Neurological Cases Vol1 No2 DECEMBER 2014
ISSN 2374-3522 (PRINT) ISSN 2374-3530 (ONLINE) http://www.researchpub.org/journal/nc/nc.html

The Eagle Sign (of Radiologic Diagnosis) Aids in the Management of Spontaneous Intracranial Hypotension: A Case Report

Joseph C. Wildenberg, *MD, PhD* [1,*], Seetharam C. Chadalavada, *MD, MS* [1], Scott Akers, *MD, PhD* [2], Pratap M. Yagnik, *MD* [3], and Paolo G. Nucifora, *MD, PhD* [2]

***Abstract*—Spontaneous intracranial hypotension can occur without preceding trauma or medical intervention. Patients often present with positional headaches but may be misdiagnosed with migraines/tension headaches due to a lack of clinical suspicion. Neuroimaging is likely to be the first step in diagnosis and is highly accurate. We present a 31-year old man with spontaneous intracranial hypotension who was admitted with positional headaches and concerning findings on a head CT. Additional imaging revealed classic findings of intracranial hypotension including separation of the superior cerebral veins from the sagittal sinus. A thoracic spine MRI showed an epidural fluid collection suggestive of a CSF leak. The patient was treated with an epidural blood patch and his symptoms improved.**

***Keywords* — Intracranial hypotension, CSF, Neuroimaging**

I. INTRODUCTION

Intracranial hypotension (IH) results from a decrease in cerebral spinal fluid (CSF) pressure, often through loss of CSF from the central nervous system[1,2,3]. This disorder is most commonly encountered following a lumbar puncture, though spontaneous loss through a CSF leak without trauma or recent medical intervention is also possible. When IH occurs spontaneously, without clues in the patient's history to guide clinicians, initial diagnosis is difficult, and delayed or misdiagnosis is common.

Cross-sectional neuroimaging with MRI generally provides diagnostic accuracy, although CT is usually the initial study obtained[1,3]. In this report, we present a case of IH in a young man who came to the emergency department with new positional headaches without any history of trauma or recent medical procedure.

II. CASE REPORT

A 31-year old man presented to the emergency department with new headaches that started approximately one month prior. He denied any trauma and had no other significant past medical history. The headaches were more severe when standing or sitting upright and improved when he lay down. He also noted that consumption of caffeine temporarily reduced the headache severity. He initially experienced some associated "ear popping" and neck pain but those symptoms resolved several days after the headaches began.

On examination the patient was well appearing. He did not have any neurological deficits on physical exam and was not excessively sensitive to light or loud noise. A non-contrast head CT did not show any focal lesions, though prominent venous sinuses raised concern for a venous sinus thrombosis (Fig. 1). A subsequent contrast-enhanced MRI and MR venogram of the head revealed diffuse enhancement of the meninges, sagging of the cerebellar tonsils, and engorgement of the cerebral venous vasculature (Fig. 2). No prior imaging was available therefore the chronicity of the sagging cerebellar tonsils could not be determined. In addition, the superior cerebral veins were separated from the superior sagittal sinus with expansion of the subdural space.

Without a clinical history to explain these findings, a diagnosis of spontaneous IH was made. MRI of the thoracic spine was then performed in an attempt to localize the presumed CSF leak. While no explicit leak was seen, there was a large epidural fluid collection posterior to the mid-thoracic spinal cord. Although a lumbar puncture is commonly performed to confirm IH, the presence of this epidural fluid collection was deemed sufficient to make this diagnosis. The patient was then taken to the interventional suite for a 10 mL spinal epidural blood patch placed at L3-4. He experienced some immediate symptomatic relief providing both diagnostic confirmation as well as treatment. After a few days his headaches recurred, again worse on standing and relieved by lying down. He received another epidural blood patch with immediate relief of his pain. Telephone follow-up after three weeks confirmed that the patient had near-complete sustained relief of headaches.

1. Housestaff, Department of Radiology, Hospital of the University of Pennsylvania, Pennsylvania, PA, USA. 2. Attending physician, Department of Radiology, Philadelphia VA Medical Center, Philadelphia, PA, USA. 3. Attending physician, Department of Neurology, Philadelphia VA Medical Center, Philadelphia, PA, USA.

*Correspondence to Dr. Joseph Wildenberg (e-mail: joseph.wildenberg@uphs.upenn.edu).

ISSN 2374-3522 (PRINT) ISSN 2374-3530 (ONLINE) http://www.researchpub.org/journal/nc/nc.html

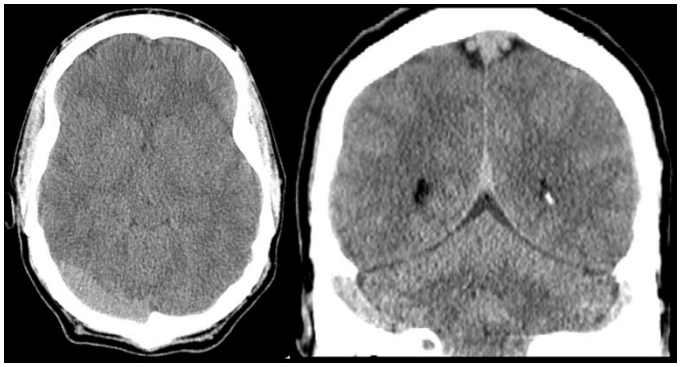

Fig. 1. Axial and coronal unenhanced CT images of the head. There is no global mass effect or midline shift. The coronal image demonstrates engorgement and separation of the superior cerebral veins from the superior sagittal sinus. The right transverse sinus is also engorged.

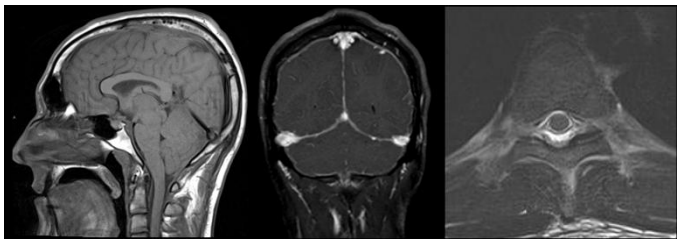

Fig. 2. Pre-contrast T1 sagittal (left) and post-contrast fat-saturated T1 coronal (middle) MR imaging of the brain show sagging of the cerebellar tonsils and engorgement/separation of the superior cerebral veins from the superior sagittal sinus. Axial T2 imaging of the thoracic spine demonstrates a posterior crescentic epidural fluid collection suggesting the presence of a spinal CSF leak.

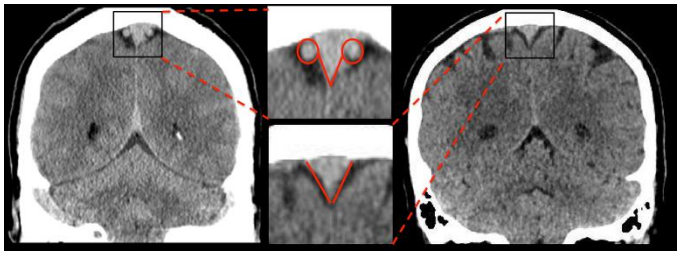

Fig. 3. Coronal CT images of the patient (left) and a healthy control (right) demonstrate the appearance of engorgement and separation of the superior cerebral veins from the superior sagittal sinus in intracranial hypotension. The magnified views and overlay highlights and outlines the Eagle sign.

III. DISCUSSION

Orthostatic headaches are a well-recognized symptom of IH following lumbar puncture. In the absence of relevant clinical history, however, positional headaches suggestive of spontaneous IH are more likely to be initially diagnosed as migraines or tension headaches. It has been estimated that spontaneous IH may have an incidence of 5/100,000 per year, with a peak age near 40 and affecting women more often than men[1]. Some connective tissue disorders such as Marfan's and type-II Ehlers-Danlos syndromes confer a higher risk for spontaneous IH[4].

Specific diagnostic criteria for spontaneous IH have been proposed, but in the absence of a definite extradural CSF collection, requires a lumbar puncture with a low opening pressure[5]. The findings presented in this case highlight the ability for cross-sectional neuroimaging to diagnose spontaneous IH without invasive diagnostic procedures. Studies have suggested that findings on MRI of the brain can have a sensitivity of 70-95% and specificity above 90%[5,6]. The most common findings can be remembered with the mnemonic SEEPS: Subdural fluid collections, Enhancement of the pachymeninges, Engorgement of venous structures, Pituitary hyperemia, and Sagging of the brain[1,7,8]. In addition to these findings, the patient presented here showed expansion of the subdural space around the superior sagittal sinus with separation of the superior cerebral veins from the sinus, a finding we have termed the "Eagle Sign" (Fig. 3).

Lumbar puncture continues to be the reference standard for the diagnosis of IH. The invasive nature of this procedure, and the potential to temporarily worsen the patient's symptoms, have led to an increased use of neuroimaging as the first diagnostic step. As with this case, neuroimaging findings highly suggestive of IH may obviate the need for a lumbar puncture.

Many cases of spontaneous IH resolve without treatment, contributing to difficulty with estimation of incidence. Conservative treatment consisting of rest, vigorous oral hydration, and caffeine intake can be effective if given time[1,2,3]. With severe symptoms, however, the mainstay of treatment remains the administration of 10-20 mL of autologous blood into the spinal epidural space, called an epidural blood patch[9]. Although only effective in approximately one-third of cases, this procedure can be repeated as necessary with increasing patch volumes[10]. Recurrence is possible though this seems relatively uncommon[1]. Focal leak as seen on MRI appears to be associated with a better prognosis for symptomatic recovery than diffuse leak[11].

Spontaneous intracranial hypotension is an uncommon but not rare cause of positional headaches and clinicians should keep this etiology in mind when patients present without a clinical history of trauma or recent medical intervention. Brain imaging with MRI or CT has a high diagnostic accuracy and the Eagle Sign, engorgement of the superior sagittal sinus and superior cerebral veins with expansion of the subdural space, is a readily seen finding in these patients.

REFERENCES

[1] Schievink WI. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. *JAMA*. 2006;295:2286.
[2] Mokri B. Spontaneous low pressure, low CSF volume headaches: spontaneous CSF leaks. *Headache*. 2013;53(7):1034-53.
[3] Rahman M, Bidari SS, Ouisling RG, and Friedman WA. Spontaneous intracranial hypotension: dilemmas in diagnosis. *Neurosurgery*. 2011;69(1):4-14.
[4] Schievink WI, Gordon OK, and Tourje J. Connective tissue disorders with spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension: a prospective study. *Neurosurgery*. 2004;54:65-70.

[5] Schievink WI, Maya MM, Louy C, Moser FG, and Tourje, J. Diagnostic Criteria for Spontansous Spinal CSF Leaks and Intracranial Hypotension. *AJNR*. 2008;29:853-856.
[6] Farb RI, Forghani R, Lee SK, Mikulis DJ, and Agid R. The venous distension sign: a diagnostic sign of intracranial hypotension at MR imaging of the brain. *AJNR*. 2007;8:1489-93
[7] Fishman RA, and Dillon WP. Dural enhancement and cerebral displacement secondary to intracranial hypotension. *Neurology*. 1993;43:609-611.
[8] Mokri B, and Atkinson JLD. False pituitary tumor in CSF leaks. *Neurology*. 2000;55:573-575.
[9] Sencakova D, Mokri B, and McClelland RL. The efficacy of epidural blood patch in spontaneous CSF leaks. *Neurology*. 2001;57:1921-23.
[10] Griauzde J, Gemmete JJ, Chaudhary N, Wilson TJ, and Pandey AS. Large-volume blood patch to multiple sites in the epidural space through a single-catheter access site for treatment of spontaneous intracranial hypotension. *AJNR*. 2014:Ahead of print.
[11] Schievink WI, Maya MM, Louy C. Cranial MRI predicts outcome of spontaneous intracranial hypotension. *Neurology*. 2005;64:1282-84.

Questions** (please choose a single answer)**:

1. The most common cause of intracranial hypotension in the outpatient setting is:

   A. Trauma
   B. Spontaneous
   C. Post lumbar puncture
   D. Infection

2. All of the following are MRI findings suggestive of intracranial hypotension EXCEPT:

   A. Pachymeningeal enhancement
   B. Uncal herniation
   C. The Eagle Sign – separation of the superior cerebral veins from the superior sagittal sinus
   D. Pituitary hyperemia

3. Which of the following is NOT a conservative treatment for intracranial hypotension:

   A. Fluid resuscitation
   B. Caffeine
   C. Rest
   D. Diuretics

4. If an initial blood patch is unsuccessful at relieving symptoms of intracranial hypotension, the next step is:

   A. Repeat blood patch
   B. Intrathecal saline injection
   C. Exploratory surgery
   D. Trendelenburg positioning

Answers: 1. C; 2. B; 3. D; 4. A.