Abdominal Pseudocyst: A Rare Complication of Ventriculoperitoneal Shunts

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ABSTRACT

Ventriculoperitoneal shunts (VPS) remain a common procedure done as part of the management of hydrocephalus. VPS failure most commonly occurs within the first year and abdominal pseudocysts (APC) is a rare and underreported complication. In this report, we present a five-year-old female with a VPS who presented with acute onset of abdominal pain, vomiting, and fever and a diagnosis of APC was established. In this paper, we demonstrate our approach to the management of APC, including a review of the literature for alternative treatment options.

Keywords: Ventruloperitoneal shunt, Abdominal peritoneal pseudocyst, Hydrocephalus

INTRODUCTION

Ventriculorperitoneal shunt (VPS) placement is a standard procedure performed for the treatment of hydrocephalus. Around 30,000 VPS procedures are carried out in the United States per year[1]. VPS failure rates that occur within the first year are estimated at 11-25% and are higher in the pediatric population than adults, with the most common cause being shunt obstruction[2]. A rare and perhaps underreported complication of VPS insertion is the formation of an abdominal pseudocyst (APC).

An APC is a collection of cerebrospinal fluid (CSF) that forms around the distal end of a VPS, and is lined by fibrous tissue. APC has been reported to occur in 1 to 4.5% of pediatric cases and is often associated with significant morbidity[2]. Theories proposed to explain the aetiology behind pseudocyst formation include the presence of an infection, or non-specific chronic inflammation towards a component in the shunt or within the CSF itself. To allow CSF drainage to continue after the formation of an APC, the distal shunt has to be re-positioned within the peritoneum or in a non-peritoneal space, such as the pleura or the atrium. An alternative is the complete removal of the shunt and the use of an external ventricular drainage system.

We herein present a complex patient with VPS failure due to a large APC, highlighting effective strategies for management including a review of the literature for alternative treatment options.

THE CASE

A five-year-old female presented to the emergency department complaining of acute onset generalized abdominal pain, vomiting, and foul-smelling urine. The patient's past medical history is significant for a congenital lumbar spine meningocele associated with hydrocephalus. Both were managed surgically on day three of life with surgical closure of the spinal defect and VPS placement. From a urinary point of view, the patient was being managed with clean intermittent catheterization (CIC) for her neurogenic bladder and prophylactic antibiotics for bilateral dilating vesico-ureteric reflux (VUR). The patient also developed an infarct of the left front temporoparietal area, in the region supplied by the middle cerebral artery, at four years of age. Although investigated extensively, the cause for her infarct is still unknown.

On admission, she was febrile with temperatures reaching up to 39°c and had generalized abdominal tenderness. Urinary tract infection was confirmed by urine cultures positive for Pseudomonas Aeruginosa. She was placed on a 14-day course of intravenous antibiotics (gentamicin and ceftazidime), and repeated cultures were negative. Despite this, the patient had persistent abdominal pain, abdominal distention, and spiking temperatures.

An abdominal ultrasound (Figure 1) was therefore performed which revealed a large intra-abdominal septated collection. This was followed

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by a CT scan which confirmed the presence of a complex intra-abdominal pseudocyst measuring 5.5 cm by 7.5 cm, with the distal end of the VPS coursing within it (Figure 2). Considering the size of the collection and the clinical status of the patient, an ultrasound guided pigtail catheter was inserted and drained 200 ml of clear fluid. A repeat ultrasound scan showed a collapsed pseudocyst, and cultures from the aspirate were negative. Following the pigtail insertion, the patient's clinical status improved significantly, her abdominal pain subsided, and the fever settled. Over the following days, CSF was continuously draining through the pigtail, and in effect became an external CSF drain. Distal shunt re-siting was delayed for four weeks to allow further patient recovery and resolution of the intra-abdominal inflammatory process.

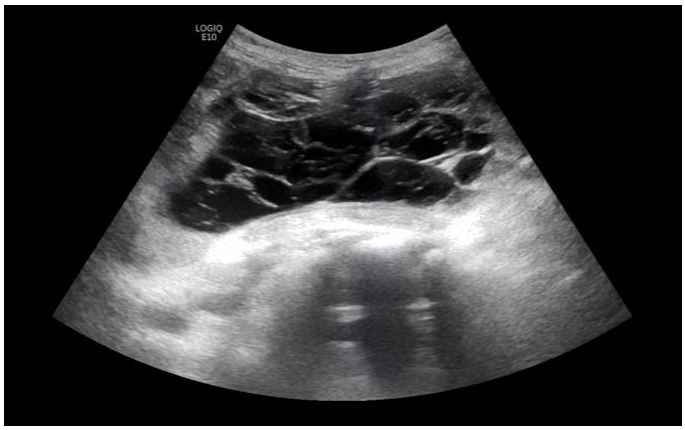

Figure 1: An abdominal ultrasound showing a large intra-abdominal septated collection

Pre-operatively, a repeat ultrasound showed the pseudocyst just below the umbilical level. As such, laparoscopic camera port access was obtained through a right sided incision. Significant intra-abdominal adhesions were noted making it nearly impossible to identify normal anatomy. The distal tip of the VPS was identified, carefully pulled out of the pseudocyst and re-sited to the right lower quadrant. The patient recovered well in the initial post-operative period. Four days post-operatively, an abdominal ultrasound was performed due to new onset abdominal pain which revealed a newly forming right sided collection around the re-sited VPS tip. Another ultrasound 3 days later confirmed that this new collection was growing in size. This together with the intra-operative findings indicate that the patient's peritoneal cavity has become inadequate for CSF absorption.

The patient was scheduled for immediate distal shunt re-siting from the peritoneal cavity to the pleural cavity (Figure 3). The procedure was uneventful, and the distal end of the tube was externalized and used to aspirate the remaining intra-abdominal collection. VPS valve cultures were positive for Pseudomonas Aeruginosa, and hence the patient was re-started on a 14-day course of intravenous gentamicin and piperacillin-tazobactam.

The patient made an excellent recovery over the ensuing 2 weeks with a functioning ventricular-pleural shunt (VPLS) and no recurrent abdominal collection. The patient was followed up in the out-patient's clinic 6 months later, with no new complaints, and the shunt has been working well so far.

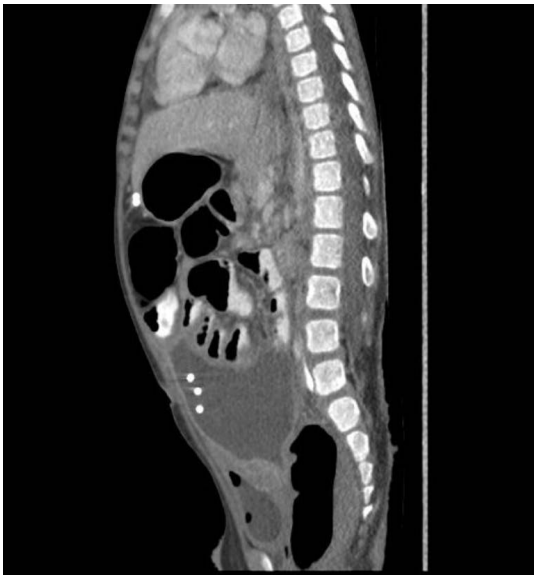

Figure 2: Abdominal contrast CT scan of a complex intra-abdominal pseudocyst measuring 5.5 cm by 7.5 cm, with the distal end of the VPS coursing within it

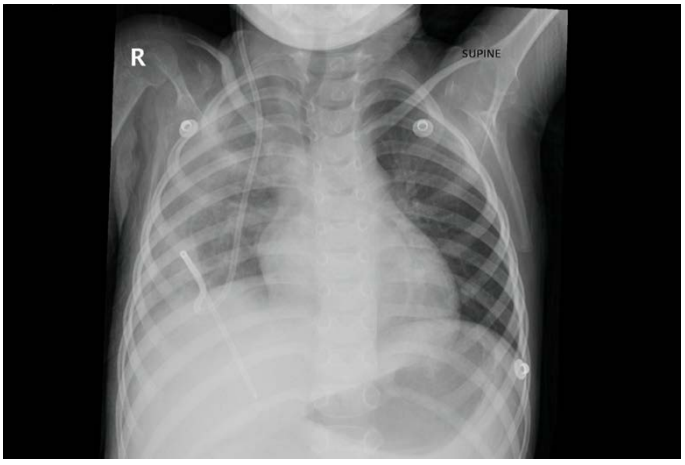

Figure 3: Chest Xray showing distal ventriculopleural shunt

DISCUSSION

APC is a rare complication of VPS and was first recognized by Harsh in 1954[3]. It is usually a late complication and occurs a year or more after shunt placement or revision[2]. APC is a collection of CSF around the distal tip of the catheter lined by fibrous tissue. Patients may exhibit non-specific systemic signs such as fever and vomiting with or without abdominal distention or a palpable mass[2]. Plain X-ray of the abdomen is often the first line of investigation and may show an opacified parenchymal round mass around the tip of the catheter[4]. Ultrasound of the abdomen is reliable in establishing the diagnosis of APC, as it is able to delineate the cyst size and location and show its proximity to the catheter. Further anatomical detail and pre-operative planning can be obtained by performing an abdominal CT scan, although this is left to the surgeon's discretion[5].

The aetiology of APC has been debated. Grunebaum et al. first described APC as a chronic inflammation of the abdominal serosa secondary to the tip of the VPS[6]. Egelhoff et al. suggested that the primary trigger for APC formation is a subclinical chronic infection, which may occur even in the absence of systemic signs of infection[7]. Goldfine et al. described it as an allergic or nonspecific inflammation to the catheter or to a component of the CSF[8]. Others have described increased CSF protein as the cause of APC formation[9]. Unfortunately, the aetiology of APC remains unclear to date.

The absence of epithelium in a pseudocyst means it is unable to absorb CSF nor secrete fluid[4,10,11]. Hence, repositioning the distal tip of the catheter elsewhere within the peritoneum, or in an alternative epithelial space, is essential for CSF shunting to continue[5]. An alternative approach is the complete removal of the shunt and the use of an external ventricular drainage system[5].

The surgical procedure chosen relies heavily on the presence or absence of infected CSF[5,11]. Infection can be confirmed by CSF culture, or more accurately by a culture of the catheter tip[10]. If cultures are positive, a common treatment strategy includes systemic antibiotics, removal of VPS, and placing an external ventricular drainage (EVD) system[4,5,9,11-13]. Once CSF cultures become sterile, shunt re-siting can be performed[4,9]. However, some authors believe that EVD is best avoided as it poses additional unnecessary risks to the patient, such as superinfection of the CSF[4]. Instead, they recommend replacement and repositioning of the shunt to the pleura or atrium after a period of systemic antibiotic therapy[4].

In cases where cultures are negative, shunt externalisation is unnecessary, and shunt repositioning alone can be carried out[4,5,9,11,12]. However, APC may result from a subclinical infection and as such we believe antibiotic prophylaxis is always necessary[11]. In our case, a percutaneous drain was inserted as a substitute to shunt externalisation and after confirmation of negative cultures and sufficient antibiotic therapy, laparoscopic repositioning of the catheter tip was performed. Image guided percutaneous drainage has several other advantages including alleviating acute symptoms, controlling intra-abdominal sepsis, and obtaining aspirate for culture[5,10,11]. It also allows a period of watchful waiting which we believe is vital to avoid entering a hostile abdomen early in the patient course.

Percutaneous drainage of the pseudocyst alone is inefficient because recurrence is inevitable once the drain is removed[4]. Thus, re-siting the catheter tip is ultimately necessary. During this procedure, the pseudocyst can be intraoperatively drained and excised, or left to resorb spontaneously[5,9]. Gaskill concluded that by re-positioning the catheter, the APC resorbed spontaneously without the need of surgical excision or drainage[13,14]. However, it must be monitored post-operatively, and if symptoms arise, it can be percutaneously drained[5].

Site of shunt placement or replacement is case dependent. VPS is favored as it has the lowest rates of intraoperative and postoperative complications[5]. Although the formation of an APC suggests an inefficiency in the peritoneum's ability to shunt CSF, it is not considered an absolute contraindication for re-using the peritoneal cavity[3,10]. This inefficiency lasts for an unknown period of time[3,10], which creates a challenge to the treating surgeon and indeed when counselling patients as the suitability of the peritoneum cannot be known prior to re-siting[3].

Regardless of the timing of the catheter repositioning, the recurrence rates of APC are still high [3,4,7,9,10,14], and this is independent of the operative techniques, the presence or absence of infection, or the use of antibiotics[3]. However, a known risk factor for APC formation is a history of multiple shunt revisions[11]. Therefore, in such patients and especially when significant intra-abdominal adhesions are evident intra-operatively, diverting the shunt to the pleural cavity or the atrium is probably a better alternative[9].

In our case we initially chose to re-use the peritoneal cavity as the patient did not have a history of shunt failure or shunt revisions. We also chose delayed re-siting as opposed to immediate, to allow time for the inflammatory process to settle. By doing so, we were aiming at decreasing the chances of converting to laparotomy which would have rendered the patient's peritoneum inadequate for shunting. However, as the APC recurred within a few days, we concluded that the patient's peritoneum was incapable of acting as a shunt and decided to divert the shunt into the pleural cavity instead.

CONCLUSION

As of yet, no established algorithm exists for the management of VP shunt associated pseudocysts in children. We propose that percutaneous drainage, as performed in our case, is an effective and safe strategy in the acute period. It allows resolution of the inflammatory process and avoids the risks of entering a hostile abdomen early in the patient course. Delayed shunt revision may be attempted and can easily be performed laparoscopically, but is associated with a high failure rate. Patients must be counselled regarding this risk and the high likelihood of requiring a new shunt type following pseudocyst development.

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