Transcranial Doppler Ultrasonography Identifies Symptomatic Cavum Septum Pellucidum Cyst: Case Report

Abstract

Objective: Cavum Septum Pellucidum (CSP) cysts are considered normal anatomic variants, comprising as many as 15% of the adult and 85% of pediatric populations. On rare occasions, the cavum can obstruct CSF outflow from the lateral ventricles causing elevated intracranial pressure (ICP) and headaches. The purpose of this paper is to present a challenging case of new onset symptomatic CSP in a previously healthy adult male without papilledema and elevated ICP detected by transcranial Doppler (TCD) ultrasonography.

Clinical Presentation: A previously healthy 44 year-old man presented to the neurology service with debilitating positional headaches that were mitigated solely by recumbent positioning. A magnetic resonance imaging scan (MRI) of the brain revealed a cavum septum pellucidum. A lumbar puncture was performed and revealed normal ICP. No papilledema was evident on fundoscopic examination. A CSF flow study revealed normal dye opacification pattern without evidence of CSF leak.

Intervention: Without other clinical indicators of high ICP, but a history suspicious for symptomatic CSP, TCD study was performed and revealed abnormally low cerebral blood flow velocities (CBFV's) and significantly elevated pulsatility indices (PI's) for patient's age indicative of high ICP. Endoscopic fenestration of the septum pellucidum was performed improving the patient's headaches and normalization of the PI's and CBFV's to normal (p<0.01).

Conclusions: Symptomatic CSP is a difficult diagnosis to make based on existing diagnostic paradigm. TCD in the absence of other objective confirmatory studies, can aid in the diagnosis and provide information about the success of fenestration of the cavum septum.

Introduction:

Cavum Septum Pellucidum (CSP) cysts are normal anatomic variants present in 10-15% of adults. On rare occasions, these cysts can become symptomatic, obstructing cerebrospinal fluid (CSF) outflow via a ball-valve mechanism. In general, patients present with symptoms suspicious for high intracranial pressure (ICP), (headache, usually postural, papilledema) and/or behavioral changes in the setting of a radiographically confirmed septum pellucidum. Several reports have indicated that in appropriately selected patients, endoscopic or open fenestration of both septa of the septum pellucidum can relieve symptoms.

The following case describes a patient who presented with signs and symptoms of severe headaches that improved with recumbence in the setting of a CSP. Despite all evidence confirming an intracranial low-pressure state, transcranial Doppler ultrasonography (TCD) confirmed high pressure within the supratentorial space. The patient’s symptoms improved dramatically following endoscopic fenestration concomitant with the improvement in the TCD variables pulsatility indices (PI's) and cerebral blood flow velocities (CBFV's).

Case Report

An otherwise healthy 44 year-old-man with chronic, low intensity headaches presented following significant worsening of his
headaches. These headaches were intolerable while the patient was upright, and improved when he lay flat. He had no history of head trauma. He had tried multiple medications (Non steroidal anti-inflammatory drugs, steroids, narcotics) without significant relief of pain. A fundoscopic examination revealed sharp disc margins without evidence of papilledema. A magnetic resonance imaging (MRI) of the brain (Figure 1a and 1b) revealed a CSP cyst and enlarged ventricles. A lumbar puncture was performed revealing an opening pressure of 15 cmH₂O with no headache improvement subsequent to drainage of 20 cc CSF. A CSF flow study was performed revealing a normal flow pattern of CSF without evidence of leak.

At this point, with all evidence supporting low pressure headaches in the setting of a known CSP cyst, TCD study of all major intracranial vessels was performed revealing abnormally low CBFV's and significantly elevated PI's. Based on this finding, a compartmentalized elevated supratentorial pressures contributing to low CBFV and normal infratentorial and spinal CSF pressures was presumed. An endoscopic fenestration of the septum pellucidum was performed without any complication, and the patient noticed immediate relief of headache while in upright position. Follow-up TCD analysis performed 1 day post-operatively revealed normalization of the CBFV's and PI's. A paired-sample t-test was conducted to determine whether the

Figure 1: (a) T2 axial MRI through and (b) T2 coronal FLAIR MRI of the lateral ventricles obtained prior to endoscopic fenestration. Note the dilated cavum septum pellucidum (single head black arrow) with the anterior portion of the septum (two head white and black arrows) bulging to cover the foramina of Monroe (single head white arrow).

Figure 2: (a) Boxplot of Pulsatility Indices for right anterior circulation on pre and postoperative measurement and (b) Boxplot of Pulsatility Indices for left anterior circulation pre and postoperative measurement.
mean PI's were significantly different from initial examination to follow-up exam. The results indicated that the mean value for the initial PI for the right anterior circulation (M = 1.163, SD = .17) was significantly greater than for the mean for the follow-up PI for the right anterior circulation (M = .98, SD = .15), t(5) = 4.052, p = .01) (Fig. 2a). The results also indicated that the mean for the initial PI for the left anterior circulation (M = 1.32, SD = .34) was significantly greater than for the mean for the follow-up PI for the left anterior circulation (M = .83, SD = .12), t(5) = 4.247, p = .008) (Fig. 2b). In addition, the results indicated that the mean CBFV’s for the right anterior circulation (M=44.43, SD=13.09) and left anterior circulation ((M=44.57, SD=12.49) were significantly greater post operatively than the mean CBFV’s pre-operative state for the right (M=31.57, SD=3.81), t(5) = 5.82, p < .001) and left anterior (M=26.29, SD=9.12), t(6) = 5.88, p < 0.001) circulations

Though the patient has residual headache, his debilitating headache has not returned in the 9 months since his surgery. His postoperative MRI's (Figure 3a and 3b) reveal that the CSP is no longer deviated or obstructing CSF flow from the lateral ventricles.

Discussion

The occurrence of CSP cysts in both adults (10-15%) and children (up to 85%) is not rare. It is, however, rare for these cysts to become symptomatic. Prior to 1996, only 18 reported cases appeared in the literature. Since that time, the majority of reports have focused on minimally invasive treatment modalities. Signs and symptoms include those associated with elevated ICP (papilledema, positional headache worse when lying flat) and/or memory dysfunction secondary to pressure on the fornices. In this case, the patient presented with signs and symptoms of low-pressure headaches in the setting of an identified CSP cyst on MRI. All initial testing indicated that there was a normal CSF flow pattern in the setting of normal ICP. However, the TCD study that indicated that, the pressure in the lumbar cistern might not have been indicative of the supratentorial pressure.

The primary purpose of TCD ultrasonography is to determine the velocity of flowing blood by quantitative interpretation of arterial pressure waveforms. Although the qualitative contour of the TCD waveform during ICP elevation falls into a recognizable pattern, their interpretation depends on the experience and expertise of TCD examiner/interpreter. Objective, reproducible and verifiable measures of TCD waveform changes are necessary for TCD findings to be used with certainty for evaluation of intracranial hypertension. One method of quantifying these changes is utilization of the PI. The PI is the reflection of downstream resistance. PI takes into account the peak systolic CBFV (pCBFV) and the end-diastolic CBFV (edCBFV) and compared the changes in these variables against the change in the standard measure of entire waveform, such as mean CBFV. Changes in arterial pulsatility, especially during intracranial hypertension, will affect both pCBFV and edCBFV, which are easily identified in TCD waveform, and are reflected by the equation PI = pCBFV - edCBFV / meanCBFV.

This case highlights several points pertaining to the use of TCD and physiology of CSF compartments. First, though the central nervous system compartments are interconnected (supratentorial, infratentorial, spinal, etc.), there may be subtle pressure differences that are not communicated well throughout the closed system. Second, TCD can be used to evaluate ICP either independent of or in conjunction with other invasive and non-invasive imaging studies. In one study performed at our institution, TCD's were obtained on all severely head injured patients' from Operation Iraqi Freedom. Though the primary goal of that study was to determine the rate of vascular injury, a secondary, unpublished component included the use of PI's as an independent indicator of increased ICP.
Conclusion

This case represents a novel use of TCD to confirm the clinical suspicion of a symptomatic CSP cyst in the setting of several confounding clinical variables. TCD examination, if performed by an experienced ultrasonographer, can be used to detect subtle alterations in ICP.

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References: