak should be explored in patients with orthostatic headaches (worse upright) and lower back pain in the context of a sacral canal fluid-intensity lesion demonstrated on imaging. Advanced imaging modalities such as CT/MR myelogram or surgical exploration may be required to confirm a CSF leak. MRI head can be normal in cases of suspected low CSF pressure and MRI spine may not be effective in localizing a CSF leak. A misdiagnosis of a benign and simple sacral cyst could lead to suboptimal or inappropriate

management. Identifying a dural defect and CSF leak can lead to appropriate intervention, such as targeting epidural patches at the site of leak, and prevent progression and morbidity.

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

Appendix Authors

Name	Location	Contribution
Tommy Lik Hang Chan, MBBS	Stanford University	Drafted the manuscript for intellectual content
David Dongkyung Kim, MD	Western University	Revised the manuscript for intellectual content

Appendix *(continued)*

Name	Location	Contribution
Syed Hashmi, MD	Stanford University	Interpreted and obtained the images
Ian Carroll, MD	Stanford University	Major role in acquisition of information, revised the manuscript for intellectual content

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