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CLINICAL REPORT
SPINE IMAGING AND SPINE IMAGE-GUIDED INTERVENTIONS

Spontaneous Intracranial Hypotension Associated With Vascular Malformations

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ABSTRACT

SUMMARY: Spinal CSF leaks from dural tears or CSF-venous fistulas are the most common causes of spontaneous intracranial hypotension. Rarely, CSF leaks have also been associated with vascular malformations, which have primarily been discussed in case reports or small series. In this clinical report, we report the clinical features, imaging findings, and treatment of 6 children and adults with CSF leaks associated with vascular malformations in the spine and skull base depicted on CT myelography and cisternography.

ABBREVIATIONS: SIH = spontaneous intracranial hypotension

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INTRODUCTION

Spontaneous intracranial hypotension (SIH) is most commonly secondary to a spinal CSF leak either from a dural tear or CSF-venous fistula.[1] Apart from these spontaneous defects, SIH has also been described with vascular malformations, namely venous and lymphatic malformations, that are either isolated lesions or in patients with syndromes.[2-7] Despite the prior reports, understanding of these diseases has been hampered by incorrect nomenclature. Regarding SIH, previous publications have inappropriately reported vascular malformations associated with "Chiari 1 malformation", when in fact the lesions were resulting in SIH.[8, 9] Vascular malformations are also often mischaracterized despite nomenclature from the International Society for the Study of Vascular Anomalies (ISSVA).[10] For example, CSF leaks from underlying "hemangiomas" and "lymphangiomatosis" have been described,[9, 11] which now may not be appropriate nomenclature, owing to our greater understanding of the genomics of vascular malformations. Lastly, myelographic evidence of CSF leaking into a vascular malformation has only been briefly reported in some reports. In this clinical report, we present 6 cases of SIH secondary to spinal or skull base CSF leaks from an underlying vascular malformation and discuss the proper terminology, myelographic techniques, and possible therapeutic options.

Case Series

Institutional review board approval was granted, which waived the right for informed consent. The study population consisted of SIH patients associated with a vascular malformation between 2000-2024. Inclusion criteria were the following: (1) diagnosis of SIH according to the International Classification of Headache Disorders, 3rd edition,[12] (2) spinal or skull base CSF leak identified on CT myelography or cisternography; (3) vascular malformation appropriately categorized according to the latest ISSVA classification after multi-disciplinary review in the vascular anomalies clinic or by a radiologist experienced with vascular anomalies. Brain MRIs were assessed using the SIH Bern scoring system.[13]

Six cases of CSF leaks associated with an underlying vascular malformation were identified (Table). There were 4 females and 2 males, and the mean patient age was 20.8 years (range, 7-40 years). Three of the patients were pediatric (<18 years). Four of the CSF leaks were in the spine, and 2 were in the skull base. There were 3 CSF leak cases with a venous malformation and 3 with a lymphatic malformation, and all 3 patients with a lymphatic malformation had a complex lymphatic anomaly.[14] All patients had a paraspinal or skull base vascular malformation. Two of the patients were previously reported by our group and are being included to evaluate this unique cohort holistically.[6, 7]

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Table: Patient demographics of CSF-vascular malformation fistulas

Case	Age (yr)	Sex	CSF Leak Location	Vascular Malformation	Syndrome	**Myelographic/ Cisternographic Findings**	**SIH Bern Score & Other Findings**	**Treatment and Outcome**
1	23	F	T11-T12 neural foramen	Venous	None	Delayed leakage from the thecal sac to the venous malformation	6, syrinx, large paraspinal venous malformation in right thoracolumbar spine	Fibrin glue patch with moderate relief and improved brain MRI findings
2	40	M	T1-T2	Venous	None	Right T1-T2 CSF-venous fistula	6, syrinx, upper thoracic dorsal epidural venous malformation	Fibrin glue patch with cure
3	33	M	Temporal skull base	Venous	None	Leakage into left retromandibular venous malformation. Left temporal dural ectasia.	4, syrinx, extensive left temple, facial, neck venous malformation	Targeted sclerotherapy without significant relief
4	13	F	Petrous apex	Lymphatic	Generalized lymphatic anomaly	Brisk leakage from the left petrous apex extending to neck musculature to the C3 level	4, syrinx, diffuse intraosseous lymphatic malformations in the skull base, spine, viscera, and right lower extremity	Surgery being planned
5	9	F	T10-T11	Lymphatic	Kaposiform lymphangiomatosis	Brisk leakage from a right T10-T11 meningeal diverticulum to a paravertebral lymphatic malformation	6, syrinx, neck and visceral lymphatic malformations	Blood patches with transient relief; Surgery with cure. Improved brain MRI findings.
6	7	F	L4-L5	Lymphatic	Gorham Stout	Brisk leakage into the right L4-L5 paraspinal soft tissues and bones	6, syrinx, extensive destructive intraosseous lymphatic malformations	Blood patches with transient relief; *n*-BCA liquid embolic patch with marked improvement. Improved brain MRI findings.

Case 1

A 23-year-old female presented with 8 months of positional headaches that prompted an MRI of the brain and spine that showed severe brain sag, cerebellar tonsillar descent, and a syrinx, yielding the initial diagnosis of "Chiari 1 deformity". She was being evaluated by the neurosurgery service for potential suboccipital decompression. Follow up brain imaging was interpreted by a member of the CSF Leak team, and a diagnosis of SIH was made with a Bern score of 6 without dural enhancement (FIG 1). In review of spine imaging at 18 months of age, there was an enhancing epidural lesion in the thoracolumbar spine that was thought to represent an epidural hematoma or a "hemangioma". Twenty-one years later, she presented with her headache and had a spine MRI that showed marked dural ectasia at the T12 level with a previously undiagnosed paraspinal venous malformation. A right decubitus CT myelogram was performed with initial scanning of the thoracolumbar spine that showed subarachnoid contrast at the T12 dural ectasia but no leakage. Delayed imaging after three minutes and an additional small volume of contrast showed a CSF leak in the right T11-T12 neural foramen commencing from the thecal sac and extending to portions of the venous malformation near the pleura. A targeted fibrin glue patch was administered with moderate relief in symptoms to date 3 months after the procedure.

Case 2

A 40-year-old male with a longstanding history of "Chiari 1 deformity" presented to our CSF leak program after a suspicion of SIH was made by the clinical team. Brain and spine MRIs showed severe brain sag, syrinx, and a Bern score of 6 without dural enhancement. Review of the medical records stated he had a suboccipital decompression at the age of 22 that did not resolve the brain sag or syrinx. Seven years later, the patient had a T1-T3 decompression for a dorsal epidural lesion that was diagnosed radiologically and at pathology as a "hemangioma". In retrospect, this lesion represented a venous malformation (Supplementary Fig 1). After presentation to our CSF leak program, a right decubitus CT myelogram was performed and showed a right T1-T2 CSF-venous fistula that was successfully treated with targeted fibrin glue patching. Although the CSF-venous fistula did not drain into the venous malformation given its prior resection, it was thought to be related given it was located at the same thoracic level and the intracranial SIH findings were persistent despite the prior surgery. The follow up decubitus CT myelogram showed resolution of the CSF-venous fistula.

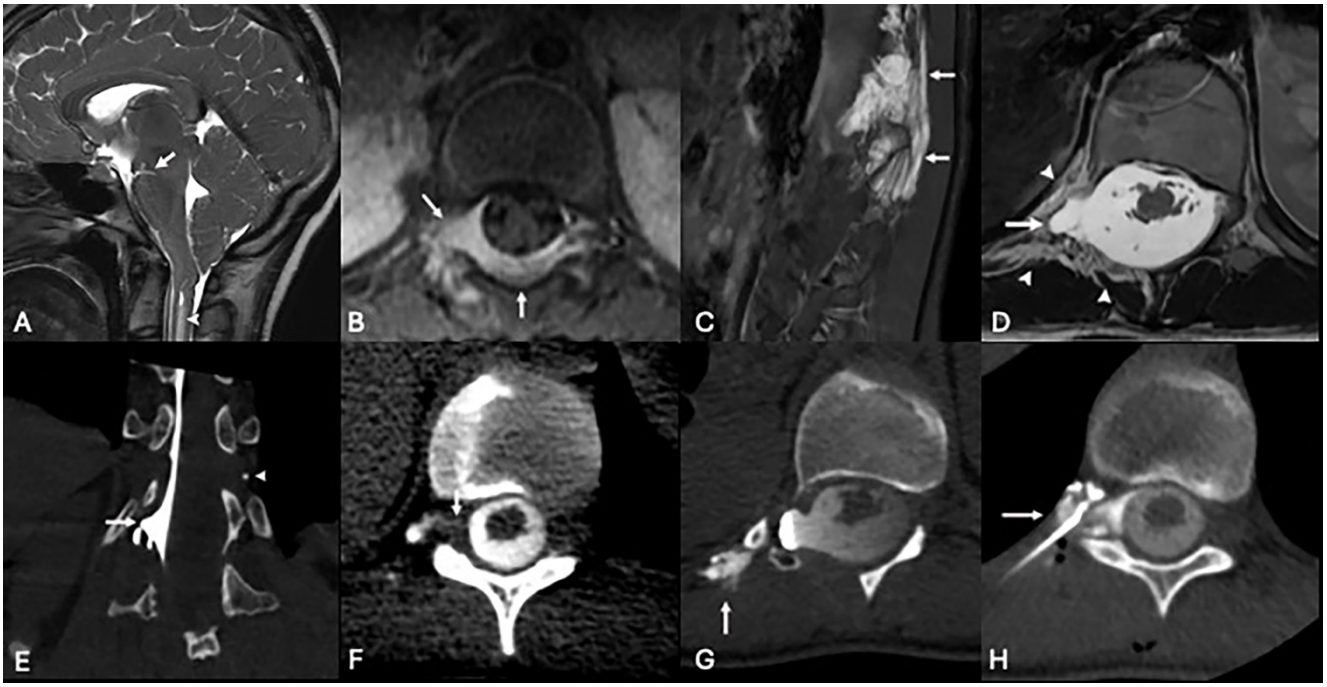

FIG 1. Patient 1: Spinal CSF-venous malformation fistula. A 23-year-old female had a history of an "epidural hematoma" at 18 months of age. 21 years later she developed postural headaches. *A*, Sagittal T2-weighted image of the brain shows severe mid brain sag (arrow), along with cerebellar tonsillar descent and a cervical cord syrinx (arrowhead). *B*, Axial contrast-enhanced fat-suppressed T1-weighted image at 18 months of age shows an enhancing lesion in the T12 epidural space (arrows) that extended into the right paraspinal soft tissues. C, Sagittal short tau inversion recovery image as an adult shows a large right paraspinal lesion (arrows) that was consistent with a venous malformation. *D*, Axial T2-weighted image shows the paraspinal venous malformation (arrowheads) and right dural ectasia at the T12 level (arrow). *E*, Right decubitus CT myelogram in the coronal plane shows opacification of the right T12 dural ectasia (arrow) but no leakage. A left-sided phlebolith is noted (arrowhead), which is consistent with the underlying paraspinal venous malformation that extended to the left side. *F*, *G*, Delayed myelographic images 3 minutes later show a right T11-T12 fistula between the thecal sac and the venous malformation (F, arrow) with additional opacification of the venous malformation more laterally near the ribs (G, arrow). *H*, Fibrin glue patch filling the CSF-venous malformation fistula (arrow) provided moderate relief.

Case 3

A 33-year-old male with a large left scalp, facial, and neck venous malformation presented with headaches and upper and lower extremity numbness. MR imaging of the brain and neck (Supplementary Fig 2) showed an extensive venous malformation along the left temporal scalp, erosion of the calvarium, and contiguity with the brain parenchyma. Imaging also showed severe brain sag, cerebellar tonsillar descent, and a cervical syrinx that was diagnosed as a "Chiari 1 deformity". Scans showed a Bern score of 4 without dural enhancement. A suboccipital decompression was performed that did not resolve the radiologic and clinical findings. Bilateral decubitus myelograms did not show a spinal CSF leak, but a CT cisternogram showed leakage into the venous malformation within the retromandibular fossa. Dural ectasia was also observed along the left temporal lobe with multiple cephaloceles. Targeted sclerotherapy has been performed for the venous malformation, but the skull base leak has not yet been repaired.

Case 4

A 13-year-old female presented with headaches and facial numbness. She was previously diagnosed with generalized lymphatic anomaly, which was first suspected after presenting with diffuse microcystic lymphatic malformations in the right lower extremity and pelvis with a resultant bladder fistula that caused chyluria. Multiple hematologic and electrolyte abnormalities were present on laboratory analysis, which were in keeping with generalized lymphatic anomaly as opposed to Gorham Stout disease, another complex lymphatic anomaly. Her brain and spine MRI showed brain sag, cerebellar tonsillar descent, and a syrinx, yielding the initial diagnosis of "Chiari 1 deformity" (FIG 2). The case was evaluated in the Vascular Anomalies Clinic, and SIH was diagnosed with a Bern score of 4 without dural enhancement. The spine MRI showed diffuse intraosseous lymphatic malformations but no extradural collection or large meningeal diverticula. Bilateral decubitus CT myelograms were performed, which did not reveal a CSF-lymphatic malformation fistula. In the same procedural session, the skull base was imaged and revealed frank CSF leakage from the left petrous apex that extended into the left neck musculature to the C3 level. In retrospect, the CSF leak was observed in the neck on the myelograms, but the leak site was not seen due to incomplete visualization of the skull base. The prior brain MRI was also reviewed and showed a lymphatic malformation in the left petrous apex. Skull base surgery is planned for treatment.

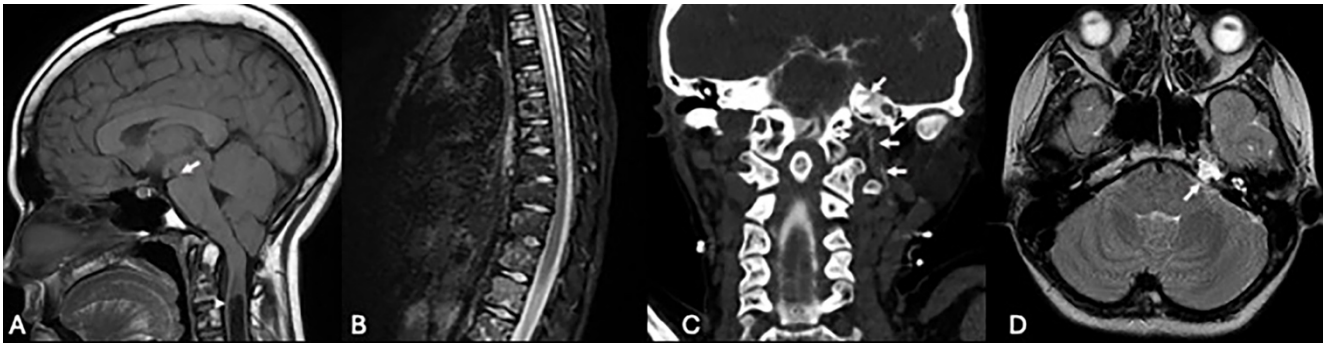

FIG 2. Patient 4: Skull base CSF-lymphatic malformation fistula in a 13-year-old female who had a history of generalized lymphatic anomaly for multiple years who presented with more recent onset of headaches. *A*, Sagittal T1-weighted image of the brain shows mid brain sag (arrow), along with cerebellar tonsillar descent and a syrinx (arrowhead). *B*, Sagittal short tau inversion recovery image of the thoracic spine shows diffuse intraosseous lymphatic malformations. Bilateral decubitus CT myelograms did not show a CSF fistula (not shown), but a CT cisternogram of the skull base (C) showed frank leakage from the left petrous apex into the left neck musculature (arrows) that extended to the C3 level. *D*, Axial T2-weighted image shows a left petrous apex lymphatic malformation (arrow) that was observed in retrospect.

Case 5

A 9-year-old female with history of kaposiform lymphangiomatosis, which was diagnosed during infancy after presenting with Kasabach-Merritt phenomenon and diffuse lymphatic malformations throughout the body, endorsed headaches for several years that were initially thought to be migraines. Brain MRI was eventually performed, and a diagnosis of SIH was rendered with a Bern score of 6, including severe brain sag but no dural enhancement. Spine MRI showed a syrinx and diffuse intraosseous lymphatic malformations but no extradural collection or visible meningeal diverticula. A ventral prone CT myelogram was performed that showed rapid CSF accumulation in the right T10-T11 neural foramen and paraspinal soft tissues that opacified the thoracic duct to the level of the left innominate vein, resembling a lymphangiogram (FIG 3). In retrospect, a paraspinal lymphatic malformation on the prior MRI was visible at the right T10-11 level. Treatment with targeted blood patches provided incomplete relief; therefore, surgical ligation of the CSF lymphatic fistula resolved symptoms after a period of rebound hypertension.

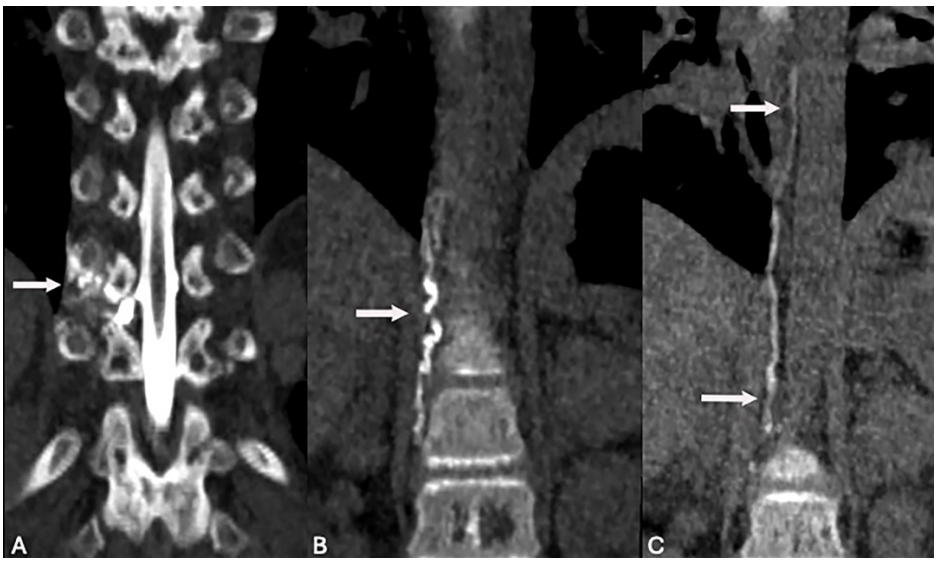

FIG 3. Patient 5, Spinal CSF-lymphatic malformation fistula in a 9-year-old female with a history of kaposiform lymphangiomatosis who had chronic headaches. Brain and spine MRI demonstrated severe brain sag, syrinx, and intraosseous spinal lymphatic malformations (not shown). *A-C*, Coronal images from a prone CT myelogram showed rapid egress of contrast from a right T10-T11 meningeal diverticulum to the paravertebral space (A, arrow). The thoracic duct also opacified to the level of the innominate vein (B, C, arrows), resembling a lymphangiogram. In retrospect, there was a lymphatic malformation identified in the right T10-T11 paravertebral space on MRI (not shown).

Case 6

A 7-year-old female with Gorham Stout disease presented with postural headaches. During infancy, she had marked irritability and MRs of the brain and spine showed brain sag, cerebellar tonsillar descent, and a syrinx that was initially diagnosed as "Chiari 1 deformity" for which a suboccipital decompression was performed. The SIH Bern score was 6, and there was no dural enhancement. At age 7, spine MRI showed diffuse destructive intraosseous lymphatic malformations throughout the lumbar spine. A prone CT myelogram showed CSF leakage from the right L4 nerve root into the vertebral body and paraspinal soft tissue lymphatic malformations (Supplementary Fig 3). Targeted blood patches resulted in transient relief. Subsequently, targeted *n*-BCA liquid embolic was percutaneously injected via a spinal needle into the fistula, which resulted in marked improvement in symptoms. Over the following 15 years, the patient has intermittently suffered from both low- and high-pressure symptoms, which have been treated with intermittent blood patches, lumbar and ventricular peritoneal shunting, and medical therapy.

DISCUSSION

Spinal CSF leaks associated with adjacent vascular malformations are uncommon. In this clinical report, we demonstrated six patients with CSF leaks at CT myelography or cisternography directly caused by spinal or skull base vascular malformations.

Similar to routine CSF-venous fistulas, the pathogenesis of CSF-vascular malformation fistulas has not been well characterized, but invasion of either the dura and or nerve root sleeve by the malformation may result in the fistulous connection. This is not uncommon for vascular malformations in other parts of the body. For example, patient 4 developed chyluria from a microcystic lymphatic malformation invading the bladder wall. Similarly, patient 1 had an epidural and paraspinal venous malformation at the age of 18 months but did not manifest her SIH symptoms until two decades later when a fistulous connection between the thecal sac and the venous malformation was detected. Patient 2 also had an epidural venous malformation and developed a CSF-venous fistula years later. Further studies are needed to explain how these fistulas form; however, one clear observation is that the presence of an epidural or paraspinal vascular malformation is a potential risk factor for a CSF fistula.

Myelographic timing and density are complementary factors for detection of CSF-venous fistulas.[15] Similarly, timing may be important for CSF-venous malformation fistulas. In patient 1, the CSF-venous malformation fistula was not visible on the early myelographic phase immediately after contrast administration but was apparent on a delayed scan 3 minutes later. We suggest multiphase scanning may be helpful in this patient cohort. Analogously, venous malformations can often demonstrate delayed enhancement at MR imaging due to their venous nature.[16]

In all patients, there was a delay in the diagnosis of SIH. Five of six patients were incorrectly diagnosed with a "Chiari 1 deformity", 3 patients had a suboccipital decompression as a result, and 2 patients were being evaluated for a suboccipital decompression prior to the correct diagnosis being established. Confusion between Chiari 1 and SIH occurs frequently despite criteria differentiating these two entities,[17] particularly among pediatric patients, where SIH is less common than in adults. One major distinction between Chiari 1 deformity and SIH is the mamillopontine distance. In SIH, this distance is often reduced, while in Chiari 1, it is typically normal.[17] Four of our 6 patients had severe brain sag, which we characterized by an absent mamillopontine distance and could be due to the severity of the hypotension. The presence of a cervical cord syrinx was another imaging feature that also likely contributed to the incorrect diagnosis of Chiari deformity. SIH can rarely result in syrinx in patients without vascular malformations,[18] but it is not fully understood why a syrinx was present in all patients in this cohort with vascular malformations. Perhaps, chronic leakage into a vascular malformation results in lower CSF pressure and greater descent of the cerebellar tonsils. In the pediatric patients, another theory could be due to reduced posterior fossa calvarial development, which is still developing in childhood and may lead to more cerebellar tonsillar descent due to the intracranial hypotension. This is the same rationale for why some studies prefer the term Chiari 1 deformity rather than malformation, as it is a postnatal developmental mismatch between the rates of growth of neural tissue and the osseous posterior fossa rather than a congenital finding.[19] Further studies are needed to explore these potential theories of why syrinxes are possibly more prevalent in patients with CSF leaks from vascular malformations, but regardless, it may be an indicator to search for a vascular malformation as a potential etiology for the SIH.

Dural enhancement was absent in all of our patients, which is typically the most prevalent brain MRI feature in SIH.[20] We speculate this imaging sign was absent in our patient cohort due to delayed diagnosis, as it has been reported that dural enhancement can resolve with time, even without treatment.[21]

Classifying vascular anomalies can be challenging due to historical terms. For example, vascular malformations are often misclassified as hemangiomas, a vascular tumor. In one study that evaluated published articles with the term "hemangioma" in 2009, >70% were misclassified.[22] In our case series, 2 patients had their venous malformation misclassified as a hemangioma. The ISSVA classification of vascular anomalies is subdivided into vascular tumors and malformations. Under the vascular malformation category, there are various types, but venous and lymphatic malformations are most common. This nomenclature challenge has also been observed in patients with spinal and skull base leaks associated with vascular malformations, which makes it difficult to surmise the exact incidence since correct terminology is not always utilized. Moreover, in patients with vascular anomaly syndromes, this can be more challenging given the wide variety and the greater diagnostic complexity. We suggest that patients with CSF leaks from an underlying vascular malformation be referred to a vascular anomalies clinic for care of the vascular malformation, in addition to the CSF leak treatment, if possible, as certain vascular malformations may be amenable to medical therapies, such as, mTOR inhibitors.

In 5 of the 6 patients, there was a direct fistula between the CSF and the vascular malformation, while one patient had a CSF-venous fistula that is typical for other SIH patients without vascular malformations. Prior descriptions of these fistulas have also been labeled CSF-venous fistulas. While this is technically true, as there is a connection between the CSF and the venous system in patients with a venous malformation, we prefer the more technically correct term of CSF-venous malformation fistula. And if there is a lymphatic connection, then CSF-lymphatic malformation fistula. This more accurately characterizes the physiology and leads to better tracking for research purposes.

Two of the six patients had skull base leaks, specifically, in the petrous apex and temporal bone. Skull base leaks do not typically result in SIH, as the spinal level in which the CSF pressure changes from negative to positive relative to the atmospheric pressure is typically in the upper cervical spine. CSF leaks occurring above this zero-pressure and hydrostatic indifference point, therefore, do not typically result in a CSF leak when the patient is upright.[23] In one study of 273 patients with SIH by Schievink et al, there were no cases of skull base leaks.[24] Subsequently, however, this same group reported a single case of a large posterior fossa CSF leak resulting in SIH.[25] In a more recent study, 31 patients with skull base leaks had a mean Bern score of 0.9, and the single patient that had dural enhancement had an infratentorial CSF leak.[26] We postulate that if a skull base leak is severe that it could possibly overcome the pressure gradient between the brain and spine and result in SIH. Further research is needed to validate this theory.

There are various treatment options for CSF-vascular malformation fistulas. In 3 cases, the fistula was targeted with blood or fibrin glue patches with some relief. In our one patient with a more typical CSF-venous fistula, fibrin glue patching was curative. In Patient 5 with a CSF-lymphatic malformation fistula, surgery was successfully performed. Surgical treatment is not without risks, however, as one reported case of surgery for a spinal CSF-venous malformation fistula was complicated by

extensive blood loss.[2] In certain cases, intravenous onyx embolization may be an option if there is a venous pathway to the fistula. Further studies are needed to determine treatment efficacy for CSF-vascular malformation fistulas.

CONCLUSIONS

CSF-vascular malformation fistulas are a rare cause of SIH. It has been proposed previously that a CSF leak should be suspected in patients with epidural venous malformations—either syndromic or isolated—if they present with clinical symptoms of SIH.[3] We agree with this recommendation but further suggest that any person presenting with headache or other SIH symptoms in whom an underlying vascular malformation syndrome or an isolated paraspinal or skull base vascular malformation is present, that a CT myelogram and/or cisternogram be performed to identify a possible CSF leak into that entity.

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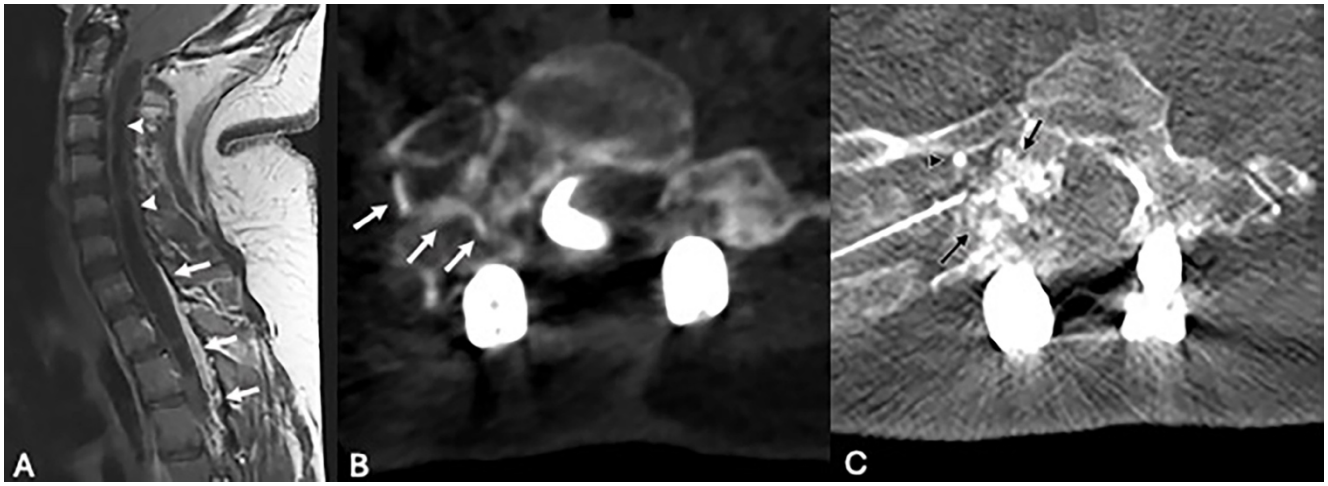
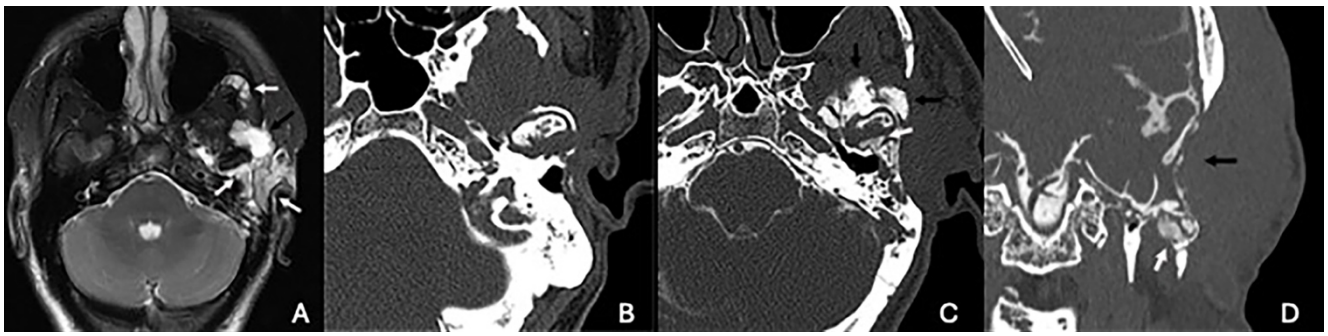
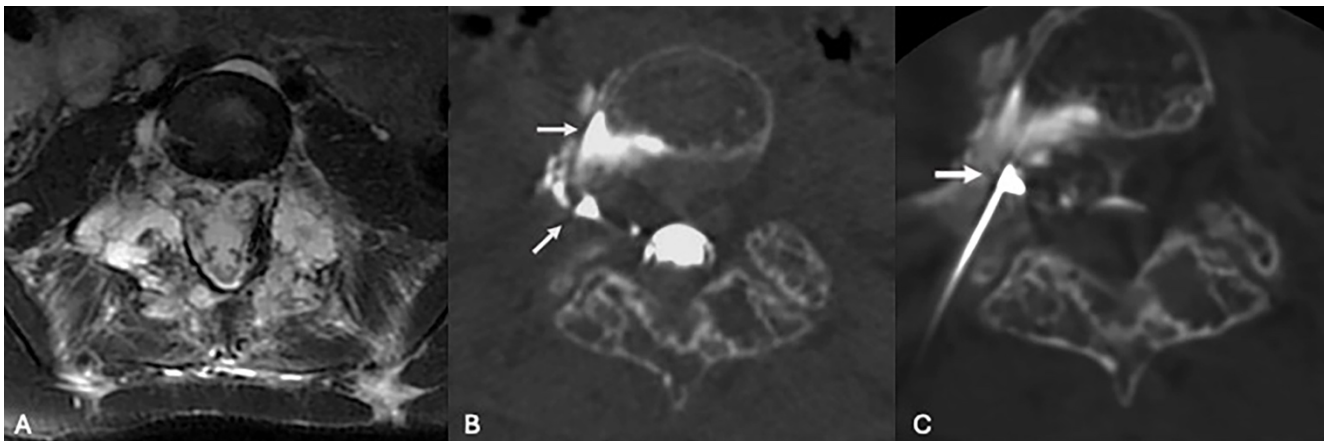
SUPPLEMENTAL FILES



SUPPLEMENTARY FIG 1. Patient 2: Spinal CSF-venous malformation fistula in a 40-year-old male who had a history of an upper thoracic epidural venous malformation that was initially thought to represent a hemangioma. The patient had a suboccipital and upper thoracic decompression for a presumed Chiari 1 deformity and hemangioma, respectively. *A*, Sagittal contrast-enhanced T1-weighted image of the cervical and thoracic spine 11 years prior shows a dorsal enhancing lesion in the upper thoracic spine (arrows). This did not represent epidural fat on additional fat-suppressed images (not shown). A syrinx is noted (arrowheads) as well as cerebellar tonsillar descent. *B*, Axial right-decubitus CT myelogram several years later shows a right T1-T2 CSF-venous fistula (arrows). Posterior spinal fusion is noted. *C*, CT-guided fibrin glue patch shows injectate within the right neural foramen and epidural space (arrows) and intravascular glue within the fistula (arrowhead). The CSF-venous fistula resolved on a post-treatment CT myelogram (not shown).



SUPPLEMENTARY FIG 2. Patient 3: Skull base CSF-venous malformation fistula in a 33-year-old male who had a large scalp, facial, and neck venous malformation who presented with headaches and upper and lower extremity numbness. Brain and spine MRI showed severe brain sag and a syrinx (not shown). *A*, Axial T2-weighted fat suppressed image shows extensive venous malformations in the left temple, infratemporal fossa, and retromandibular fossa (white arrows), along with a cephalocele (black arrow). *B,* Axial noncontrast CT shows no opacification in the retromandibular fossa. Erosion of the mandibular condyle is noted from the venous malformation. *C, D* Axial and coronal images from a prone CT cisternogram show abnormal enhancement in the left retromandibular fossa (white arrows) corresponding to the venous malformation that is suggestive of a CSF-venous malformation fistula. Opacification of the cephalocele within the infratemporal fossa is also seen (C, black arrows). Extensive osseous erosion of the left temporal calvarium secondary to the venous malformation is noted. (D, black arrow).



SUPPLEMENTARY FIG 3. Patient 6, Spinal CSF-lymphatic malformation fistula in a 7-year-old female with a history of Gorham Stout disease who had chronic headaches. Brain and spine MRI demonstrated brain sag and syrinx (not shown). *A*, Axial T2-weighted fat-suppressed image shows extensive lymphatic malformations in the bones and paraspinal soft tissues at the L4-L5 level. *B*, Axial prone CT myelogram shows extensive contrast leakage into the right L4-L5 neural foramen, paraspinal soft tissues, and vertebral body (arrows). C, Axial prone CT-guided treatment image shows injected *n*-BCA liquid embolic (arrow) within the fistula, which is on a background of contrast leakage from the preceding myelogram.