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Case Report

Solving a complex, rare, and challenging scenario in a normal pressure hydrocephalus with recurrent shunt dysfunction and multiple revisions – The opposing role of evolving low-pressure hydrocephalus and idiopathic raised intra-abdominal pressure

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Received : 29 June 2022
Accepted : 26 October 2022
Published : 25 November 2022

DOI
10.25259/SNI_586_2022

Quick Response Code:

ABSTRACT

Background: Maintenance of pressure gradient within the intracranial chamber, shunt hardware, and the abdominal cavity plays a significant role in the optimal functioning of the ventriculo peritoneal shunt. We report a rare and a complex scenario in a patient with normal pressure hydrocephalus (NPH) who had recurrent and refractory ventricular peritoneal shunt dysfunction. Following a meticulous analysis, this was attributed to a very rare, and, first to be documented in the literature, a combination of an evolved very low pressure hydrocephalus (VLPH) system and asymptomatic raised intra-abdominal pressure (IAP).

Case Description: A 72-year-old male presented with NPH syndrome, associated with recurrent shunt dysfunction. A thorough systematic evaluation, which included intracranial pressure monitoring and IAP monitoring, revealed the presence of VLPH and a concomitant elevated IAP that was asymptomatic. This unique situation required changes in surgical strategy, which included correction of VLPH state, insertion of the anti-siphon device, and the placement of the distal end of the shunt into the pleural cavity. This led to solving the "mystery" of recurrent shunt dysfunction in this complex scenario.

Conclusion: It is imperative to perform the pressure analysis of the intracranial chamber, shunt hardware, and even the abdominal cavity, especially, in every case of refractory shunt revisions. Possibilities of a rare cause such as VLPH or an asymptomatic raised IAP acting alone or in combination must be considered. Only then, the final cerebrospinal fluid diversion strategy should be decided.

Keywords: Hydrocephalus, Intra abdominal pressure, Low pressure hydrocephalus, Shunt dysfunction, Ventriculo peritoneal shunt

INTRODUCTION

Hydrocephalus is an active distention of the ventricular system of the brain resulting from an inadequate passage of cerebrospinal fluid (CSF) from its point of production within the cerebral ventricles, to its point of absorption into the systemic circulation.[7]

Maintenance of pressure gradient within the intracranial chamber, shunt hardware, and abdominal cavity plays a significant role in optimal functioning of the ventricular peritoneal shunt. While the most frequent cause of shunt dysfunction is either related to altered pressure dynamics in the intracranial cavity or hardware dysfunction, minimal attention has been accorded to raise intra-abdominal pressure (IAP) as a causative factor. We report a rare and complex and unusual scenario in a patient presumed to have normal pressure hydrocephalus (NPH) with numerous failed shunt revisions, the cause of which was finally attributed to an extremely rare combination of an asymptomatic increased IAP and, a newly evolved very low pressure hydrocephalus (VLPH).

CASE HISTORY

A 72-year-old male presented with memory disturbances, imbalance, and urinary urgency for the past 4 months, indicating classical features of NPH. This was further confirmed by radiographic imaging [Figure 1], Pre and post lumbar puncture, gait assessment, and Mini Mental State Examination Score. Considering the improvement, a programmable vertriculoperitoneal shunt at set pressure of 110 mm of H2O was placed. Immediate post-operative, improvement in the gait was noted; however, CT brain [Figure 2], neither showed any evidence of reduction in the ventricular size, nor presence of features of shunt over drainage. After 2 months, patient presented in emergency in a debilitated condition. At this point, he was diagnosed with gross bilateral subdural hygroma caused by shunt over drainage at the valve pressure of 110 mmH_2O which was treated by increasing shunt pressure to 180 mmH_2O and surgical evacuation of hygroma [Figure 3].

A month later, he presented again with altered sensorium and ventriculomegaly [Figure 4]. Multiple attempts to readjust the pressure setting failed, no mechanical obstruction of shunt was noted. The condition was treated with shunt revision and laparoscopic repositioning of peritoneal end of shunt was attempted. At this moment, the possibilities of intermittently dysfunctional shunt, arrested hydrocephalus, onset of Parkinson's plus syndrome, encephalopathy, or metabolic disturbances were considered. CSF study and metabolic workup were done which showed no abnormalities. Electroencephalographs showed slowing in both hemispheres.

Considering a difficult case of hydrocephalus and recurrent shunt failure, it was imperative to assess every single compartmental pressure that directs the flow of CSF. To get a "true" value of the intracranial pressure (ICP) recordings and not allowing any effect of the existing valve settings, an external ventricular drainage (EVD) was placed in the contralateral left frontal horn, while the exteriorized tube

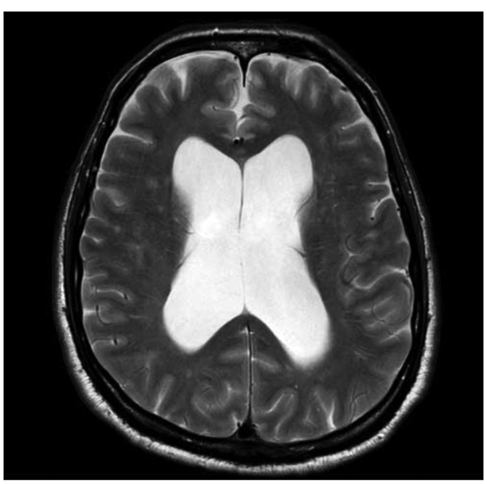

Figure 1: Preoperative MRI: Ventriculomegaly with minimal periventricular lucency.

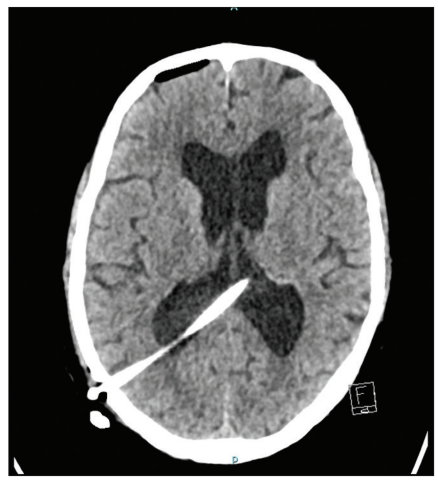

Figure 2: Immediate post ventriculopleural shunt: No change in ventricular size, dilated frontal horn, and temporal horn.

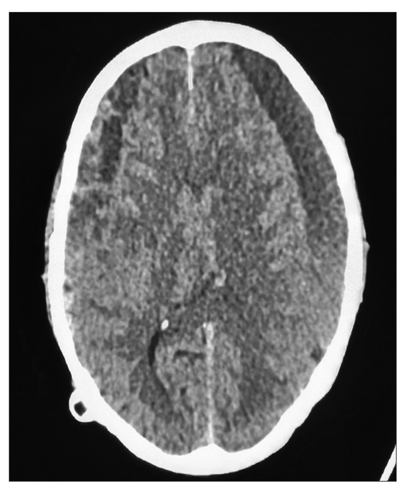

Figure 3: Post ventriculopleural shunt over drainage of the shunt with collapsed ventricles and bilateral subdural hygroma.

was kept clamped. ICP measured through EVD was mere 3 mmHg, whereas the IAP was 25 cm of H_2O (18 mm Hg). The evaluation was performed for the cause of high IAP and was inconclusive. This high IAP (18 mmHg) with a VLPH (<3 mmHg) contributed to loss of differential pressure resulting in recurrent shunt dysfunction.

This extraordinary situation was circumvented initially by correcting the VLPH by the sub-zero draining method of Pang *et al*.[6] This involved lowering the EVD bag sequentially (in 5 cm gradient) below the reference level of tragus, until, there was a clinical and radiological improvement. Moreover, then, again gradually, raising it up (5.0 cm/day), till the negative or low-pressure state is reversed back. Once the ICP values reached up to the 9.0 mmH_2O, with documented improvement in patients clinioradiological state [Figure 5], plan was made to adjust the existing valve to 80 mmH_2O, place an antisiphon device (to prevent position related overdrainage), and finally, replace the shunt to ventriculopleural shunt (−5 mmHg negative pressure) [Figure 6].

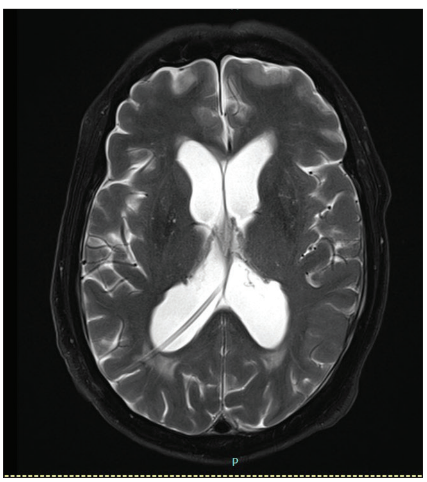

Figure 4: MRI brain: Pan ventriculomegaly with patent shunt *in situ*.

Significant improvement in the condition of the patient was noted within 1 week of surgery, patient was later discharged ambulating, with minimal assistance. Patient has been following up as outpatient for the past 2 years without any neurological deficits or shunt dysfunction.

DISCUSSION

We describe a rare and extraordinary case of a patient with LPHS and intra-abdominal hypertension. The combination of these two opposing pressure gradients resulted in recurrent shunt dysfunction.

Why does CSF flow within the shunt tube?

CSF flows from high pressure cavity to the low pressure cavity. The drainage of CSF in Ventriculoperitoneal (VP) shunt is regulated by ICP, hydrostatic pressure (HSP), and IAP. The tally of ICP with HSP needs to be more than IAP for effective drainage of CSF. This can be mathematically summed up as:

Draining pressure (+) = ICP (+) + HCP (+) (p fluid density × g gravity × h height) – IAP (+) (Eq. 1)[4]

When the draining pressure is positive, then there is forward flow of CSF within the shunt tube.

Situational analysis in our case

Role of ICP – State of low pressure/negative pressure hydrocephalus (NePH)

Pang *et al*.[6] proposed that brain parenchyma has viscoelastic properties which resist or permit distortion due to changes in brain turgor. Brain turgor increases with increase in extracellular fluid. Loss of brain turgor prevents reduction in ventricular size, producing a low pressure system.

To promote adequate drainage of CSF, it is essential to have the drainage system pressure to be lower than the ICP. Hence,

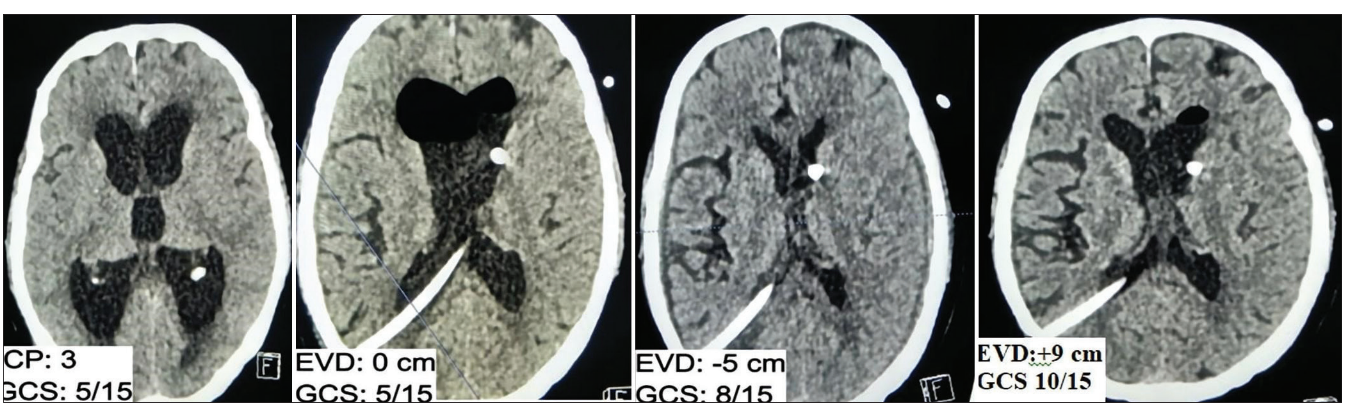

Figure 5: Progressive change of the height of external ventricular drainage tube in cm with increasing intracranial pressure. F(foot) in the image denotes direction towards which the image is being scrolled.

Figure 6: Post ventriculopleural shunt and chest X-ray showing the intrapleural. F(foot) in the image denotes direction towards which the image is being scrolled.

the treatment of NePH is to gradually increase the ICP by initiating the CSF drainage at pressure lower than ICP. This principle was followed in our case and the ICP was gradually increased to subnormal pressure.

Role of hydrostatic pressure

The hydrostatic pressure is directly dependent on the fluid density, gravity, and the height of the fluid column. Due to the position of the patient, the height of the fluid column can vary and therefore hydrostatic pressure can be variable in sitting and supine position.[2]

Role of IAP

Normal range of IAP is 0–7 mm Hg.[1] Certain and normal individuals especially those who have high body mass index can harbor asymptomatic raised IAP.[8] Sahuquillo studied IAP before insertion of the peritoneal end of the VP shunt through direct measurement of the IAP, and noted certain individuals especially those with high body mass index can harbor asymptomatic raised IAP. Mirzayan MJ *et al*, noted that with increase in BMI, there was an increase in intra abdominal pressure, leading to shunt dysfunction.[5]

IAP in standing and sitting pressure ranges from 16 to 20 mmHg.[3] IAP difference between supine and standing position is average 4–6 mmHg.[3] For efficient functioning of the shunt, it is essential to maintain the pressure difference between ICP and IAP of 8–10 mmHg in sitting position. For effective flow of CSF from the shunt, the ICP and HCP essentially need to be more than 20 mmHg. With increase in IAP, in most situations, a high HCP promotes drainage of the VP shunt. Therefore, it is very rare to have a scenario where the HCP is not able to overcome IAP. The theoretical effects of raised IAP on VP shunt function are well known. Shunt dysfunction has been noted due to raised IAP in patients with obesity, chronic constipation, chronic urinary retention, and bowel inflammation.[9]

What had happened in this case?

In our case, IAP could have been normal/high normal at the time of the index shunt placement and also at the time of over drainage. Combination of "normal" ICP and the HCP was able to move the CSF into the abdominal cavity. However, progressively, the patient became non-ambulatory and obtunded probably due to evolution of VLPH. At this moment, two opposing pressure gradients were acting simultaneously – a very low ICP and a high IAP with resultant malfunction as illustrated in Equation 2.

Draining pressure (−) = ICP (−) + HCP (+) (p fluid density × g gravity × h height) – IAP (−) (Eq. 2)

Strategy for shunt placement in this scenario

To change the direction of flow, ICP needed to be more positive, and a distal reservoir more negative. Therefore, the VLPH state was corrected as described above and made more positive, while the pleural cavity was chosen as the reservoir as it has a negative pressure[8] leading to a state of Equation 1 and an ideally functioning shunt. Since there was a history of over drainage at 120 mmH2O and now the setting was 80 mm H2O (Mean ICP value 90 mmH2O), a siphon guard was inserted to avoid posture related over drainage. This strategy definitively worked and proven by the 2 years of follow-up data with the patient with an improved and stable clinicoradiological state.

Were there options?

Endoscopic third ventriculostomy? (ETV)

ETV is a treatment for low pressure hydrocephalus (LPH) especially in patients who have undergone 3rd–4th ventricular tumor excision surgeries and intraventricular bleed. Due to the uncertainty over the results of ETV and concerns with the over drainage, we adopted a more "measurable" and a definitive procedure based on the strong hypotheses that were created.

Lumboperitoneal (LP) shunt?

LP shunt is a treatment for VLPH state. Considering a situation of LPH with high IAP approaching into peritoneum has high chances of failure. We had shunt *in situ* that was not infected, without blockage, and otherwise "working well." Having worked on building up strong hypotheses, and limited data on the efficacy of LP shunt, we were less inclined to adopt LP shunt.

CONCLUSION

VP shunt for hydrocephalus is known for multiple complications. LPH itself is arduous to diagnose. In addition, the presence of "occult" elevated IAP can be a nightmare for the surgeon and the patient. Measuring IAP becomes essential in complex failed cases of shunt surgery to prevent complications. This case highlights the importance to understand the physiology behind the working of a VP shunt and the mechanisms behind its dysfunction especially if there is no mechanical obstruction. Pre-operative monitoring of ICP and IAP becomes essential to avoid repeated surgical procedures and reduce hospital stay and financial strain especially in patients with history of multiple shunt revision procedures, abdominal surgeries, and obesity.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Doshi AB, Sarkar H, Shelke Y, Pothare A, Kulkarni A. Solving a complex, rare, and challenging scenario in a normal pressure hydrocephalus with recurrent shunt dysfunction and multiple revisions – The opposing role of evolving low-pressure hydrocephalus and idiopathic raised intra-abdominal pressure. Surg Neurol Int 2022;13:546.