Identification and surgical ligation of spinal CSF-venous fistula

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INTRODUCTION

Spontaneous intracranial hypotension/hypovolemia (SIH) often presents with positional or orthostatic headaches. The positional component may be subtle, evolve gradually, or be absent entirely but is usually part of the diagnosis.[7,11,12,14,20,22] The headache is often triggered or exacerbated by a cough or Valsalva maneuver.[5] SIH is somewhat of a misnomer given that opening CSF pressure is normal (7–20 cm of H₂O) in the majority of cases[11,12,17] and it is most often due to spontaneous CSF leaks; thus, low intracranial CSF volume is another description.[11,21] Non-specific symptoms may be present, including neck or face stiffness, facial pain or numbness, tinnitus or muffled sounds, visual changes such as blurry vision, paresthesia of upper or lower extremities, hand weakness, brain fog, and gait disturbances.[29,30,33]
Schievink et al. classified spontaneous CSF leaks into four categories [Table 1]. CSF leaks in the skull base often present with CSF rhinorrhea or otorrhoea, especially when the patient bends forward, and are not always associated with positional headaches.[31]

SIH due to CVF presents both diagnostic and therapeutic challenges because positional headaches also occur in postural orthostatic tachycardia syndrome,[19] tonsillar herniation seen in chiari malformation,[33] cervicogenic headaches, and other primary headache disorders including new daily persistent headache.[19] Moreover, the radiographic characteristics of SIH, subdural fluid collection, enhancement of pachymeninges, engorgement of venous structure, pituitary hyperemia, and sagging of the brain (SEEPS)[32] are only indirect clues to an underlying leak. The MRI finding of cerebellar ectopia alone, in a patient with orthostatic headaches, may erroneously point to an etiology that may not be the primary reason for patients’ symptoms. Therefore, ascertaining the underlying cause is critical to establish the diagnosis and initiate appropriate treatment.

Spinal MRI can determine if a large spinal fluid collection is present and may suggest a region of the spine where the leak is located but, in general, cannot pinpoint a leak site. CT myelography (CTM) is the first-line imaging study to diagnose a leak site; however, CTM may be normal in patients with CVF. Hyperdense contrast in the renal collecting systems can suggest an underlying leak but does not help to localize the site.[33] Digital subtraction myelography (DSM), which carries similar risks as conventional myelography, has made it possible to identify CVF through real-time high-resolution imaging utilizing intrathecal contrast.

MATERIALS AND METHODS

We present three cases of CVF, discuss their management, and review the current CVF literature. Furthermore, we propose a systematic approach to the evaluation and management of this unique and potentially treatable cause of SIH.

RESULTS

Clinical vignettes

Case 1

A 25-year-old female presented with a 3-year history of orthostatic headaches with brain MRI findings of pachymeningeal enhancement, brain sagging, pituitary engorgement, and venous distention that were suggestive of SIH. Her CTM was unrevealing. She underwent three blood patches without significant symptomatic improvement. Two years later, she presented with hand numbness, right-sided headaches, and paresthesias of all four extremities and was found to have a large cervicothoracic syrinx. There were no occipital headaches, and a neurological examination was normal except for decreased sensation. She had a Chiari-1 decompressive surgery that did not relieve her symptoms. She underwent a magnetic resonance myelography (MRM) that revealed a T7-T8 meningeal diverticulum and an enhancing left paraspinal vein draining into the azygos system [Figure 1]. Opening CSF pressure was <4 cm of H2O [Table 2]. The preoperative angiogram had confirmed that the artery of the Adamkiewicz was at L2. There was another spinal artery at left T8, one level below the site of the fistula.

Surgical procedure and follow-up

She was brought to the operating room supine, intubated with arterial and venous access established. After preoperative antibiotics and steroids administration, fluoroscopic guidance was utilized to localize the T7-T8 interspace where the T7 nerve root exits. The access was gained through a prior surgical incision, which was extended superiorly and just lateral to the dural sac to expose the nerve root complex. There was significant venous engorgement, which was coagulated and dissected around the nerve root to make room for placing a temporary aneurysm. During the 10 min wait, the somatosensory evoked potentials (SSEPs) and motor evoked potentials (MEP) had remained stable. A permanent aneurysm clip was then placed and further reinforced with a zero-silk tie around the nerve root. The area was thoroughly irrigated with antibiotic solution and packed with muscle and Tisseel. The wound was closed in layers using 3-0 Vicryl for deeper layers and with subcuticular 4-0 Caprosyn. She tolerated the procedure well; her intraoperative electrophysiological monitoring remained stable throughout the procedure. She was extubated and transferred to the recovery room.

The patient’s headaches, hand numbness gradually resolved, and within 3 months, she was asymptomatic. Repeat MRI showed a complete resolution of the syringomyelia and ascent of the herniated cerebellar tissue [Table 2].

| Table 1: Classification of spontaneous CSF leaks, adopted from Schievink et al.[19] |
|---------------------------------------------------------------|
| **Type 1** | Dural tear |
| a. Ventral | |
| b. Dorsolateral | |
| **Type 2** | Meningeal diverticulum |
| a. Simple | |
| b. Complex | |
| **Type 3** | CSF-venous fistula |
| **Type 4** | Indeterminate/unknown |
| **Extradural CSF** | Yes: positive |
| No: negative | |
A 64-year-old male presented with a prolonged history of migraines and severe vertex headaches aggravated by bending, coughing, sneezing, and straining. His positional symptoms and MRI brain findings (SEEPS) suggested SIH. He received a short course of steroids and two blood patches without relief of his symptoms. On MRM, he was found to have a left T7-T8 spinal meningeal diverticulum, a vascular malformation of the lower thoracic spine, and contrast opacification extending along the left paraspinal vein into the azygous system, representing a CVF [Figure 2]. The opening CSF pressure was 9 cm of H₂O [Table 2]. Preoperative spinal angiogram and embolization were performed, during which a small coil was placed for easy intraoperative identification. He subsequently underwent T7-T8 hemilaminectomy for CVF repair.

Surgical procedure and follow-up

As described above, after intubation, arterial and venous access, and administration of preoperative antibiotics, the fluoroscopic guidance was utilized to identify the small coil placed during the preoperative angiogram. The level was further confirmed by counting vertebral bodies and ribs corresponding to the level of the coil. Through a midline incision, the dissection was performed along the spinous process over the lamina, above and below the targeted nerve root of the fistula. Using a Cloward retractor, lamina was exposed, and the hemilaminectomy from outside to inside fashion was performed to identify the medial dura of the thecal sac. Kerrison punches were used to unroof the bone up to expose the lesion with large veins and the dilated nerve. Under the operating microscope, enough space was created around the lesion allowing for two 10 mm straight aneurysm clips to be placed to disrupt the connection between the nerve root and the vessel. After a wait of 10 min to confirm that MEPs and SSEPs remained normal, a permanent aneurysm clip was placed, which was further reinforced with a 2-0 silk tie. The dilated nerve root was further cauterized, and the area was packed with muscle, and the dural sealant was applied. After copious antibiotic irrigation, the wound was closed in layers using 3-0 Vycril sutures and subcuticular 4-0 Caprosyn for the skin. He was extubated and transferred to the recovery room. On follow-up, his severe headaches and tinnitus had resolved [Table 2].

Case 3

A 54-year-old female presented with unremitting headaches, and brain MRI findings (SEEPS) were suggestive of SIH. She underwent multiple blood patches without relief of her symptoms. An MRM was performed, revealing bilateral extensive spinal meningeal diverticula with a possible CVF at T9-10, for which she underwent a right T9 nerve root ligation. However, her symptoms recurred shortly thereafter, and a follow-up CTM demonstrated a left T9-T10 spinal meningeal diverticulum with opacification of an adjacent
Figure 2: Case 2. MR Myelogram axial T1 VIBE (a-d) demonstrates an irregular left T8-T9 spinal meningeal diverticulum (solid arrow) and opacification of an adjacent left paraspinal vein extending into the azygous system (dotted arrow), suspicious for a CSF-venous fistula.

Figure 3: Case 3. Axial CT Myelogram demonstrates a left T9-T10 spinal meningeal diverticulum (solid arrow) and opacification of an adjacent left paraspinal (dotted arrow), suspicious for a CSF-venous fistula.

left paraspinal vein, suspicious for a CVF [Figure 3]. Her opening CSF pressure was <4 cm of H2O [Table 2]. She, therefore, underwent a left T9 foraminotomy with clipping of her fistula with an aneurysm clip.

Surgical procedure and follow-up

Since the patient had prior surgery on the right side at that level, the same incision was used to gain access down to the fascia. Fluoroscopic guidance was used to identify the level utilizing the clip as a marker that was placed on the contralateral side. The level was further confirmed using the visual anatomic landmarks, identifying the left TP at T9, and dissected laterally without identifying the dura in the midline to a straight lateral approach. The entire surgery was performed under the operative microscope, the T9-T10 joint space corresponding to the nerve root was identified, and a small bony window was created. Hemostasis using bone wax and coagulation of epidural veins was achieved, and the plane around the nerve root was cleared.

A large straight nonfenestrated clip across the nerve root complex was applied. The MEPs and SSEPs remained stable during the 5 min wait time; therefore, the clip was left in place. The venous structures lateral and deep to the nerve root clip were further coagulated, and the area was packed with muscle and Tisseel to obliterate the region completely. The area was copiously irrigated with antibiotic solution and then closed in layers, using Vicryl 0 and 2-0 for deep layers and the skin with 3-0 Vicryl and a 4-0 Caprosyn. She tolerated the procedure well, was rolled onto her back, extubated, and moved to the recovery room without any complications.

Her postoperative recovery has been unremarkable, and all her symptoms have completely resolved. Her tonsillar herniation persists, but she remains asymptomatic [Table 2].

DISCUSSION

Spontaneous CVF was first described in a small case series as a cause of SIH.\textsuperscript{[30]} Since then, several studies have followed.\textsuperscript{[5,7,9,11,24,27,29]} Whereas the majority are believed to be spontaneous due to arachnoid granulations and dilated epidural veins, iatrogenic CVF have also been reported following myelography.\textsuperscript{[14,18]}

According to recent estimates, spinal CVF may represent between 2.5% and 20% of SIH cases.\textsuperscript{[3,23,30]} Small case series and retrospective studies have shown that CVF are more common in females in their early fifties and have a predilection for the thoracic spine, especially the lower thoracic levels.\textsuperscript{[8,11,12,16,23,29]} The cervical or lumbar spine are unusual locations for CVF.\textsuperscript{[5,7,11]} These findings are further confirmed by Duvall et al., in their series of 53 cases of CVF out of a total of 156 SIH patients with a prevalence of 35%, which could be an overestimation due to a selection bias.\textsuperscript{[5]} However,
a large study of 568 cases of SIH has noted CVF to be present in 2.5% of cases – where extradural CSF pooling was present in only 5% of the cases – therefore making their prevalence to be one fifth or one-fourth of all cases of SIH.\textsuperscript{[3,6,23,26,29]}

In addition, a series of 22 cases also found that 90% of them lacked clear CSF leak, and 82% were associated with a dural sleeve diverticulum.\textsuperscript{[2,11]} The opening CSF pressure was often normal, with a mean of 7.8 cm of H$_2$O.\textsuperscript{[1,38]} Of the five SEEPS criteria, pachymeningeal enhancement (73%), venous distension sign (77%), and brain sagging (64%) have been found to be more consistent signs, along with a low to normal CSF opening pressure. In our series, all patients presented with these signs and a low to normal CSF opening pressure that prompted further investigation, and the diagnosis was confirmed using MRM.

CVF may be associated with a single draining vein or a network of dilated veins surrounding the spinal nerve root sleeve in the form of a diverticulum leading to the rapid absorption of extravasated CSF within the venous system.\textsuperscript{[28,32]}

The locations of abnormal venous plexuses associated with CVF as detected on CTM were paravertebral (45%), central within the cord (32%), and lateral to the spine (23%).\textsuperscript{[7]}

Since CVF are a potentially treatable cause of protracted SIH, we propose actively looking for an enhancing paraspinous vein when performing CTM or MRM, which is reported to be an important diagnostic sign.\textsuperscript{[1,3,27]} A series designed specifically to look for a hyperdense paraspinous vein (HDPSV) sign showed that it was present in 7% of SIH cases on CTM, and none were found in the control group, thereby making it a highly probable\textsuperscript{[38]} but a nonspecific sign of CVF. However, Schievink \textit{et al}. report that HDPSV does not always correspond to the exact location of the fistula, occasionally present on the contralateral side or a level above or below that of the CVF.\textsuperscript{[27]}

In a series of 53 patients, Schievink \textit{et al}. made a case for lateral decubitus DSM due to its rapid imaging and high temporal and spatial resolution when compared to conventional imaging. In addition, they found that conventional CTM or MRI failed to locate the CSF leak in the majority (80%) of cases. Ten of the 53 patients in their series were confirmed to have CVF, concluding the prevalence to be about 19%, which may be an overestimation due to selection bias. Similar to the findings of Kranz \textit{et al}.\textsuperscript{[3,7]} they also found that their cohort had a mean age of 52 years, with the majority being in females (9/10) and the majority (7/10) having low opening pressure (<6 cm of H$_2$O).\textsuperscript{[26,29]}

In our case, we found MRM to be particularly sensitive in identifying the site of the leak. In addition, CTM and DSM exposes patients to a significant amount of ionizing radiation, which can be avoided with MRM that only uses intrathecal gadolinium contrast.\textsuperscript{[14]}

The literature describes the two techniques used to delineate leaks are “initial” and “problem-solving.” Various authors note CTM to be the initial technique that helps approximate the lesion, whereas problem-solving techniques such as DM and DSM help pinpoint the lesion.\textsuperscript{[10]} This contrasts with our experience where MRM was able to locate the lesion in all cases exactly. The patients in our cohort initially received CTM, which was inconclusive. An MRM was performed that revealed the site of CVF. One patient received a Chiari decompression surgery only to have her symptoms recur, at which point, she underwent a laminectomy. A CVF was subsequently encountered after exploration and treated permanently with surgical nerve root ligation with an aneurysm clip and a silk tie, highlighting the fact that MRM can be helpful in diagnosing and treating patients.

It has also been reported in a small case series that CVF present with accompanying venous or venolymphatic vascular malformations.\textsuperscript{[23]} The vascular malformation, which was noted only in the first case in our series, was suspected to be an independent finding with no relation to CVF etiology. In addition, it has also been suggested that type 1 leaks arising from a dural tear predispose to the formation of type 3 leaks, that is, CVF may result from communication between arachnoid granulations and dilated perispinal vascular channels. DM or DSM are best suited to detect fistulas of this sort.\textsuperscript{[14]}

There is little consensus in the literature on how to best diagnose these CVF; some authors suggest lateral decubitus CTM, whereas others propose DSM to increase their detection. However, both of these techniques expose the patient to a substantial amount of ionizing radiation, and DSM is operator-dependent. In contrast, we were able to localize CVF using MRM by off-label but well-tolerated gadolinium injection intrathecally, under CT guidance, thereby preventing any extensive deleterious effects resulting from this approach. The patient was rolled multiple times after the starting Trendelenberg position to disperse the contrast in the subarachnoid space before transferring to the MR suite to perform MRM on a 3T Biograph mMR scanner (Siemens). Multiple fat-suppressed T1 sequences were reviewed in real-time as well as delayed thin-section axial sequences were obtained.\textsuperscript{[1]} Based on our experience, we propose that patients presenting with symptoms suggestive of SIH should receive routine CT or MRI imaging to look for SEEPS. If pachymeningeal enhancement, brain sagging, and distended vein sign are found, a presumptive diagnosis of SIH should be made, and a search for an active leak and exclusion of other etiologies should ensue. This should be followed with MRM with gadolinium contrast injected intrathecally, and active extravasation and location of HDPSV should be located. Once the CVF is located, the patient should be counseled for surgical management with nerve root ligation using an aneurysm clip and/or a silk tie.
as noninvasive therapies such as blood patching and fibrin glue are rarely useful long term.\textsuperscript{[3]} However, some authors advocate that a targeted blood patch should be tried first because it has reasonable success when a leak is identified before moving to a more invasive surgical procedure which nonetheless can be curative.\textsuperscript{[2,11,34]} Kumar et al. describe a patient with SIH and IVC occlusive disease, in whom findings of SIH reversed on IVC endovascular stenting. While not specifically imaged, the authors hypothesized the SIH occurred due to a CVF, and endovascular stenting served as a treatment for this patient’s SIH. Therefore, they conclude that CVF can be considered as a differential consideration when SIH is identified in the setting of IVC occlusive disease, with targeted endovascular obliteration serving as a possible therapeutic option.\textsuperscript{[15]}

On the other hand, if no enhancing paraspinal vein or a leak is identified on MRM, other diagnoses should be considered. If a leak is identified, the patient should receive a trial of a blood patch or fibrin glue. If, however, a leak is not identified, follow-up DSM should take place. Alternatively, if no HDPSV is found, but a leak is identified, proceed directly to blood patch or fibrin glue injection because the likelihood of a CVF is low and the chance of sealing the dural defect causing SIH is not insignificant. However, if the patient continues to have symptoms, definitive treatment endovascularly or with surgical repair, as shown below, should follow [Figure 4].

The use of aneurysm clips has emerged as an excellent treatment choice that prevents needle holes encountered while stitching the dura. Prior authors have suggested "ligating"\textsuperscript{[14]} or "sacrificing"\textsuperscript{[16]} the nerve root when the risk of neurological impairment is minimal; however, specific techniques have not been expounded upon extensively in the literature. A study by Wang et al.\textsuperscript{[16]} describes the following ligation method: a 2-0 silk suture tied around the nerve root with subsequent proximal followed by distal division of the nerve root. They propose aneurysm clipping only if necessary, especially at the proximal portion of the divided nerve root.\textsuperscript{[16]} Our approach was similar to this method; however, our primary method of ligation was that of clipping. The rationale is based on the anatomic pathology being treated. Aneurysms are diverticula, and CVF are abnormal communications associated with a diverticulum, so it is reasonable to treat them similarly.

Fistula clipping in our series was performed by first locating the fistula site using MRM. A preoperative angiogram identified the location of the artery of Adamkiewicz, and a coil was placed to mark the fistula site. Dissection was performed as medially as possible lateral to the dural thecal sac. Given that venous engorgement may occlude visualization of the fistula, these veins were coagulated, dissected with a Rhoton instrument, and the nerve hooks were placed around the nerve root complex in this instance. Case 1 exemplifies this. Once located, the fistula was clipped using a temporary aneurysm clip followed by neuromotor monitoring to confirm intact function [Figure 4]. This was followed by permanent clipping, which was followed by a 2-0 silk tie reinforcement when needed. Cases 1 and 2 were treated with both interventions, while Case 3 only required a clip.

On follow-up, patients reported complete resolution of their preoperative symptoms, suggesting that clipping the fistula with a permanent aneurysm clip with or without a 2-0 silk suture reinforcement shows promise in treating SIH caused by CVF. Although our cohort is small, we argue that ligation of the fistula with preferably two forms of occlusion, with clipping being the predominant form, shows promise in permanent amelioration of symptoms.

**CONCLUSION**

We present three patients treated for SIH caused by CVF and discuss their clinical presentation, diagnostic challenges, and management in the light of current literature. A systematic approach leads to a search for enhancing paraspinal vein and direct visualization using real-time MRM. This approach can obviate the need for unnecessary surgery. We found MRM to be quite helpful in visualizing the HDPSV as well as locating the fistula site. We propose using DM/DSM in only those cases where MRM has been inconclusive in resolving the site of the fistula. The intraoperative localization can be further aided by placing a coil at the fistula site during preoperative angiography. The definitive surgical management should proceed with ligation of the defect using one or more permanent aneurysm clips after ensuring normal intraoperative monitory when temporary aneurysm clips are placed. Reinforcement with a 2-0 silk suture adds another layer of securing the fistula to permanently ameliorate patients’ symptoms. This technique seems to have long-term efficacy in ameliorating symptoms of SIH.

**Declaration of patient consent**

Patient’s consent not required as patients identity is not disclosed or compromised.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.
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How to cite this article: Majeed K, Hanz SZ, Roytman M, Chazen JL,
Greenfield JP. Identification and surgical ligation of spinal CSF-venous
fistula. Surg Neurol Int 2021;12:514.