Case Report

Management of abdominal pseudocyst in shunt-dependent hydrocephalus

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Abstract

Background: Abdominal pseudocyst (APC) is an uncommon manifestation of a ventriculoperitoneal (VP) shunt that is attributed to an inflammatory response, usually the result of infection.

Case Description: A 13-year-old girl with a VP shunt presented with progressive abdominal distention, pain and vomiting. The shunt was inserted at infancy for congenital hydrocephalus. A shunt infection was treated with externalization of the shunt, antibiotics and subsequent shunt replacement. At the age of four, the shunt was revised for a distal malfunction. Nine years later, abdominal CT and ultrasound demonstrated large multiseptated cysts. The shunt was externalized and 1.8 L of sterile, xanthochromic peritoneal fluid was drained. The cerebrospinal fluid was clear, colorless, acellular and sterile with normal protein and glucose levels. Two days later, the distal portion of the shunt was replaced back into the pleural cavity. Five months later a pleural effusion formed. Thoracentesis was performed and there was no evidence of infection. The shunt was subsequently converted to a ventriculoatrial system. The patient has remained well for over 3.5 years.

Conclusion: APC represents an important complication of VP shunts, with an unclear etiology that can occur nine years after shunt surgery. This paper presents an update on the management of APCs.

Key Words: Abdominal pseudocyst, hydrocephalus, pediatrics, shunt complication, ventriculoperitoneal shunt

INTRODUCTION

Ventriculoperitoneal (VP) shunting is the preferred and most successful method for managing hydrocephalus. Abdominal complications include peritonitis, ascites, bowel and abdominal wall perforation and inguinal hernias. An uncommon but recognized complication is an abdominal pseudocyst (APC), with cerebrospinal fluid (CSF) collecting and being poorly or not absorbed across the serosa. This results in an increased pressure within the APC, reducing forward pressure gradient and optimal shunt function.

The clinical presentation may resemble that of an acute abdomen: Abdominal pain with/without a palpable mass, abdominal distention with/without tenderness, nausea and/or vomiting, decreased appetite, constipation, fever and signs of shunt malfunction such as lethargy and headache.
The term pseudocyst implies that the cyst is surrounded by a wall of nonepithelial tissue, such as intestinal serosa and peritoneum. The wall may have evolved from an inflammatory reaction to a focal peritonitis or low-grade infection. An increase in CSF protein contents may also contribute to a sterile inflammatory response. Central nervous system malignancies such as astrocytomas and primitive neuroectodermal tumors have also been thought to predispose the abdomen to the development of an APC.

Most suggest an inflammatory component causing the formation of the APC. Histopathological evidence demonstrate the presence of inflamed serosal surfaces, fibrous tissue without epithelial lining, lined with acute and chronic inflammatory cells and granulomatous tissue with fibroblasts, collagen and inflammatory cells. It has been suggested that both the lack of epithelial lining and the presence of inflammatory cells may hinder CSF absorption.

**CASE REPORT**

A 13-year-old girl born at 32 weeks gestation with congenital hydrocephalus and VP shunt presents with abdominal distension, pain and vomiting. Her shunt was inserted at 12 days of age. This was followed by a shunt infection that was treated by shunt externalization and antibiotic therapy. The shunt was later reinserted back into the peritoneum. At the age of four, the shunt was revised because of a distal malfunction. There were no other abdominal or shunt surgeries.

On examination, the patient was awake and alert, afebrile and neurologically stable with no evidence of raised intracranial pressure. The abdomen was markedly distended with diffuse tenderness and decreased bowel sounds. Ultrasound revealed significant dilatation of the small bowel loops and echogenic peritoneal fluid. CT showed large loculated/multiseptated abdominal cysts pushing the bowels and liver superiorly [Figure 1].

The VP shunt was externalized. The CSF was clear and the APC partially drained 1.8 L of yellowish fluid, and her abdominal symptoms subsided. CSF gram stain did not reveal any organisms and all cultures were negative. The leukocyte and erythrocyte count was 1 and 0, respectively. CSF protein and glucose levels were normal [Table 1]. The shunt was converted to a ventriculoperitoneal (VPl) system and the patient discharged home. Follow-up chest X-ray revealed an increasing right-sided pleural effusion with no evidence of a shunt malfunction. Thoracentesis was performed and cultures were negative for bacteria, including slow growing and rare organisms such as tuberculosis. The VPl shunt was converted to a ventricular atrial (VA) shunt, with no complications. The patient has remained well for over 3.5 years.

### DISCUSSION

The reported incidence of an APC formation ranges from 0.33% to 68%. In most cases, the etiology of an APC is not identified. It is possible that the abdomen loses its absorptive capabilities for the CSF fluid because of adhesions and/or subclinical peritonitis. Since most APC are identified soon after shunt revision, an infection may be responsible. The most common bacteria species isolated include *Staphylococcus epidermidis* or *Aureus* and *Propionibacterium acnes*. Documented infection occurrence varies between 8% and 100% [Table 2].

Egelhoff et al. states that the simple finding of an APC itself suggests an infectious process, even in the absence of signs of infection. Likewise, McLaurin et al. state that the presence of an APC in a patient with a VP shunt indicates shunt infection, even in the absence of clinical evidence of infection. It is possible that a low-grade infection of the shunt may be underdiagnosed with a single CSF culture or that the infection be transient or latent.

Plain radiographs are useful to rule out other causes of acute abdomen and help determine the continuity of the catheter tube. However, they are often normal or do not contribute any additional valuable information. Ultrasound is an easy way for a quick and reliable diagnosis of an APC. It is able to demonstrate the presence of localized peritoneal fluid collections, as well as its

### Table 1: Cerebrospinal fluid analysis results from EVD

| CSF cell count | CSF glucose concentration | CSF protein | CSF culture |
|----------------|---------------------------|-------------|-------------|
| Clear, colorless | 3.9 mmol/L | 0.12 G/L | No growth |
| CSF leucocyte-1 | | | No organism |
| CSF erythrocyte-0 | | | No WBC |

Figure 1: (a) Abdominal CT (coronal view) demonstrating the multiseptated cysts pushing the bowels and liver superiorly. (b) Abdominal CT (sagittal view) demonstrating the abdominal pseudocyst occupying most of the abdomen.
size, contents and relation of the catheter to the walls.[28]
Sonographic signs include a sonoluent mass with/without
septations, the so-called railroad sign and a fluid-echo
line.[17] More commonly, it can be visualized as a smooth
pseudomembrane echo-free fluid collection.[28] CT, while
considered as definitive as ultrasound,[11] is more costly,
and offers radiation exposure.[20] Both CT and ultrasound
can be used to percutaneously drain the APC.[8] Nuclear
shuntograms[16] and upper gastrointestinal series[9] can
also be used for evaluation.

Surgical treatment options include repositioning the
distal peritoneal catheter in a different abdominal
quadrant, shunt removal, external ventricular drainage
and conversion to either a VA or VPI system.[9,21] If
an infection is present, the shunt is externalized and
systemic antibacterial therapy administered. This
has proven to be a very effective treatment.[3] In the
absence of a confirmed infection, shunt revision alone
without prior shunt externalization is an appropriate
treatment.[19,20] Removal of the shunt catheter leads to
spontaneous resorption of the APC, and subsequent
placement of the catheter back into the peritoneum
without therapeutic drainage of the APC may lead to a
functional VP shunt again.[13,22] In some instances, the
APC itself may be watched or drained percutaneously
if it becomes symptomatic. Coley et al.[3] believe that
image-guided aspiration of the APC may be used to
alleviate acute symptoms while awaiting elective
revision of the shunt. If a VP shunt is not feasible, then a VPI
shunt is a preferred alternative,[20] with VA shunts used as
a last resort.[2]

Most reported cases of APC occur within 6 months
of the last abdominal surgery[27] with a median time
of 16 months; the longest reported cases in literature
being 8 years[21] and 15 years[7] [Table 2]. The
delayed presentation of the APC may be related to:
(1) the remote shunt infection and previous shunt
revision causing peritoneal adhesions and/or scarring;
(2) subclinical, sterile peritonitis, which may occur slowly
due to the proteinaceous content of the CSF; with the
abdomen not being able to adapt to the inflammatory
depth and ultimately unable to provide an adequate
absorptive pathway for the CSF; (3) a slow-growing,
rare organism that did not elicit the classic signs and
laboratory abnormalities associated with infection[7]
or (4) a delayed immunologically mediated reaction to the
shunt material.

**CONCLUSION**

APC represents an uncommon complication of VP
shunts of unknown aetiology in most cases. It may also
present as an acute abdomen. Factors that contribute
to decreased CSF absorption include shunt infection,
previous abdominal surgeries or multiple shunt revisions.

Our patient did not have any signs or symptoms of
infection. Analysis of CSF cultures, cell count, protein and
glucose concentration were all normal. The formation of
the APC may represent a non-infectious peritoneal reaction,
associated with a chronic inflammatory process that is
poorly understood. The unique feature of the presented
case is the delayed presentation with an acute abdomen
nine years after the last shunt revision. In addition, despite
the extreme sized APC, the patient did not present with a
shunt malfunction or signs of raised intracranial pressure.

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