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Pelvic pseudocyst presenting with vaginal drainage of cerebrospinal fluid in an adolescent: A rare complication of ventriculoperitoneal shunt

Nancy Nimer [a], Robin T. Petroze [b], Kathryn LaRusso [b], Jean-Pierre Farmer [c], Pramod Puligandla [b, *]

[a] Department of General Surgery, University of Montreal, Montreal, QC, Canada
[b] Department of Pediatric Surgery, Division of General and Thoracic Surgery, Montreal Children's Hospital, McGill University Health Centre, Montreal, QC, Canada
[c] Department of Pediatric Surgery and Neurosurgery, Montreal Children's Hospital, McGill University Health Centre, Montreal, QC, Canada

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ABSTRACT

The case report describes a 16-year-old female with a ventriculoperitoneal (VP) shunt due to congenital hydrocephalus. She presented with abdominal pain and vaginal leakage of cerebrospinal fluid (CSF). CT imaging demonstrated multiple intra-abdominal and pelvic CSF pseudocysts as well as possible erosion of the VP shunt into the vagina. She was taken to the OR for externalization of the shunt and resection of pseudocysts. Surprisingly, the VP shunt tubing was not related to the vagina at all. Instead, there was a large, inflamed pseudocyst within the pelvis and CSF was draining through the fallopian tubes which were located within it. This cyst was only partially resected to protect the fallopian tubes and surrounding structures. Post-operatively, cultures of the CSF demonstrated an infection by *Propionibacterium,* pathogens that form part of the normal skin flora, but rarely of the vaginal flora in adolescent girls (Huang et al., 2014) [1]. Vaginal drainage stopped and the patient made a full recovery. This case highlights the very rare finding of a CSF pseudocyst decompressing through the fallopian tubes and provides an overview of the complications associated with VP shunts.

1. Introduction

Complications related to ventriculoperitoneal (VP) shunts occur frequently in the pediatric population, with rates approaching 30% at 1 year and 60% by 10 years [2]. The two most common complications are shunt obstruction and shunt tract infection. However, pseudocyst formation is an uncommon but important complication, occurring in about 2.3% of patients with a VP shunt [8]. We present the rare case of a teenage girl with abdominal pain and vaginal discharge as the presenting complaint secondary to a large pelvic pseudocyst decompressing into the fallopian tube.

2. Case report

2.1. History and physical exam

A 16-year-old female with a history of congenital hydrocephalus requiring neonatal VP shunt insertion presented to the emergency department with a four-week history of vague, crampy abdominal pain that was progressively worsening. For the three days prior to her presentation, it was accompanied by copious, clear vaginal discharge and new onset constipation. Further history revealed that she did not have frequent problems with her VP shunt and that the shunt only required 2 revisions since its initial insertion, the most recent occurring 15 months prior. A review of systems noted that she had just started her regular menstrual cycle. She also denied any other symptoms including dizziness, headache, nausea or emesis. On physical examination, the patient was afebrile, had normal vital signs and a normal neurological exam. Importantly, her abdominal examination revealed a mildly distended abdomen with suprapubic fullness, which was otherwise non-tender and devoid of any peritoneal irritation. A pelvic examination revealed clear vaginal discharge, but was otherwise within normal limits.

* Corresponding author. Division of Paediatric General and Thoracic Surgery, The Montreal Children's Hospital, 1001 Décarie Boulevard Room B.04.2318, Montreal, QC, H4A3J1, Canada.
E-mail address: pramod.puligandla@mcgill.ca (P. Puligandla).

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2.2. Imaging and early investigations

Laboratory investigations included a complete blood count which was within normal limits. A CSF shunt series at that time did not show any discontinuity or significant kinking of the shunt tubing. A CT scan of the abdomen and pelvis was ordered. It revealed multiple, thick-walled, discrete fluid collections along the intra-abdominal trajectory of the VP shunt, the largest measuring 6.7 x 8.5 × 6.5 cm within the pelvis cul-de-sac (Figs. 1 and 2). While there was no clear evidence of abscess, bowel perforation or vaginal perforation, the radiologist commented on a possible fistula between the largest collection in the cul-de-sac and the vagina. There was also hydrosalpinx, with the density of this liquid being similar to that found in the vagina.

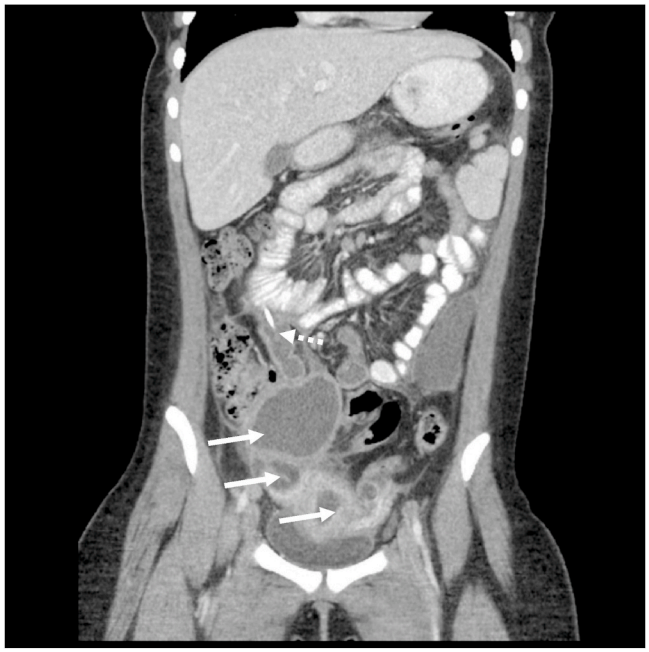

Fig. 1. Multiple well-formed collections (solid white arrows) along the intra-abdominal trajectory of the VP shunt (dotted white arrow).

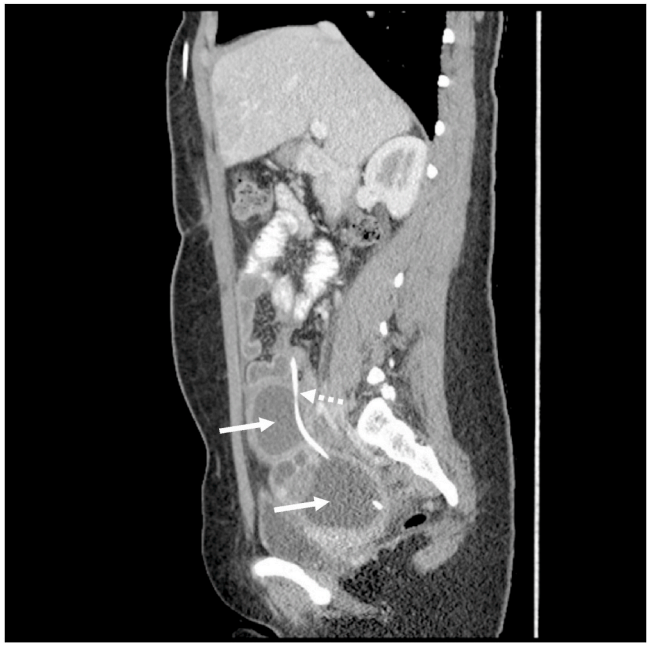

Fig. 2. Largest collection within pelvis cul-de-sac (white arrows).

2.3. Perioperative course

The patient was admitted to the hospital and started on intravenous antibiotics for a possible shunt infection. She was taken the following day for laparoscopic exploration of the multiple CSF pseudocysts. Intra-operatively, there were dense adhesions along the tract of the VP shunt catheter with multiple large pelvic pseudocysts (Fig. 3). There was no evidence of peritonitis. The pseudocysts were systematically resected and drained being careful not to damage adjacent intra-abdominal viscera (Figs. 4 and 5). Fluid was sent for gram stain and culture. The scarring was most intense in the pelvis which made the identification of the fallopian tubes and ovaries extremely difficult. The cysts were also adherent to the cecum and sigmoid

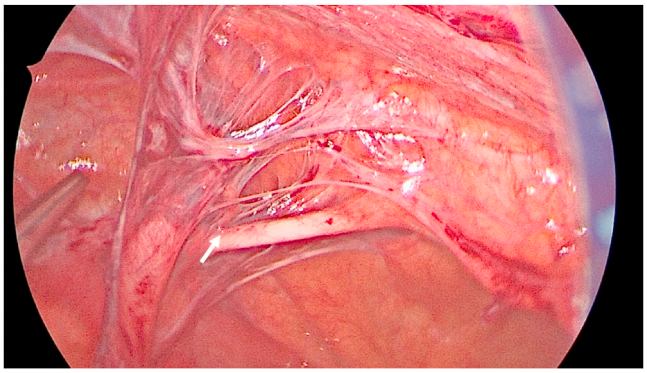

Fig. 3. Intraoperative dense fibrous adhesions between the shunt tubing (white arrow) and pseudocyst multiple large pelvic pseudocysts.

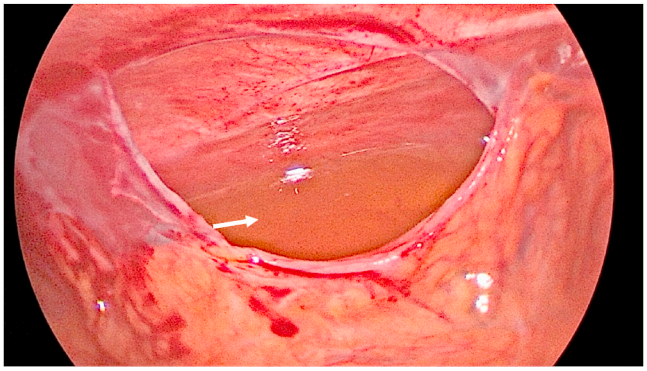

Fig. 4. Fluid within one of the pseudocysts (white arrow).

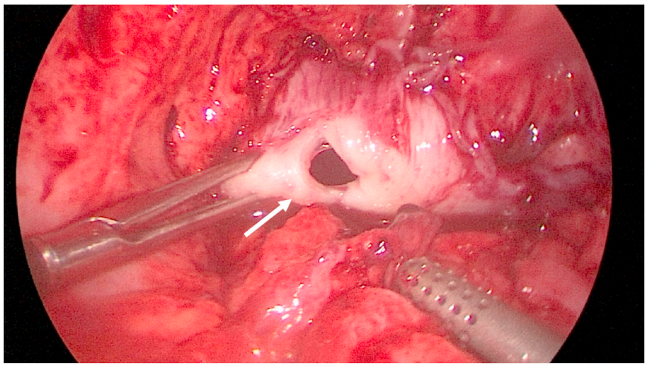

Fig. 5. Removal of pseudocyst indicating thick-walled collection (white arrow) and adherence to surrounding structures.

colon. Only subtotal resection of the pelvic pseudocysts could be achieved due to fear of damaging the pelvic organs. However, the resection was adequate enough to rule out vaginal fistulisation as the end of the shunt tubing was not near pelvis. This was confirmed with intra-operative contrast instillation via the vagina (Fig. 6). At the completion of the operation, the VP shunt was externalized.

The patient's post-operative recovery was uneventful and the vaginal drainage stopped in the early post-operative period. While the CSF cultures from the valve did not become positive, cultures of the intra-abdominal VP shunt tubing were positive for *Propionibacterium acnes,* a skin organism. The patient was subsequently treated with penicillin. The revision of the intra-abdominal portion of the VP shunt occurred three weeks later after repeat cultures were negative.

3. Discussion

Developing either at the ventricular or peritoneal end, most complications of the VP shunt can be broadly categorized as either mechanical or infective [4]. Mechanical complications include shunt obstruction, disconnection, migration and, rarely, hollow viscus perforation [4–6]. Infective complications include shunt tract infection, ventriculitis, peritonitis, and abscess formation [3–5]. Other complications include CSF ascites, subdural hematoma, and pseudocyst formation [3–7].

We present the first published case of spontaneous decompression of an intra-abdominal CSF pseudocyst via the fallopian tubes in an adolescent female with a VP shunt. There is one prior report in the literature of a distal shunt catheter migrating into a fallopian tube and causing hydrosalpinx and similar copious, clear vaginal discharge [9]. In that case, the surgeons performed a salpingectomy in order to remove the catheter. Our patient's clinical presentation was unique as she presented with progressive abdominal pain, clear vaginal discharge, and multiple large pseudocyst on imaging, but no overt symptoms of distal shunt failure. We hypothesize that decompression of the pseudocyst via the fallopian tubes, uterus and vagina mitigated the development of neurological sequelae that would normally be observed with distal shunt dysfunction.

Pseudocyst formation is a rare complication that occurs when CSF resorption by the peritoneum is disrupted. Ultimately affecting approximately 2.3% of patients with a VP shunt, CSF pseudocysts may present with either neurological or gastrointestinal symptoms, or both [8]. In younger patients, symptoms are most commonly related to increased intracranial pressure (i.e. lethargy, headache, nausea and vomiting) from distal shunt obstruction [3,5]. Gastrointestinal symptoms generally occur in older children and adolescents and include anorexia, abdominal pain, distension, constipation, or even a palpable mass or fullness [10].

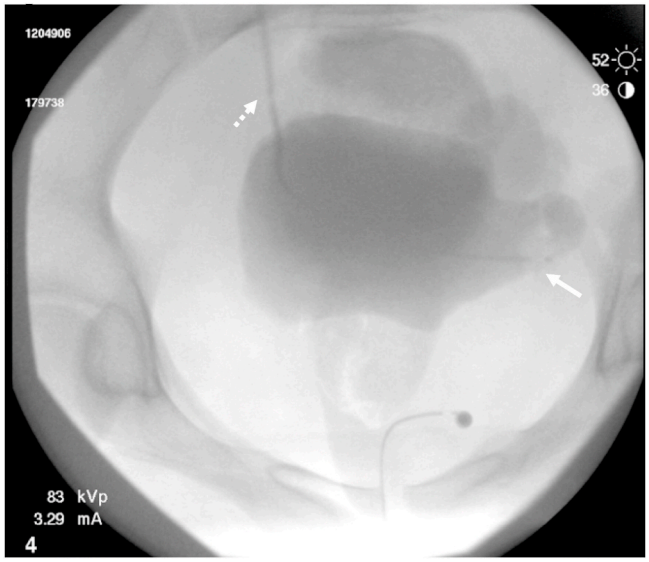

Fig. 6. Fluoroscopic view of pseudocyst with injection of contrast into the distal shunt tubing (dotted white arrow). There is opacification of the left fallopian tube demonstrating hydrosalpinx (solid white arrow), but no opacification of the vagina, bowel or bladder.

While there are several mechanisms that may lead to the development of CSF pseudocysts, (e.g. intra-abdominal adhesions, chronic inflammation, increased CSF protein), infection is the most frequent [5,11,12]. Skin flora (e.g. coagulase-negative staphylococci, *Staphylococcus aureus*, and *Propionibacterium acnes*) are most commonly implicated and usually result from surgical contamination [13]. The growth of *Propionibacterium acnes* in the peritoneal fluid cultures of our patient suggests that a shunt infection was the likely cause of the multiple pseudocysts encountered. Furthermore, infection by *P. acnes* differs from other peritoneal bacterial infections since it usually presents in older patients (>1-year-old) and can occur after a prolonged latent period following surgical manipulation. Indeed, our patient's most recent VP shunt revision had occurred 15 months prior to her most recent presentation. Moreover, infections with *P. acne* are typically indolent and often demonstrate normal serum C-reactive protein and leukocyte values [14].

The management of CSF pseudocysts is dependent on the sterility of the CSF. If cultures demonstrate infection, the shunt is externalized and the patient managed with systemic antibacterial therapy [2]. However, culture results may take several days before they are positive and they may be falsely negative depending on bacterial load. Pseudocysts that cause gastrointestinal symptoms can be managed by a variety of methods. These include shunt externalization followed later by repositioning the peritoneal catheter to an alternative location (e.g. different abdominal quadrant, the pleural space or the right atrium). Shunt removal should only be considered if there is a clear determination of shunt independence which is only in 3–9% of patients [15].

Surgical intervention is generally required for more severe symptoms. The suggestion of a vaginal fistula from VP shunt erosion on preoperative imaging was the main impetus for semi-urgent operative intervention in this case. While a VP shunt has previously been reported to cause a vaginal perforation in a post-hysterectomy patient, our intra-operative findings strongly suggested that the vaginal leakage was the result of decompression of CSF via the fallopian tube that was “trapped” within a large pseudocyst. This hypothesis is supported by our findings at laparoscopy during which we were able to clearly see the distal end of the VP shunt away from any of the pelvic organs. A large pseudocyst occupied the entire pelvis and dissection revealed small communications with others scattered throughout the abdominal cavity. Furthermore, intra-operative contrast studies failed to demonstrate any fistula. Lending further support to our hypothesis is the fact that the patient's vaginal leakage promptly stopped after pseudocyst excision.

4. Conclusion

CSF pseudocysts can lead to several different complications and their management often requires surgical intervention. We present the first report of a teenage female who presented with abdominal pain and clear vaginal discharge secondary to the decompression of multiple CSF pseudocysts via the fallopian tube. The fallopian was spared. Excision and unroofing of the pseudocysts and externalization of the shunt also led to prompt cessation of vaginal drainage early in the postoperative period with no further recurrence on imaging after 1 year.

Patient consent

Consent to publish the case report was obtained from the patient.

This report does not contain any personal information that could lead to the identification of the patient.

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Disclosures

The authors have no financial disclosures.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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