Communicating hydrocephalus due to cerebral venous sinus thrombosis treated with ventriculoperitoneal shunt

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Abstract

Cerebral venous sinus thrombosis (CVT) is a rare cerebrovascular disease with variable presentation. CVT rarely causes hydrocephalus. Communicating hydrocephalus due to CVT is extremely rare. We describe a patient of CVT presenting with chronic headache and communicating hydrocephalus. The patient was successfully treated with ventriculoperitoneal (VP) shunt. A 40 year old man presented with moderate to severe headache since six months and progressive visual loss since two months. Head Computed tomogram showed mild hydrocephalus without obstruction. Lumbar puncture (LP) demonstrated elevated pressure but was otherwise normal. Magnetic resonance venogram showed extensive CVT. Repeated CSF drainage and thecoperitoneal shunt did not relieve the severe headache hence a VP shunt was placed. Post shunt headache subsided with resolution of hydrocephalus. CVT can present as communicating hydrocephalus. Gradual reduction of intra-ventricular pressure by repeated LPs followed by VP shunt can safely treat hydrocephalus due to CVT.

Key Words

Cerebral venous sinus thrombosis, hydrocephalus, ventriculoperitoneal shunt

Case Report

A 40-year-old gentleman presented with history of insidious onset headache for 6 months and diminution of vision for 2 months. The headache was holocranial, continuous and of mild to moderate intensity. It was not associated with nausea, vomiting, photo or phonophobia. Since 2 months, he noticed progressive diminution of vision in both eyes. He did not have seizures, altered sensorium or any focal deficit. There was no history of fever. He was previously healthy and was not on any medications. On examination, blood pressure was 120/80 mmHg, general and systemic examination was normal. On neurologic examination, there were no signs of meningism. Our patient presented with chronic headache and diminished vision. Our patient is unique as he was successfully treated with VP shunt without any complication, unlike the previously reported case.

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Magnetic resonance venogram (MRV) showed near-complete thrombosis of the right transverse sinus and partial thrombosis of the left transverse and the sigmoid sinus [Figure 2]. Cerebrospinal fluid (CSF) manometry demonstrated opening pressure of 30 cm of CSF in the lateral decubitus position. Twenty of CSF was drained and the pressure reduced to 15 cm. The CSF did not show any cells, while proteins were 5 mg/dL and sugar was 99 mg/dL (blood sugar of 130 mg/dL). India ink preparation and cryptococcal antigen were negative. Post CSF drainage, the headache promptly decreased in intensity. Two days later, the headache recurred. CSF manometry was repeated, where the opening pressure was 27 cm. CSF was drained and pressure was reduced to 15 cm of H2O. There was relief of headache for the next 2 days. Two days later, the headache recurred and CSF manometry (pressure of 30 cm) with drainage of CSF was repeated for the third time, which resulted in prompt relief of headache. In view of persistently elevated CSF pressure and presentation similar to IIH, a thecoperitoneal (TP) shunt was performed. The patient had relief of headache following TP shunt. Post TP shunt, he was started on low-molecular heparin followed by warfarin for venous sinus thrombosis. He did not have headache for the next 4 weeks but there was no improvement in vision. After 4 weeks, the headache recurred and was severe in intensity. CT scan of the brain did not show any significant change. In view of failure of TP shunt to relieve headache and persistence of hydrocephalus, a ventriculoperitoneal (VP) shunt was placed. Post VP shunt, there was relief of headache and reduction in ventricular size but no improvement in vision. Six months later, he is free of headache although there is no improvement in vision. He is on oral anticoagulation for chronic venous sinus thrombosis.

**Discussion**

Although the presentation of CVT is highly variable, communicating hydrocephalus is extremely rare in CVT. Hydrocephalus in CVT is usually obstructive, secondary to parenchymal lesions like cerebral edema or hemorrhagic infarction in the thalami and basal ganglia. These lesions cause obstruction of CSF outflow at the level of the foramen of Monro or third ventricle, leading to obstructive hydrocephalus.[3] CVT with hydrocephalus without parenchymal edema, infarction or hemorrhage, as in our case, is extremely rare and only one case is reported previously.[1] In CVT without parenchymal lesion, the obstruction to the CSF pathway is in the arachnoid granulation at the end of the CSF pathway. Hence, there is no pressure gradient between the subarachnoid spaces and the ventricles and the hydrocephalus does not develop.[7] In experimental superior sagittal sinus ligation in dogs in the acute stage, no ventricular enlargement occurred. However, in the chronic stage, nine dogs out of 12 dogs showed enlargement of the ventricles.[8] Similarly, in chronic venous sinus thrombosis, malabsorption of CSF through arachnoid villi and impaired venous circulation can lead to hydrocephalus. However, why this does not cause hydrocephalus in majority of cases of CVT is not clear.

Our patient presented with chronic headache and post-papilloedema optic atrophy without any features of a focal lesion or meningitis. A communicating hydrocephalus [Figure 1a] without a parenchymal lesion, meninginal enhancement or subarachnoid hemorrhage (SAH) was detected on CT scan. Lumbar puncture showed raised pressure and no evidence of infection or SAH. MRV [Figure 2] showed extensive venous sinus thrombosis.

The previously reported patient had presented with thunder clap headache, seizure and drowsiness.[9] This presentation was thought to be typical of SAH. The initial CT did not depict any diagnostic clues suggestive of sinus thrombosis. The CSF analysis did not reveal SAH or meningitis. Hence, hydrocephalus was treated in the usual way with external ventricular drainage (EVD). This resulted in intracerebral hemorrhage. A repeat CT showed appearance of a typical empty triangle sign. Subsequent MRV confirmed the tentative diagnosis of venous sinus thrombosis. The authors hypothesize that sudden reduction in intraventricular pressure due to EVD led to a critical increase of the transluminal pressure of intraparenchymal veins. Owing to the increase of the pressure gradient, these vessels ruptured and caused intracerebral hemorrhage.[9] In our patient, we suspected CVT as a possible cause of hydrocephalus. We performed lumbar puncture (LP)
with CSF drainage to lower the intracranial pressure. The gradual reduction of intraventricular pressure by repeated LPs was followed by a VP shunt. This prevented creation of a sudden pressure gradient between the intraparenchymal veins, venous sinuses and the ventricles that is responsible for secondary intracerebral hemorrhage.

**Conclusion**

CVT can present as communicating hydrocephalus without parenchymal lesion. Gradual reduction of intraventricular pressure by repeated LPs followed by VP shunt can safely treat hydrocephalus due to CVT.

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