Management of Raised Intracranial Pressure—When Three is Better than Two

Dear Editor,

We had a 31-year-old lady who presented with a 2 months history of headache, neck pain, episodic dizziness, diplopia, and transient visual obscurations associated with multiple emetic episodes.

She had long standing obesity and had been diagnosed with polycystic ovarian disease (PCOD) for which she was taking cyproterone acetate and metformin for a year prior to this presentation. About 2.5 months before this presentation, she had had H1N1 influenza with respiratory failure for which she was intubated and mechanically ventilated. Over the next week, her respiratory functions improved and she was extubated and discharged.

At this presentation, she was conscious and alert and was obese. Her blood pressure, heart rate, and respiratory rate were within normal limits. Bilateral papilledema was noted on fundoscopy and constriction of visual fields on perimetry [Figure 1]. The neurological examination was unremarkable except for hyporeflexia of the deep tendon jerks.

She was emergently evaluated for the cause of raised intracranial pressure (ICP). Magnetic resonance imaging (MRI) of the brain did not reveal any parenchymal or extra parenchymal mass lesions, hydrocephalus, loss of flow voids to suggest cerebral venous sinus thrombosis (CVST), or abnormal flow voids suggesting a dural arteriovenous fistula. There were tell-tale signs of raised ICP—a partially empty sella and flattening of the sclera at the optic nerve head [Figure 2]. Cyproterone was stopped at admission.

A lumbar puncture was done to document the opening pressure and look for chronic meningitis. According to modified Dandy’s criteria, ruling out competing diagnosis is one of the criteria to establish a diagnosis of idiopathic intracranial hypertension (IIH), which is a diagnosis of exclusion, and was also a possibility, keeping her obesity in view.[1,2] Similarly, any patient with a raised ICP with normal CSF cellularity and biochemistry should be evaluated for the possibility of CVST, but her MR venogram (MRV) was normal [Figure 3]. On Lumbar puncture (LP), a clear free-flowing CSF with an opening pressure of 13 cm H₂O was obtained. The CSF was acellular, cytopsin was negative for malignant cells, and CSF proteins and sugar were in the normal range. CSF was negative for Xpert Mycobacterium tuberculosis/Rifampin gene (MTB/Rif), veneral diseases research laboratory (VDRL), and cryptococcal antigen. Strong suspicion of IIH warranted a repeat lumbar puncture a week later, which revealed a CSF opening pressure of 28 cm H₂O.

Her hemogram, liver and renal function tests, echocardiography, and urine examination were unremarkable. ACE levels were within normal ranges and Human immunodeficiency virus (HIV) serology, Hepatitis B surface antigen (HbsAg), and (HCV) Hepatitis C virus were negative.

Our patient’s clinical picture and one of the CSF opening pressure results raised suspicion for IIH, for which Acetazolamide was started. Since her visual field charting had revealed a significantly restricted field, relieving the CSF pressure by inserting a ventriculoperitoneal shunt was being considered. However, in lieu of contradicting opening pressure findings, CSF pressure was continuously monitored by inserting an epidural catheter in the subarachnoid space and connecting it to the CSF pressure monitor, which revealed a CSF pressure between 10 and 15 cm H₂O throughout the evaluation period of 3 days. Since the CSF pressure was not elevated on continuous monitoring, acetazolamide was stopped soon after starting the monitoring, and the pressure remained in the same range even after stopping the drug. She subsequently began to improve clinically, and on follow-up at 28 days, her symptoms were relieved, papilledema resolved, and perimetry showed improved visual fields bilaterally [Figure 3].

Patients with headaches are evaluated by direct ophthalmoscopy, but drusen may be mistaken for papilledema in a patient who otherwise may have a primary headache such as migraine. Examination of bilateral papilledema by an expert ophthalmologist should be considered especially when CSF opening pressure is normal. Bilateral papilledema is a specific marker of raised ICP, particularly in younger patients.[3]

Increased ICP is a common cause of headaches and vision loss. Presenting features like headache, transient visual blurring, and papilledema should prompt suspicion toward a raised ICP.
and emergently evaluated. Raised ICP can lead to rapid vision loss, and early intervention in such cases can save the vision. If ventriculoperitoneal or lumbar theco-peritoneal shunting is delayed due to any reason, in such cases, large volume CSF removal can buy time.

In our index case, the etiology of raised ICP had several differentials. After excluding malignant hypertension, neuroimaging (preferably MRI brain and MRV) is indicated to identify any evident space-occupying mass or CVST.\[4-6\]

Her history of severe influenza infection and intake of cyproterone acetate led to a high clinical suspicion of CVST as the cause of the raised ICP. An MR venography was performed, which was unremarkable. In this clinical situation, occluded sinuses on venography have a high positive predictive value. However, a normal venogram does not rule out a recanalized thrombus, as the imaging was done 2.5 months after the initial event. In absence of evidence of an obvious cause for raised ICP, the diagnosis of IIH could be considered, particularly so in an obese female of reproductive age group.

PCOD has a proven association with both IIH and Cerebral venous sinus thrombosis (CSVT), particularly in obese women of the reproductive age group. Usage of estrogen-based oral contraceptives, which may be combined with cyproterone acetate, may not only contribute as a risk factor for developing venous thrombo-embolic phenomenon but has also been associated with IIH.\[9\] Recent history of a viral influenza infection fitting with the temporal origin of the symptoms raises suspicion of cytokine-mediated thrombotic complication.\[10\]

Initially, we thought that there was an error in determining the opening pressure in the first lumbar puncture—with papilledema and constricted visual fields, we expected the opening pressure to be high. However, contrary to our hypothesis, we found the pressure to be in the normal range. So we repeated the second time, when it was elevated. Since the findings were contradictory, we decided to proceed with a continuous ICP monitoring.

Given the improvement in the patient’s clinical profile and non-deterioration, we concluded that the first reading was perhaps correct and the second elevated reading was a false reading, which was confirmed by the continuous CSF pressure monitoring. Normal opening pressure on a lumbar tap, in the context of raised ICP, can be due to incorrect positioning or obstruction in the needle. On the other hand, straining, coughing, and pressing of the legs against the abdomen can lead to spuriously high CSF opening pressure. Opening pressure may also spuriously fall in cases of acute dehydration or low blood pressure. Therefore, a single observation of any reading of CSF pressure, not concordant with clinical findings, warrants a repeat lumbar puncture or continuous monitoring before subjecting the patient to any invasive surgical procedure.

Figure 1: Upper panels show the fundi with papilledema and the lower panels show the visual field charting with perimetry and the dark points represent the loss of vision and thus the lower panels show constricted visual fields.

Figure 2: Magnetic resonance imaging of the brain showing normal parenchyma with no mass lesions, no hydrocephalus, and no features to suggest venous sinus thrombosis or abnormal flow voids.

Figure 3: Upper panels show the fundi with resolved papilledema (normal fundus) and the lower panels show perimetry in which the constricted visual fields shown in Figure 1 have recovered.
In our case, the most likely etiology of raised ICP was a CVST, which had spontaneously recanalized and the CSF pressure thus came to normal range. The papilledema took time to recover as the arachnoid granulations were probably blocked and took time to recanalize.

When a patient is seen in real-time with headache, papilledema, and constricted field of vision, the clinician faces the threat of losing time and thus a possibility of permanent vision loss in the patient. When a raised CSF pressure is found in such a patient with impending vision loss and since a raised ICP is an identifiable proximate cause for vision loss, it is tempting to rapidly perform the intervention such as CSF shunting to prevent further damage. However, in some cases, the underlying etiology may be self-limiting. In some cases, the patient may be in the resolving phase of the illness and may improve spontaneously in the natural course of the illness. In a given case, it may not be possible for the clinician to know at that time point if the patient is in the ascending phase or resolving phase of vision loss given a single constricted field of vision. So, the first reading of CSF opening pressure signaled us that it is possible that the patient could be having resolving papilledema. The patient did not have further worsening after admission and her visual acuity was 6/6 throughout the hospital stay. This prompted us to do continuous CSF pressure monitoring and thus avoid an invasive surgical procedure.

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**Conflicts of interest**

There are no conflicts of interest.

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