

## **Intracranial Hypotension in the Setting of Concurrent Perineural Cyst Rupture and Subarachnoid Hemorrhage**

Case Report

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## ABSTRACT

Although most patients with intracranial hypotension typically present with headaches, the rest of the clinical spectrum is characteristically non-specific and often quite variable. In a patient with concurrent pathologies that can produce a similar clinical picture, a high index of suspicion must be maintained to garner the correct diagnosis. The authors report a case of intracranial hypotension in the setting of concurrent perineural cyst rupture and subarachnoid hemorrhage. A 63-year-old woman with a family history of ruptured intracranial aneurysms presented after a sudden thunderclap headache and was found to have diffuse subarachnoid hemorrhage. Imaging revealed anterior communicating and superior hypophyseal artery aneurysms. Following the uneventful clipping of both aneurysms, the patient experienced a delayed return to her neurological baseline. After it was noted that the patient had an improved neurological examination when she was placed supine, further workup confirmed intracranial hypotension from perineural cyst rupture. The patient improved and returned to her neurological baseline after undergoing a high-volume blood patch and remained neurologically intact at postoperative follow-up. Although intracranial hypotension is known to be commonly associated with cerebrospinal fluid leak, its causal and temporal relationship with subarachnoid hemorrhage has yet to be elucidated.

**KEY WORDS:** aneurysm; coma; epidural blood patch; intracranial hypotension; perineural cyst; subarachnoid hemorrhage; Tarlov cyst

**RUNNING TITLE:** Intracranial hypotension with perineural cyst rupture and subarachnoid hemorrhage

## INTRODUCTION

The confounding nature of the symptoms of intracranial hypotension, along with the infrequency with which it occurs, may make the condition a diagnostic quandary for neurosurgeons. Although most patients typically present with headaches, the rest of the clinical spectrum is characteristically non-specific and often quite variable. This can lead to a significant delay in diagnosis, resulting in increased morbidity and possible mortality [1]. In a patient with concurrent pathologies that can produce a similar clinical picture, a high index of suspicion must be maintained to garner the correct diagnosis. We present the case of a 63-year-old woman who developed intracranial hypotension after subarachnoid hemorrhage (SAH) in whom diagnosis of a large perineural cyst in the upper thoracic spine was delayed. In this report, the authors aim to explore the relationship between the two entities.

## CASE PRESENTATION

*History and Presentation.* This 63-year-old woman with a family history of ruptured intracranial aneurysms presented to our institution after the sudden onset of a severe “thunderclap” headache. On arrival, the patient complained of headache and neck stiffness with intermittent nausea and vomiting. She was somnolent but easily arousable and otherwise neurologically intact (Glasgow Coma Scale score of 14). A computerized tomography (CT) scan of the brain obtained upon admission demonstrated diffuse SAH with blood in the sylvian fissures bilaterally as well as in the basal cisterns (Fig. 1). CT angiography of the brain confirmed the presence of both an anterior communicating artery aneurysm and a right superior hypophyseal artery saccular aneurysm (Fig. 2). The patient’s medical history included mucocutaneous systemic lupus

erythematosus (mSLE) that was controlled with hydroxychloroquine, but she had no history of hypertension or tobacco use. All initial laboratory values were within normal parameters.

*Operation.* On admission to the intensive care unit, the patient was promptly placed on our institutional SAH protocol, including strict blood pressure control, hydration, vasospasm monitoring, and seizure prophylaxis. Preparation for surgery was delayed because of difficulty acquiring blood products for possible transfusion because mSLE caused significant agglutination during the blood type and cross matching. Three days after her admission for SAH, the patient underwent clipping of both the right superior hypophyseal and the anterior communicating artery aneurysms with placement of an external ventricular drain.

*Postoperative Course:* Postoperatively, despite an uneventful surgery with unchanged somatosensory evoked potential and motor evoked potential monitoring, there was a delay in the patient's return to her preoperative level of consciousness. She also exhibited left-sided extensor posturing to painful stimulation. Treatment with hypertonic saline and dexamethasone resulted in a gradual improvement in her neurological status, and she was extubated on postoperative day (POD) 5.

The patient continued to exhibit a waxing and waning neurological examination for several days. On POD 16, a marked improvement in her alertness level was noted primarily when she was lying supine, raising the possibility that spontaneous intracranial hypotension was causing the patient's fluctuating neurologic status. Subsequent magnetic resonance imaging of the lumbar and thoracic spine demonstrated evidence of multiple small perineural cysts at all levels, with the most prominent on the right side at T10-T11 (Fig. 3). Scout imaging also

demonstrated inferior descent of the cerebellar tonsils by approximately 1.3 cm, further supporting the diagnosis of intracranial hypotension.

After she underwent a high-volume blood patch at T11-T12, the patient exhibited significant symptomatic improvement and returned to her neurologic baseline. At 3-month follow-up, the patient was noted to be neurologically intact with stable imaging.

## DISCUSSION

Intracranial hypotension is a rare clinical entity that remains a diagnostic dilemma, which is therefore often overlooked and treated suboptimally. The spectrum of symptoms experienced by patients is broad, including both overtly acute, rapid progression to stupor and coma and insidious, clinically silent presentation with subtle histories. Headaches are the most common symptoms patients experience, but they vary significantly, ranging from orthostatic in the majority of patients to chronic nonpositional in others [2]. They are typically resistant to treatment with conventional analgesics and are relieved when the patient is in the supine position [3,4]. Concurrent symptoms usually include nausea, vomiting, and neck pain and are generally considered to be relatively mild by the patient [5]. Atypical presentations, however, may progress to Parkinsonism, frontotemporal dementia, syringomyelia, hypopituitarism, seizures, coma, and even death [3,4,6-8].

Whether spontaneous or post-traumatic in nature, most cases of intracranial hypotension are believed to result from persistent cerebrospinal fluid (CSF) leakage [1]. First described in 1938 by Isadore M. Tarlov, perineural cysts are well-described causes of CSF leak and subsequent intracranial hypotension [9-12]. In this case report, we present an interesting case of

intracranial hypotension resulting from a perineural cyst rupture-induced CSF leak in the setting of an aneurysmal SAH.

Whether the SAH caused the perineural cyst rupture and subsequent intracranial hypotension in our patient is unclear. SAH does cause transient changes in intracranial pressure (ICP) throughout the course of the disease process [13]. The introduction of blood into the subarachnoid space creates immediate outflow resistance. ICPs continue to fluctuate until the blood is eventually absorbed. Whereas ICP changes are well studied on a clinical level, there may be other consequences of pressure differentials that may have delayed effects and consequences along the craniospinal axis. We speculate that the pressure differential created by the significant amount of SAH in our patient would have been large enough to cause a leak from an already compromised area of dura mater within a large perineural cyst.

## CONCLUSIONS

The myriad of symptoms that may occur in a patient with intracranial hypotension can make this condition difficult to diagnose, requiring the neurosurgeon to maintain a large index of suspicion. Although it is known to be commonly associated with CSF leak, its causal and temporal relationship with SAH has yet to be elucidated.

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**FIGURES**

Figure 1. Noncontrasted head computed tomography scans obtained on arrival illustrating diffuse subarachnoid hemorrhage within the bilateral sylvian fissures (a and b) and basal cisterns (c).



Figure 2. Computed tomography angiogram of the head with contrast enhancement illustrating an anterior communicating artery aneurysm (purple arrow) in the axial (a) and sagittal (b) views and a superior hypophyseal artery aneurysm (gold arrow) in the axial (c) and sagittal (d) views.

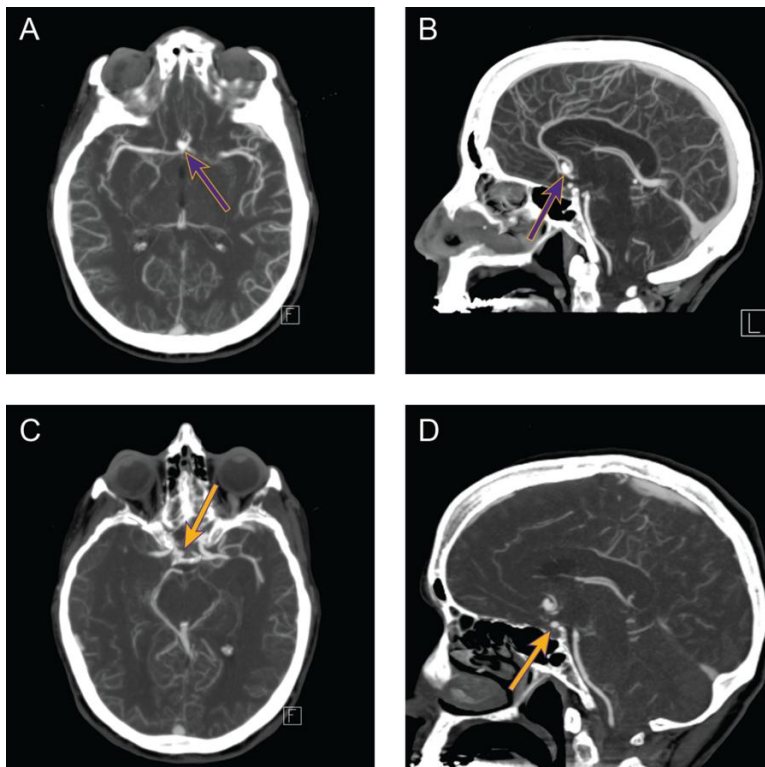


Figure 3. Sagittal (a) and axial (b) T2-weighted magnetic resonance imaging without contrast enhancement showing a right 2.4×2.4×2.1-cm cyst (gold line). Coronal (c) T2-weighted half-Fourier acquisition single-shot turbo spin-echo (HASTE) myelogram magnetic resonance imaging illustrating the large, right perineural cyst (purple arrow).

