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Case Report

Spontaneous intracranial hypotension in a patient with systemic lupus erythematosus

Dan Isaac Cohen-Addad, MD, Aslan Efendizade, DO, Arkadij Grigorian, MD, Kevin Hewitt, DO, Vinodkumar Velayudhan, DO*

Department of Radiology, SUNY Downstate/Kings County Hospital, 451 Clarkson Ave, 3rd Fl, Rm B-3304, Brooklyn, NY 11203, USA

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ABSTRACT

Spontaneous intracranial hypotension (SIH) associated with cerebrospinal fluid leak classically presents with postural headache. It is most commonly caused by the spontaneous dehiscence of a meningeal diverticulum or as a consequence of dural tears. The association between connective tissue disease and SIH is well known. However, the occurrence of SIH associated with systemic lupus erythematosus has rarely been reported. We present a 53 years old female with a history of systemic lupus erythematosus who was diagnosed with SIH. The patient was worked up with Magnetic resonance imaging and Computed tomographic myelography, and successfully treated with a nontargeted epidural blood patch. Furthermore, we review the current literature and focus on the various imaging techniques that can be used in the workup of a cerebrospinal fluid leak.

Introduction

Intracranial hypotension is a cerebrospinal fluid (CSF) pressure of less than 7 mm H_2O, and classically presents as a postural headache (relieved by lying flat) [1]. This reduced pressure is due to a leak of CSF somewhere along the neuroaxis. Spontaneous intracranial hypotension (SIH) is most commonly caused by the spontaneous dehiscence of a meningeal diverticulum or as a consequence of dural tears secondary to degenerative changes of the vertebral column. The association between connective tissue disease and SIH is well known. However, the occurrence of SIH associated with systemic lupus erythematosus (SLE) has rarely been reported. We present a case of SIH in a patient with SLE.

Case report

A 53-year-old female with a history of SLE, antiphospholipid syndrome, protein C and S deficiency, and recurrent deep vein thrombosis presented to the hospital with a several month

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* Corresponding author.
E-mail address: dancodoc@gmail.com (D.I. Cohen-Addad).

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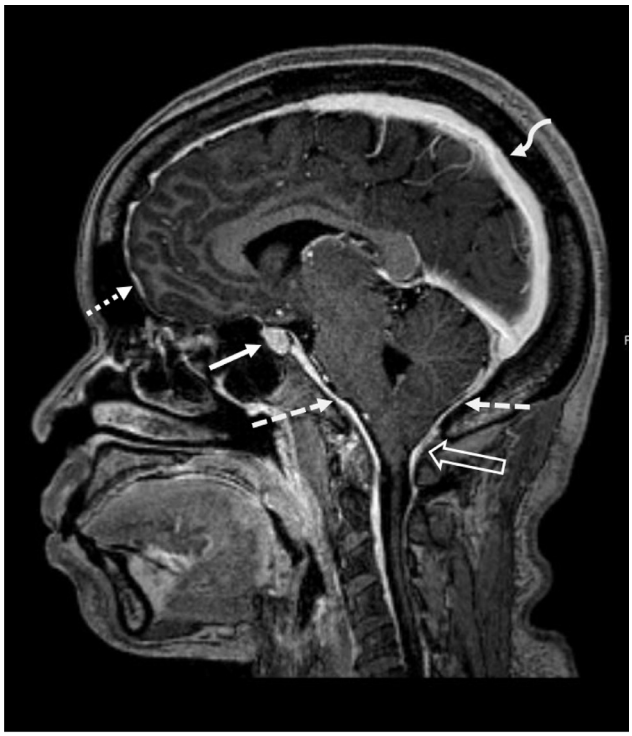

Fig. 1 – T1 postcontrast sagittal MRI in a 53 year old female, PMH of SLE, antiphospholipid syndrome, protein C and S deficiency, and recurrent deep vein thrombosis presented with a several month history of headaches showing low lying cerebellar tonsils and suprasellar structures (empty arrow). Diffuse smooth dural enhancement (dotted arrow). Enlargement of the pituitary gland (filled arrow), engorgement of the sagittal sinuses (curved arrow). Engorged clivus venous plexus, cervical epidural veins, and occipital sinus (dashed arrows).

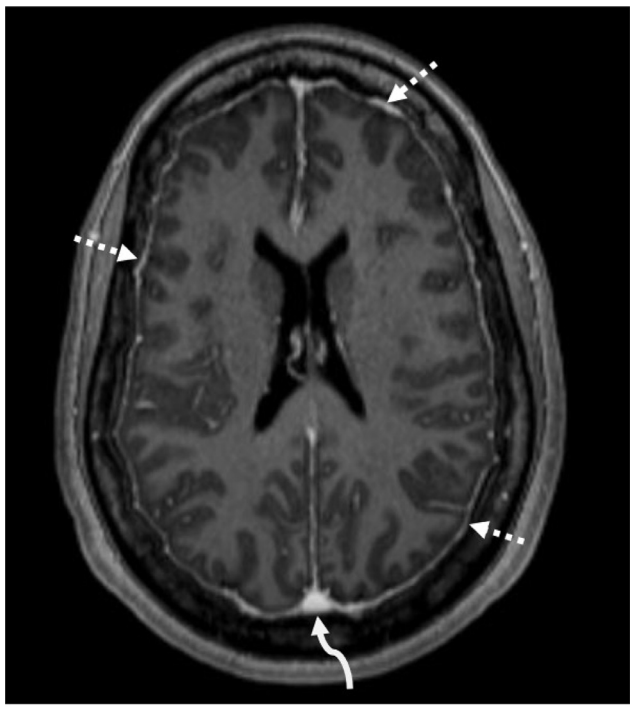

Fig 2 – T1 Postcontrast axial MRI demonstrating diffuse sooth dural enhancement (dotted arrow). Rounding of the dural sinus consistent with engorgement of the sagittal sinus (curved arrow). The constellation of the findings is suggestive of intracranial hypotension.

history of headaches. Associated symptoms included vertigo, as well as facial and hand paresthesia.

Magnetic resonance imaging (MRI) of her cervical spine revealed minimal degenerative findings. Contrast-enhanced MRI of the brain revealed diffuse pachymeningeal enhancement, with low lying cerebellar tonsils and suprasellar structures (Figs. 1 and 2). This raised suspicion for intracranial hypotension. Magnetic resonance angiography and venography did not reveal arterial stenosis or venous thrombosis. Computed tomographic (CT) cisternography excluded the skull base as a possible site of a CSF leak. Finally, a CT myelogram demonstrated a CSF leak along the thoracic spine with contrast pooling around the thecal sac from T3 to T10 (Fig. 3). The exact site of the leak was difficult to ascertain secondary to contrast pooling.

The patient underwent intervention with an epidural blood patch to seal the CSF leak (Fig. 4). Postoperatively, her symptoms resolved. For optimal management of SLE, azathioprine, and meloxicam were added to her medication regimen.

Upon 1-month follow-up, her brain MRI (Fig. 5) revealed resolution of cerebellar tonsillar ectopia, greater patency of the subarachnoid space and minimal pachymeningeal enhancement. The inner ear structures and cranial nerves were unremarkable.

Discussion

Normal CSF opening pressure in adults ranges from 70 to 200 mm H_2O [2]. Intracranial hypotension is a condition where there is a CSF pressure of less than 7 mm H_2O. It classically presents with headache in the upright position that improves upon lying flat. Patients may also exhibit nausea, vomiting, neck pain, vertigo, facial paresthesia, upper extremity radiculopathy, and even galactorrhea [3]. This phenomenon can broadly be categorized as primary (spontaneous) and secondary (iatrogenic or post-traumatic), with CSF leak as the common pathophysiologic substrate.

The Monro–Kellie hypothesis, which assumes the skull to be a rigid compartment, states that the sum of the intracranial volumes of blood, brain, and CSF are constant. Thus, a loss in the volume of 1 component may lead to compensation by another. In the case of a CSF leak, there is a decrease in the volume of CSF which leads to a compensatory dilatation of the vascular spaces (initially venous due to its higher compliance) [4], and increased flow to these vessels. Vessels within the dura mater lack a blood-brain barrier, such that the increase flow of blood to this region may also be accompanied by effusion into the dural spaces [5,6]. These processes account for the appearance of diffuse dural enhancement

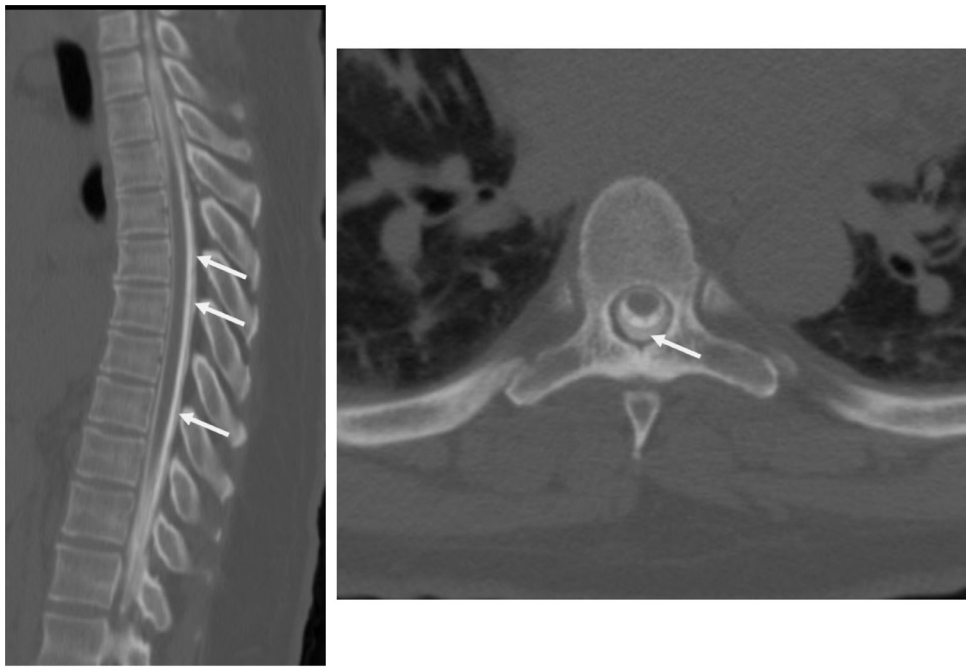

Fig 3 – CT myelogram sagittal (left) and axial (right) images demonstrating pooling of contrast around the thecal sac from approximately T3 level throughout T10 level, consistent with CSF leak.

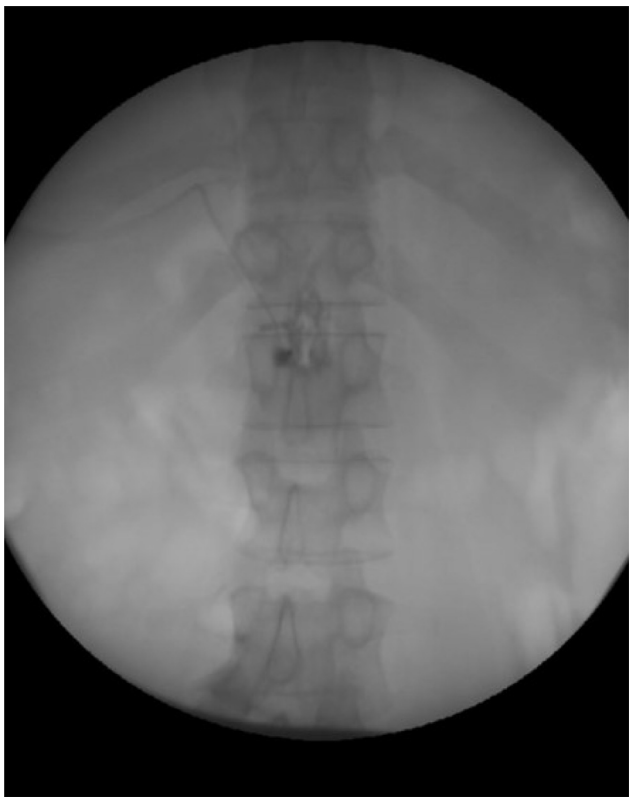

Fig 4 – Fluoroscopy image showing needle placement at T12-L1 intervertebral disc space level prior to epidural blood patch procedure.

seen on contrast imaging, along with subdural hematomas and hygromas in SIH. Another region of the brain which lacks a blood-brain barrier and displays compensatory hyperemia in the event of a CSF leak is the pituitary gland [7]. Decreased CSF and alterations in intracranial fluid dynamics leads to additional imaging findings including cerebellar tonsil ectopia, decreased size of the basilar cisterns, and downward displacement of the optic chiasm and brainstem structures. Further, sagging of the brain causes tension on subdural bridging veins, which are prone to rupture and forming hematomas. Spinal MRI may show enhancement patterns similar to those seen in the brain, with dilatation of the anterior epidural venous plexus and subdural collections [8]. The diagnosis is confirmed with a lumbar puncture revealing low opening pressure.

SIH is most commonly due to a CSF leak in the spine. The common causes of a CSF leak are dural dehiscence in the setting of a meningeal diverticulum, tears secondary to degenerative changes, and rare developmental anomalies. The spinal meninges are most susceptible around the spinal nerve roots, where the arachnoid layer may protrude through the dura, forming a diverticulum, which is susceptible to tear. The exact cause of the tear is unclear, but it may be associated with minor trauma causing stretching of the nerve root sleeves [9]. Alternatively, degenerative changes (osteophytes, disc protrusion) may cause tears in the thecal sac [10–12], most commonly along the ventral aspect of the low cervical and high thoracic spine, often associated with high flow leaks [13,14]. Particularly, thoracic disc protrusions are more likely to calcify, and therefore more likely to cause a meningeal tear [15,16]. In the setting of genetic connective tissue disorders, such as Ehlers-Danlos and Marfan syndrome [9], patients are speculated to be at greater risk of CSF leaks than in the general

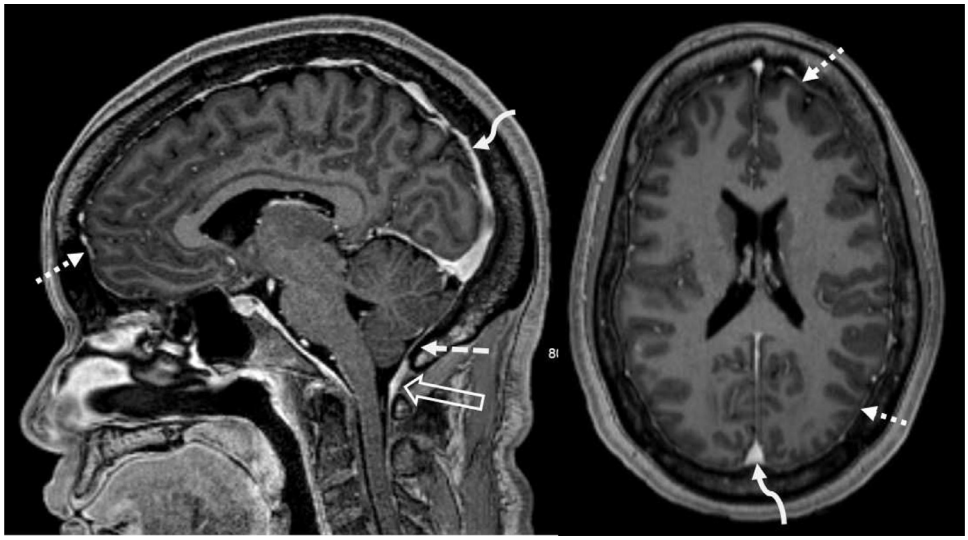

Fig 5 – T1 Post contrast sagittal (left) and axial (right) images showing resolution of tonsillar cerebellar ectopia and greater patency of the subarachnoid spaces (empty arrow). Very mild residual pachymeningeal enhancement (dotted arrow). Decreased sinuses engorgement (curved arrow) and residual occipital sinus (dashed arrows). Findings are improved compared to previous MRI consistent with resolution of intracranial hypotension.

population. Interestingly, there are also reports of CSF leaking into the peritoneum and pleural space [17]. A CT myelogram for our patient showed no evidence of other potential etiologies for a CSF leak such as meningeal diverticula, pronounced degenerative changes, or developmental anomalies. Furthermore, the patient had no documented history of connective tissue disorders.

We describe a case of SIH in a patient with a known history of SLE. To our knowledge, only 4 such cases have been previously reported [18–21]. The central nervous system manifestations of SLE are broad. For example, there are cases of both intracranial *hypotension*, as well as intractable headaches associated with intracranial *hypertension* [22]. SLE-induced intracranial hypertension is believed to be secondary to arachnoid granulation occlusion by inflammatory cells or by immune complex deposition within the choroid plexus [23]. Conversely, a potential causal relationship between SIH and SLE may be inflammation-induced weakening and perforation of the dura.

Identifying a CSF leak can be challenging, especially in the setting of SIH. The leak can occur anywhere along the neuroaxis. Furthermore, the leak can be slow, intermediate, or high-flow. The first step in diagnosis is to confirm the presence of intracranial hypotension with the use of contrast-enhanced brain MRI. Subsequently, various imaging modalities can be used in the detection and localization of a spinal CSF leak. It should be noted that pooling of contrast is common at the C1-C2 vertebral body level and can be misinterpreted as positive for a leak [24–26]. Furthermore, slow leaks require techniques with high spatial and contrast resolution, with different timing secondary to variability.

Intracranial hypotension has different treatment options based on the cause. Conservative management options include bedrest, and oral or intravenous caffeine. Both caffeine and theophylline act by opposing adenosine receptor activity, which results in arterial vasoconstriction. It is believed that the combination of hydration and adenosine receptor blockade will result in overall decrease in intracranial blood collection and venous engorgement, with simultaneous intracranial fluid volume expansion. In the case of a CSF leak, and failed conservative management, the next step would be a nontargeted epidural blood patch [27]. If this also fails, then localization is vital for a targeted blood patch.

Conclusion

We presented a case of SIH in a patient with SLE. The patient's symptoms resolved after a nontargeted epidural blood patch and improved medical management of her SLE. We also reviewed the imaging features of the entity, as well as the role of various imaging modalities in the work-up. Few cases of SLE-associated SIH have been reported in the past. As more cases are compiled, this raises the question of a causal relationship between the 2 entities. The mechanism of SIH development in these patients remains unclear, but SLE-related intracranial inflammation may weaken and perforate dura mater at the time of SLE exacerbation, and strict medical management may reduce the occurrence of such perforating lesions.

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