Cerebral parenchymal cyst: A rare complication of ventriculoperitoneal shunt malfunction in an adult

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We report a rare complication of ventriculoperitoneal (VP) shunt malfunction: an intraparenchymal pericatheter cerebrospinal fluid (CSF) cyst. To the best of our knowledge, this is the second reported case of VP-shunt-related parenchymal CSF cyst to be reported in an adult patient, and the longest reported delay in development of this complication after shunt placement. Mass effect and edema from CSF cysts on CT and even T2 FLAIR sequences suggest the diagnosis of cerebral abscess or tumor. Comprehensive MR imaging can exclude both, and lead to proper management. While this complication is rare, clinicians treating adult patients with shunts, even shunts placed decades prior, need to be aware of it.

Case report

The patient, a 20-year-old man, presented as a referral to the neurosurgical service with progressive symptoms of hemianopsia, headache, nausea, and vomiting with the presumptive diagnosis of cerebral abscess. At 5 years of age, the patient had had a right transfrontal VP shunt placed for communicating hydrocephalus secondary to meningitis.

Initial noncontrast enhanced CT of the head (Fig. 1) demonstrated homogeneous low density conforming to the right frontal white matter centered about the right frontal VP shunt. Mass effect effaced the right lateral ventricle and resulted in 8 mm of right-to-left subfalcine herniation with enlargement of the left lateral ventricle, suggestive of entrapment or shunt failure. Low density on CT, extensive mass effect, and the patient’s history led to the differential diagnosis of cerebral abscess and neoplasm. An emergent extraventricular drain was placed to decrease regional mass effect caused by the markedly enlarged ventricles (opening pressure 15 mmH2O), and the VP shunt was removed. A mechanical cause for obstruction could not be determined after catheter inspection, and the catheter and CSF were sent for culture (no organism was isolated). The patient was scheduled for surgical debridement of the presumed abscess.

MRI performed 24 hours later (Fig. 2) for pre-operative planning demonstrated a 16mm multiloculated, spherical, cystic mass. MRI performed 24 hours later (Fig. 2) for pre-operative planning demonstrated a 16mm multiloculated, spherical, cystic mass.
fluid-filled mass with extensive surrounding vasogenic edema and a thin, mildly T2-hypointense, minimally enhancing rim. The collection demonstrated T1 and T2 signal characteristics similar to CSF, centrally suppressed on T2 FLAIR, as well as facilitated diffusion (also similar to CSF). As with CT, mass effect effaced the right lateral ventricle; this resulted in 8 mm of right-to-left subfalcine herniation with asymmetric enlargement of the left lateral ventricle. Since the imaging characteristics of the fluid collection matched that of CSF, and not an abscess or tumor, and since it was located adjacent to the VP shunt tract, the presumptive diagnosis of catheter-related CSF cyst was made. The patient’s surgical exploration was cancelled.

Followup MRI 4 days after catheter removal and contralateral extraventricular drain placement (Figs. 3A, B) demonstrated decreased size of the fluid collection (7 mm), decreased vasogenic edema, and decompression of the left lateral ventricle. MRI followup 2 months later, and after new VP shunt placement, (Figs. 3C, D) demonstrated complete resolution of the cyst, surrounding vasogenic edema, and regional mass effect. The ventricles were of normal size and symmetric. Cultures of the catheter and CSF remained negative.

Discussion
This case is unusual because it is only the second reported adult to develop a CSF cyst within the brain caused by presumed shunt occlusion. It is also novel in that it demonstrates the greatest reported delay between catheter placement and cyst development: 15 years.

Various complications of ventriculoperitoneal (VP) shunts have been reported in the literature, ranging from common tube disconnections, infections, and intraperitoneal pseudocysts (1) to more unusual cases of vaginal enterocele (2), spontaneous knot (3, 4), and migration into the heart (5, 6).

Shunt catheter malfunctions are typically divided into four main categories: 1. those that result in hydrocephalus (for example, obstruction, disconnection); 2. abdominal

Figure 2. 20-year-old male with cerebral parenchymal cyst. Select images from preoperative MRI performed 24 hours after initial CT demonstrated CSF intensity structure abutting and partly encompassing the VP shunt that was isointense to CSF on T1WI post contrast (A) and on T2 (B) with full suppression centrally on T2 FLAIR (C), and facilitated diffusion on DWI/T2 weighted trace (E) and corresponding ADC map (F). Note the presence of operative fiducial markers on T1 and T2 sequences (A, B, G). There was a faint thin rim of enhancement on the T1 post-contrast sequence also visible on the T2 and T2 FLAIR with extensive vasogenic edema in the centrum semiovale (B) and corona radiate (G). Mass effect resulted in effacement of the right lateral ventricle (D) and asymmetric enlargement of the left lateral ventricle, particularly the temporal horn (D). Mass effect is exerted on the brainstem (D). Since imaging characteristics of the collection matched CSF, and not an abscess, and it was located adjacent to the shunt, the patient’s surgical exploration was cancelled.

Figure 3. 20-year-old male with cerebral parenchymal cyst. Select coronal (A) and axial (B) T2 FLAIR images 4 days after shunt removal and contralateral EVD placement demonstrated partial crenation of the cyst, decrease in vasogenic edema, and marked improvement in mass effect. Select axial T2 FLAIR images 2 months after initial EVD placement (C and D), with interval transition from EVD to VP shunt, demonstrated normal size and symmetry of lateral ventricles (C) and complete resolution of the cyst and vasogenic edema with residual tract from initial VP shunt (D).
complications (for example, pseudocyst); 3. foreign-body complications (for example, migration, infection, fibrosis, allergic reaction), and 4. surgical complications (for example, overdrainage). Cerebral parenchymal cyst can be included in the first category.

The proposed mechanism of cyst development has been likened to that of subependymal interstitial edema development in patients with acute hydrocephalus, and to pericatheter edema in acute VP-shunt occlusion. Fluid from the pressurized ventricular system seeps into the interstitial space via bulk flow surrounding the catheter. Preferential flow around the catheter may be secondary to focal weakness in the ependymal surface due to catheter perforation, or possibly partial occlusion of side holes in the parenchyma. Formation of a discrete cyst versus simple pericatheter edema is thought to require well-formed gliosis around the catheter tract and rapid development of hydrocephalus (7).

Cases of shunt-related CSF cysts often have extensive adjacent vasogenic edema. It is likely that the same bulk flow of fluid into the interstitial space from the pressurized ventricle system forms both the cyst and the edema, as both resolve with appropriate decrease in CSF pressure. The edema may be secondary to a more gradual transition of the CSF from the ventricle system into the interstitium. It is also possible that the fluid seeps directly from the cyst into the interstitial space.

Ten cases of shunt-related CSF cysts within the brain have been reported (7-11), and only one other reported case was an adult patient (10). All of the other previously reported cases have been in pediatric patients ranging from early infancy (9) to 10 years of age (7). This case also illustrates the possibility of an extremely delayed presentation of pericatheter parenchymal cyst relative to catheter insertion. Previous reports describe cyst formation within months (8, 9, 10, 11) to the first few years after catheter insertion (7, 8, 10, 11). Presumably mature catheter tracts from longstanding shunts should not exclude shunt-related CSF cysts from the differential diagnosis.

Acute shunt occlusion can lead to low density and high intensity along the shunt tract on CT and MRI, respectively, and can also lead to CSF accumulation in soft tissues external to the skull. In this case, accumulation of CSF in the brain led to mass effect, edema, and an appearance that suggested either cerebral abscess or tumor. MRI excluded both, and led to the proper management of shunt replacement rather than biopsy.

Space-occupying lesions with secondary mass effect and edema on CT and even T2 FLAIR sequences may suggest cerebral abscess or tumor. In VP-shunt patients, comprehensive MRI should be performed to evaluate these conditions and possibly diagnose shunt-related CSF cyst. Surgical exploration may be averted in some cases, and lead to the appropriate treatment.

References
1. Goeser CD, McLeary MS, Young IW. Diagnostic imaging of ventriculoperitoneal shunt malfunctions and complications. Radiographics. 1998 May-Jun;18(3):635-651. [PubMed]
2. Karp DR, Rizvi TZ, Davila GW. Symptomatic vaginal enterocoele associated with malfunctioning ventriculoperitoneal shunt and cerebrospinal ascites. Int Urogynecol J. Sep 2011;22(9):1189-1191. [PubMed]
3. Chopra I, Gnanalingham K, Pal D, Peterson D. A knot in the catheter—an unusual cause of ventriculoperitoneal shunt blockage. Acta Neurochir (Wien). Sep 2004;146(9):1053-1056; discussion 1056-1057. [PubMed]
4. Mohammed W, Wiig U, Caird J. Spontaneous knot: a rare cause of ventriculoperitoneal shunt blockage. Br J Neurosurg. Feb 2011;25(1):113-114. [PubMed]
5. Chong JY, Kim JM, Cho DC, Kim CH. Upward migration of distal ventriculoperitoneal shunt catheter into the heart: case report. J Korean Neurosurg Soc. Sep 2008;44(4):170-173. [PubMed]
6. Kano T, Kurosaki S, Iwasa S, Wada H. Migration of a distal ventriculoperitoneal shunt catheter into the internal jugular vein and heart through the external jugular vein: case report. Neurol Med Chir (Tokyo). 2010;50(10):945-948. [PubMed]
7. Iqbal J, Hassounah M, Sheikh B. Intraparenchymal pericatheter cyst. A rare complication of ventriculoperitoneal shunt for hydrocephalus. Br J Neurosurg. Jun 2000;14(3):255-258. [PubMed]
8. Sinha AK, Lall R, Benson R, O’Brien DF, Buxton N. Intraparenchymal pericatheter cyst following ventriculoperitoneal shunt insertion: does it always merit shunt revision? Zentralbl Neurochir. Aug 2008;69(3):152-154; discussion 154. [PubMed]
9. Rim HR, Hwang SK, Kwon SH, Kim HM. Intraparenchymal pericatheter cyst as a complication of a ventriculo-peritoneal shunt in a premature infant. J Korean Neurosurg Soc. Aug 2011;50(2):143-146. [PubMed]
10. Vajramani GV, Fugleholm K. Reversible CSF cyst related to a functioning ventriculo-peritoneal shunt. Acta Neurochir (Wien). Nov 2005;147(11):1199-1202; discussion 1202. [PubMed]
11. Sugimoto K, Enomoto T, Nose T. Reversible porencephaly. Alteration of the cerebrospinal fluid flow after shunt malfunction. Child's Nervous System: Child's Nervous System. 1991;7/(7):394-398. [PubMed]