Intraparenchymal pericatheter cyst as an indicator of ventriculoperitoneal shunt malfunction: A case-based update

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INTRODUCTION

Ventriculoperitoneal shunt (VPS) is the main surgical intervention for communicating or obstructive hydrocephalus and idiopathic normal-pressure hydrocephalus. It can also be performed in idiopathic intracranial hypertension (IIH) when there is no response to medical therapy or there is visual dysfunction worsening. Although standard in several conditions, the procedure shows a considerable rate of complications (~23.8% in adults).

Intraparenchymal pericatheter cyst (IPC) is a rare complication of VPS, occurring mainly in children. To date, only 35 cases have been reported in the literature, of which, 14 were in adults, and four were in IIH patients. The aim of this article is to present a case of IPC formation in an adult, as well as to review the literature on the subject.

ABSTRACT

Background: Intraparenchymal pericatheter cysts (IPCs) are a rare ventriculoperitoneal shunt (VPS) complication, with only a few cases recorded in the literature.

Case Description: We report a 22-year-old woman admitted with headache, papilledema, vision loss, and a history of leukemia. Lumbar puncture revealed idiopathic intracranial hypertension (IIH). Three months after VPS implantation, she was readmitted with headache and worsening of visual impairment. CT evidenced a IPC with perilesional edema. Intraoperatively, a shunt revision and cyst drainage were opted for. We present a discussion and literature review on this unique complication of VPS, with emphasis on management.

Conclusion: It is important to understand and consider IPCs as complications of VPS surgery, including in adult patients and IIH cases.

Keywords: Catheter obstruction, Cerebrospinal fluid edema, Idiopathic intracranial hypertension, Intraparenchymal pericatheter cyst, Ventriculoperitoneal shunt
CASE PRESENTATION

A 22-year-old woman with a history of leukemia presented with progressive headache, papilledema, and visual loss at admission. Magnetic resonance imaging (MRI) was normal [Figure 1]; however, lumbar puncture with cerebrospinal fluid (CSF) pressure measurement confirmed the diagnosis of IIH (pseudotumor cerebri), requiring treatment with VPS [Figures 1 and 2].

The patient was readmitted 3 months later due to the recurrence of headache symptoms and visual impairment. Computed tomography (CT) [Figure 2] showed a parenchymal lesion around the ventricular bypass catheter with surrounding edema, but without hydrocephalus (never present in this case). MRI [Figure 1] revealed a round-shaped lesion, with content isointense to CSF, and a thin nonenhancing wall. These findings were compatible with the diagnosis of IPC. Abdominal CT was benign, without evidence of distal causes of obstruction.

VPS revision was performed, and a proximal obstruction identified [Figure 3], then managed by catheter replacement and subsequent cyst drainage. Postoperative CT [Figure 2] revealed cyst regression and an intracystic hemorrhage. At 3-month follow-up, significant improvement in headache and visual impairment was observed, with a normal neurological examination and no surgical complications.

DISCUSSION AND LITERATURE REVIEW

VPS is a very common procedure, used mainly for treating hydrocephalus or idiopathic normal-pressure hydrocephalus. However, it is also an alternative for IIH, particularly in refractory cases or those involving visual deterioration. In the literature, hydrocephalus was the main cause for VPS surgery (31 of 35 published cases, ~91%). Despite being frequent, VPS has a significant rate of complications, commonly divided into foreign body related (e.g., migration, infection, fibrosis, and allergic reaction), abdominal (e.g., pseudocyst) and surgical complications (e.g., overdrainage), and those resulting in hydrocephalus (e.g., obstruction and disconnection).

IPCs, however, are very rare complications of VPS. Although they can also appear in Ommaya reservoirs, only VPS cases were considered in this review. Furthermore, only reports with adequate clinical and imaging descriptions were included in the study [Table 1].

IPC formation...
probably results from a weakening of ependymal cells, which, associated with VPS malfunction, cause increased CSF pressure and subsequent parenchymal collection. Then, depending on the degree of the pericatheter gliosis, a cyst can be formed, sometimes associated with vasogenic edema. Possibly, another factor is the more compliance of the white matter surrounding the catheter compared to the ependyma. The CSF diffuses through the catheter orifice created in the ependyma and accumulates in the white matter during the course of the occluded catheter. Moreover, those who have IIH without ventricular dilation may present less compliant ventricles. The intracranial pressure increases, but as there is a low resistance outlet (catheter orifice), the liquid exits there, without ventricular dilation. Therefore, the presence of ventricular dilatation may not be a precise or the sole factor to estimate the correct functioning of the shunt system. These mechanisms may explain why such cysts are more common in children, since their cerebral parenchyma and ventricles are softer and more elastic, respectively. Children account for 60% of the 35 reported cases.

Patients with this complication present with varied symptoms, the most frequent being headache (~40%), vomiting (~27%), and visual changes (~10%). There have also been reports of decreased levels of consciousness, hemiparesis, speech and gait disorders, tinnitus, and epileptic seizures. One patient was asymptomatic at re-evaluation, and the IPC was found by routine imaging tests. Interestingly, the diagnosis of this complication can occur in a variable time from shunt implant, with cyst onset being reported years, months, and even days after surgery.

Imaging tests are of utmost importance to make a differential diagnosis of parenchymal lesions in VPS patients as well as to identify concomitant conditions significant to the therapeutic approach. The CT examination can assist in the diagnosis of IPCs, and it is very often the initial imaging test. However, MRI is the most helpful examination as it allows the correct distinction between cysts and their differential diagnoses due to its better resolution. Alternative imaging techniques, such as CT cisternography and nuclear medicine scans, can be useful to recognize a cyst-ventricle communication or catheter obstruction, respectively.

CSF edema, tumors, abscesses, and dysembryogenetic lesions are all considered cyst-like lesions. Porencephalic cysts (or porencephaly) share the same clinicoradiological features as IPCs, although they can be considered a differential diagnosis and are divided into communicating or noncommunicating, depending on whether they are contiguous with the intraventricular and/or subarachnoid spaces. A cyst-ventricle communication was well described in two cases. Certain imaging conditions can present concomitantly to the pericatheter cyst, such as CSF edema and hydrocephalus. Dual cyst presentation is also possible.
Perilesional CSF edema was the only known accompanying condition in the presented case [Figures 1 and 2].

Early recognition and treatment of an IPC can provide complete clinical improvement and cyst regression. A shunt revision is the most common approach for IPCs (29 of 33, ~88%), typically in symptomatic cases with catheter obstruction. There were no instances of cyst recurrence. However, a new cyst was formed after a contralateral shunt placement and a pseudocyst identification after an apparent cyst regrowth. Both cases were managed with shunt revision (followed by a cystoperitoneal shunt placement in the first). There have only been four reported cases of IPCs after VPS surgery for IIH, all of them in adults, and a shunt revision was performed in three of them, resulting in near-complete cyst resolution. The age and management features were also similar to the presented case.

Some authors have questioned the necessity of shunt revision, especially in asymptomatic patients. Revision was not performed in four cases, three of them being managed conservatively with clinical and imaging surveillance and one being lost to follow-up. No patient who underwent conservative treatment showed symptoms of shunt malfunction. All remained asymptomatic and two evolved with cyst regression. Sinha et al. supported conservative treatment, especially in asymptomatic patients. This type of approach has the advantage of avoiding surgery, though an obstruction, even if partial, may lead to the necessity of shunt revision.

CONCLUSION

Although rare entities, it is important to understand and consider IPCs as complications of VPS surgery, including in adult patients and IIH cases. Shunt revision is the standard approach, though conservative treatment has been performed in specific conditions.

Declaration of the patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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