



## Case Report

# Cerebrospinal fluid venous fistula: Illustrative case

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## ABSTRACT

**Background:** Spinal cerebrospinal fluid (CSF) venous fistulas are an under-recognized cause for spontaneous intracranial hypotension (SIH) which may result in postural headaches.

**Case Description:** A 60-year-old-male presented with 6 years of a persistent headache. The initial brain and spine MRIs and conventional CT myelogram (CTM) showed no CSF venous fistula. However, the lateral decubitus dynamic CTM demonstrated a hyperdense paraspinous vein on the right at the T10-T11 level consistent with a CSF venous fistula. It was subsequently successfully treated with surgical ligation. Temporary CSF diversion with lumbar drain was required to treat transient rebound intracranial hypertension.

**Conclusion:** The diagnosis of a CSF venous fistulas is often missed on standard brain and spine MRI imaging, and conventional CTM. Dynamic CTM is a more effective modality to detect CSF venous fistulas. Surgical ligation is a safe and effective treatment option. Patients with long-standing SIH may encounter rebound intracranial hypertension after CSF venous fistula ligation and may require temporary CSF diversion.

**Keywords:** Cerebrospinal fluid leak, CSF leak, CSF venous fistula, CSF-venous fistula, Spontaneous intracranial hypotension

## INTRODUCTION

Cerebrospinal fluid (CSF) leaks, variously attributed to dural/nerve root sleeve defects, meningeal diverticuli, tears in the ventral dura, and CSF venous fistulas are etiologies for spontaneous intracranial hypotension (SIH) and positional headaches. As many as, 25% of patients with SIH may have CSF venous fistulas.<sup>[6]</sup> We present an illustrative case where a patient with long-standing symptomatic SIH attributed to a CSF venous fistula, diagnosed with a lateral decubitus dynamic CT myelogram (CTM), and was successfully treated with surgical ligation.

## MATERIALS AND METHODS

A 60-year-old male presented with a 6-year history of persistent postural headaches. Initial MR, CT studies, and conventional CTM studies showed no evidence of a CSF leak. The repeated lateral decubitus CTM documented multiple T10/T11 dilated nerve root sleeve cysts but no active leak. The brain MRI obtained 4 years after demonstrated stigmata of SIH (i.e., cerebellar tonsillar herniation, sagging of midbrain, diffuse patchy meningeal thickening and enhancement,

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venous engorgement of sella, and prominent dural venous sinuses). He underwent two nontargeted epidural blood patches (EBPs). After the EBP, he experienced rebound intracranial hypertension symptoms including headaches were relieved by standing but worsened in the supine position, as well as bilateral papilledema confirmed on fundoscopic examination.

He was treated with a combination of acetazolamide and a short course of dexamethasone with symptom improvement. One month later, he again underwent a dynamic CTM that demonstrated a hyperdense paraspinous vein on the right at the T10-11 level indicative of a focal CSF venous fistula.

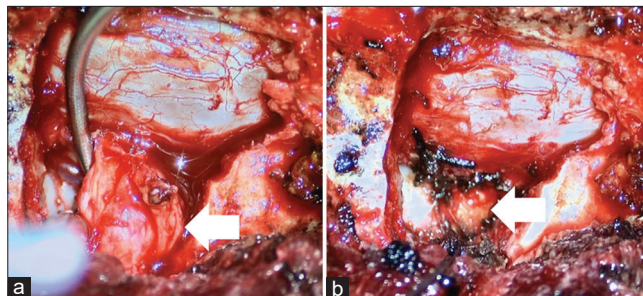
### Surgery and postoperative course

Her underwent a right T10 hemi-laminectomy with complete facetectomy to expose the right T10 nerve root. Dilated epidural veins along with a bulbous nerve root sleeve were clearly visualized. The right T10 nerve root sleeve was temporarily clamped and then fully ligated as there were no changes in neuromonitoring over 10 min [Figure 1]. This was followed by a T10-11 instrumentation and fusion for segmental spinal stabilization. Postoperatively, he again developed intracranial hypertension symptoms as anticipated (headaches and blurry vision) that required a 7-day course of CSF diversion with lumbar drain. He was also on acetazolamide, which was eventually tapered off. He had complete symptom resolution and was doing well at the 1-year follow-up visit.

## DISCUSSION

### Diagnosis of CSF venous fistulas

Diagnosis of CSF venous fistula can be challenging; since in most cases, there is no focal extradural CSF collection to indicate the presence of a spinal CSF leak (i.e., on conventional MR and CT) [Table 1]. However, dCTM may help diagnose CSF venous fistulas by differentiating central CSF venous fistulas from low-flow epidural leaks.<sup>[3]</sup> The “hyperdense paraspinous sign” on CTM/dCTM may prove suggestive of a CSF venous fistula.<sup>[6]</sup> As such, digital subtraction myelography, particularly in the lateral decubitus position, is considered the most accurate imaging modality for diagnosing CSF venous fistula.<sup>[4]</sup> New spinal compliance curves using saline infusion catheters may provide information about pressure-volume dynamics to assist in the diagnosis of CSF venous fistulas on dCTM.<sup>[2]</sup> However, delayed imaging after intrathecal injection, even a few minutes, may prevent fistula detection.<sup>[5]</sup> Finalization of scanning parameters before contrast injection and scanning immediately following contrast injection is essential to increase the likelihood of detecting the CSF venous fistula.<sup>[5]</sup>



**Figure 1:** Intraoperative photographs demonstrating (a) the cerebrospinal fluid (CSF) venous fistula within the associated bulbous thoracic nerve root and (b) ligation of the CSF venous fistula with associated nerve roots. Arrow is pointing to the location of the CSF-venous fistula in (a), where the CSF-venous fistula was ligated in (b).

**Table 1:** Summary of the findings of citations reported in this case report.

Reference	Summary
Brinjikji et al., 2021	Transvenous embolization of the paraspinous vein is a safe and effective treatment of CSF venous fistulas.
Caton et al., 2021	An analysis of craniospinal space compliance curves provides qualitative and quantitative information about pressure-volume dynamics and may aid as a diagnostic tool in patients with known or suspected CSF venous fistulas.
Kranz et al., 2017	CSF venous fistulas are an important cause of spontaneous intracranial hypotension and may be detected on both CT and dynamic myelography.
Madhavan et al., 2020	CSF venous fistulas are a difficult to diagnose, relatively uncommon cause of spontaneous CSF leaks that respond well to surgical treatment.
Mamlouk et al., 2021	Decubitus CT myelography allows for the diagnosis of CSF venous fistulas using gravity to dependently opacify the CSF venous fistula, which can be missed on traditional myelographic techniques.
Schievink et al., 2019	Hyperdense paraspinous vein signs can be seen in CSF venous fistulas, and they can be treated successfully with either surgery or glue injection.

CSF: Cerebrospinal fluid

### Treatment options

Surgical ligation and endovascular embolization are both effective treatment options.<sup>[1]</sup> Surgery involves eliminating the abnormal connection between the subarachnoid space and epidural venous plexus by ligating other nerve root proximal to the dorsal root ganglion. Endovascular management involves catheterization of the azygous vein followed by selective catheterization of the paraspinous vein and embolization of the vein with Onyx.<sup>[1]</sup> One preliminary

case series of five patients documented complete resolution of headaches in four patients and a 50% decrease in headaches in the remaining patient.<sup>[1]</sup>

## CONCLUSION

CSF venous fistula is increasingly recognized as a cause for SIH. Dynamic CTM is a more effective modality to detect CSF venous fistulas. Surgical ligation is a safe and effective treatment option. Patients with long-standing SIH may encounter rebound intracranial hypertension after CSF venous fistula ligation and may require temporary CSF diversion.

## Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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