Case Report

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An Unusual Cause Of Ascites: Cerebrospinal Fluid Pseudocyst

Sabrina Xin Zi Quek[1*], Keith Wei Jie Ching[2], Kamarjit Mangat[3], How Cheng Low[1] and Stephanie Q Ko[2]

[1]Division of Gastroenterology and Hepatology, Department of Medicine, National University Hospital, Singapore 119228, Singapore
[2]Department of Medicine, National University Hospital, Singapore 119228, Singapore
[3]Department of Radiology, National University Hospital, Singapore

[*]**Corresponding author:**
Sabrina Xin Zi Quek,
Division of Gastroenterology and Hepatology, Department of Medicine, National University Hospital, 1E Kent Ridge Road, Singapore 119228,

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1. Abstract

We report a rare case of a cerebrospinal fluid (CSF) perihepatic pseudocyst presenting as symptomatic transudative ascites in a 42-year-old patient with a history of ventriculoperitoneal (VP) shunt insertion for spina bifida associated hydrocephalus. Our patient was admitted initially for a urinary tract infection and during his hospital stay, developed recurrent symptomatic abdominal fluid collections resulting in a prolonged hospital admission for workup. Through this case we highlight the evaluation methodologies of such an unusual cause of an abdominal fluid collection, the epidemiological considerations in our patient population, and the pertinent investigations utilised to clinch the eventual diagnosis.

The majority of CSF pseudocysts in literature are reported to occur within 3 weeks to 10 years from insertion of the VP shunt, whereas our patient presented more than 25 years after the initial insertion. And while most cases are usually diagnosed via radiological imaging, this case also shows how fluid beta-2 transferrin can be used as a confirmatory diagnosis of CSF pseudocyst.

2. Keywords:
Ascites, Cerebrospinal Fluid, Pseudocyst, Ventriculoperitoneal Shunt

3. Introduction

A 42-year-old male with a history of spina bifida was admitted initially for a urinary tract infection and later developed recurrent symptomatic abdominal fluid collections resulting in a prolonged hospital admission for workup.

His spina bifida was complicated by bilateral congenital talipes equinovarus and neurogenic bladder with vesicoureteral reflux. He had multiple urological revision procedures over 20 years, the most recent being a total cystectomy and revision of ureteroileal conduit at age 32. He had a long term suprapubic catheter which was complicated by recurrent urinary tract infections. The patient also had a ventriculoperitoneal shunt (VPS) inserted for hydrocephalus prior to 1995 – this was not stated in his medical records prior to this admission, and the patient himself could not recall the exact year of VP shunt insertion.

The patient first presented with fever with hematuria and diagnosed with a urinary tract infection with an early prostatic abscess. Two weeks into his intravenous antibiotic course, he developed symptomatic abdominal distention associated with non-bilious, non-bloody vomiting. A Computer Tomography of the Abdomen and Pelvis (CTAP) was performed and revealed a perihepatic fluid collection in the right upper quadrant measuring approximately 17.4 x 18.0 x 15.6 cm (Figure 1) with mass effect on adjacent structures. Imaging of the liver demonstrated reduced attenuation suggestive of fatty change. The imaged pancreas, spleen, biliary tree and hepatic and portal veins revealed no focal abnormalities.

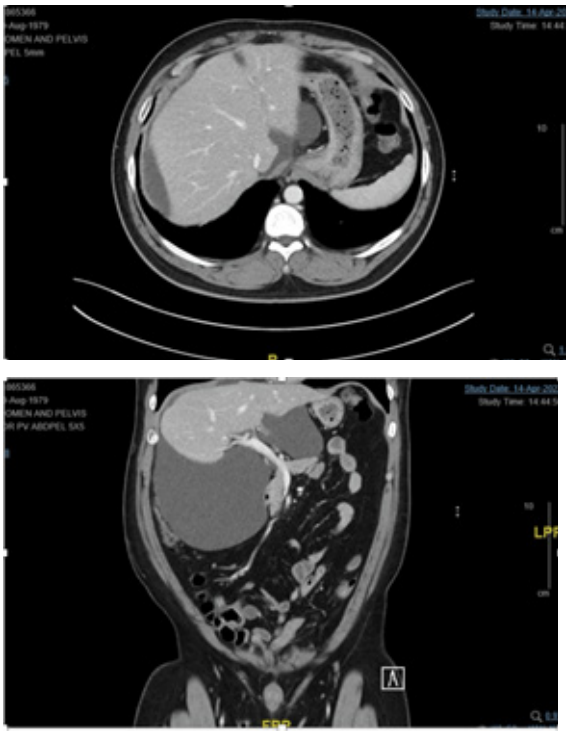

Figure 1: Initial CTAP

The patient underwent radiologically guided percutaneous drainage of the fluid collection which drained clear colourless fluid (see Figure 2). Diagnostic analysis of the fluid studies demonstrated low total protein (<8 g/L), normal lactate dehydrogenase (LDH, 61 U/L), a nucleated cell count

of 12/μL with a differential count of 47% lymphocytes, 46% monocytes, 7% neutrophils with occasional mesothelial cells. Fluid cultures, and Acid Fast Bacilli smear and cultures yielded no growth. Cytopathological studies demonstrated a chronic inflammatory yield with no malignant cells identified. Comparison of paired fluid with serum studies demonstrated a high serum ascites to albumin gradient (SAAG; serum albumin 38 g/L, fluid albumin 5 g/L) of 33 g/L.

Figure 2: IR guided peri-hepatic drain collection

A total of 5.5L of abdominal fluid was drained over 7 days and the drain was removed as there was decreasing output. However, 5 days after removal of the abdominal drain the patient reported of increasing abdominal distention. Interval imaging done revealed a slightly larger perihepatic collection measuring 18.0 x 18.2 x 19.0 cm with no other abdominopelvic collections identified (Figure 3). In view of persistent symptomatic abdominal distension radiologically guided percutaneous drainage was arranged 3 weeks after removal of his first drain. 10 days following the second drain insertion, the patient again reported persistent abdominal distention with minimal drain output. He then underwent a drain exchange and insertion of a third drain. The interventional radiologist noted that the collection was septated, hence a single drain was insufficient to completely drain the collection. Over the 3 months where there were ongoing investigations to ascertain the cause of the peri-hepatic fluid collection, the patient had a total of 6 percutaneous drain insertions and 2 drain exchanges.

The patient underwent extensive investigations for a high SAAG fluid collection, initial differentials being liver cirrhosis or non-cirrhotic portal hypertension (e.g. right heart failure or veno-occlusive disease).

A fibroscan demonstrated liver stiffness of 12.9kpa (F4 if assuming etiology as fatty liver). Transjugular hepatic venous pressure gradient (HVPG) measurements showed a hepatic vein wedge and free venous pressure of 21mmHg and 16mmHg respectively, giving a HVPG of 5mmHg (normal range). Liver biopsy showed mild mixed macro and microvascular steatosis with steatohepatitis, portal-periportal inflammation, portal-periportal fibrosis with septal linkage, ground -glass hepatocytes with graded severity of F3 fibrosis at most.

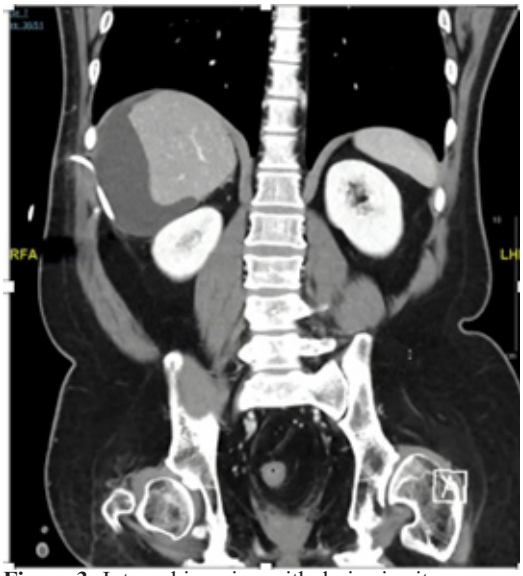

Figure 3: Interval imaging with drains in situ

Transthoracic echocardiography revealed normal left and right ventricular systolic function and preserved left ventricular ejection fraction of 65%. Percutaneous direct portal venogram demonstrated no obstruction to vessel hemodynamics, and flow direction was not suggestive of portal hypertension.

Other surgical causes of a fluid collection were also investigated. Abdominal fluid creatinine and serum creatinine were identical at 55 μmol/L, ruling out a urinoma.

4. Results

In view of the unyielding diagnostic evaluations, a multidisciplinary discussion was conducted. Significance was drawn to the clear colourless nature of the peri-hepatic fluid, raising the possibility of a cerebrospinal fluid (CSF) pseudocyst. Preceding this discussion, the patient was not known by the managing medical team to have a VP shunt, as it was not stated in the patient's case notes, nor was it deemed as a relevant component of the history at presentation. Furthermore, the VP shunt was not visualised on the initial imaging scans. Specific history elucidated from the patient revealed that the VP shunt was inserted more than 27 years ago. In consultation with our neurosurgical colleagues, a beta-2 transferrin was sent from the drained fluid which returned positive.

Re-evaluation of the initial CTAP imaging that had been performed, and a specific X-ray VP shunt series did not demonstrate presence of the shunt within the pseudocyst. Nonetheless, the patient consented for and

underwent a VP shunt exploration and revision surgery. Intraoperative findings revealed a calcified catheter and burr hole mounted ventricular catheter with good clear CSF flow at the ventricular end. The distal straight catheter was cut off with a new connector applied and the shunt was eventually re-sited into the pleural space. Drain output of the remaining abdominal drain was 250ml on the day of operation, this decreased rapidly with only blood stained fluid in the drain tubing on postoperative day (POD) 1 and having no further output on POD2. Within a week, the patient's abdominal distention was resolved. He did initially experience pleuritic chest discomfort after the re-siting of the shunt into the pleural space, but this gradually resolved. There were no other postoperative complications. An interval CTAP performed 5 days after the VPS revision surgery demonstrated a reduction in the peri-hepatic collection with only a thin sliver seen.

5. Discussion

The diagnostic approach to this case covered an extensive investigation to an abdominal fluid collection. Initial cross sectional imaging can be helpful to delineate the anatomy and look for signs of perforation, malignancy or infection. Evaluation of the abdominal fluid would include: (1) cell count and differential; (2) gram stain and cultures for infective causes; (3) albumin with a paired serum albumin to calculate the SAAG to differentiate between exudative and transudative etiologies; and (4) cytology for malignant etiologies. In TB endemic regions like Singapore, (5) tuberculosis (TB) smear, culture, and adenosine deaminase (ADA) should be considered. Other fluid studies such as LDH concentration and glucose are less specific, but when elevated may support the diagnosis of malignancy, infection or bowel perforation associated fluid collection. Visual inspection of the abdominal fluid is helpful, such as bloody fluid in a hemoperitoneum, milky in chylous ascites or a clear colourless fluid in CSF as in this case.

Our patient's fluid studies demonstrated a transudative ascites with no evidence of infection or malignancy, with a normal serum albumin. Investigation for cirrhosis included a fibroscan, transjugular liver biopsy and HVPG measurements; non-cirrhotic causes of portal hypertension include an echocardiogram and a portal venogram. In our patient, after these investigations were unyielding, other bodily fluids, such as urine, bile and CSF were then considered as alternative etiologies of the fluid collection. In retrospect, these alternatives may have been considered earlier, when there were no laboratory or clinical signs of liver disease or right heart failure.

5.1. Cerebrospinal Fluid (CSF) Pseudocyst

A ventriculoperitoneal (VP) shunt may be inserted in patients with hydrocephalus secondary to intracranial tumours, congenital malformations (e.g. spina bifida), meningoencephalitis, intracranial hemorrhage etc [1]. CSF pseudocyst is a known but uncommon complication of VPS with a complication rate of <1- 4.5% [2]. It presents more commonly within 3 weeks up to 10 years from insertion, with the longest reported onset being 21 years [3]. The paediatric population more commonly presents with symptoms of elevated intracranial pressure, whereas the adult population predominantly presents with abdominal symptoms such as pain and distension [4-7]. In the literature, the diagnosis is typically made by imaging (Ultrasound or CT) showing distal tip of the VPS within a homogeneous intraperitoneal collection [2,3,5,6].

Our patient is unique in several ways. Firstly, his VP shunt was inserted at least 27 years ago (it was present prior to the first records we have in 1995), making this the longest reported time from insertion for this complication to occur. Secondly, because of his complex surgical history and recurrent urinary tract infections, his pseudocyst was loculated requiring multiple drains for symptomatic relief. Therefore, though radiologically guided drainage has been proposed as a management option, it was not effective for our patient [8].

Lastly, though imaging usually plays a key role in the diagnosis of such a complication, our patient's VP shunt could not be seen on the multiple CT scans he underwent, during the ultrasound guided abdominal drain insertions or in the dedicated XR VP shunt series [9, 10]. The suspicion of a CSF pseudocyst had to be confirmed with beta-2 transferrin. Fluid beta-2 transferrin is a protein found in CSF and inner ear perilymph. It has been used in the field of Otolaryngology and neurosurgery for diagnosis of CSF leakage [11-15]. Its absence in other bodily fluids makes it a specific test for CSF and it has also been shown to be sensitive for detecting CSF even at small volumes with Warnecke et al reporting a test sensitivity of 0.97 and specificity of 0.99 for CSF [16]. In our patient, it was a useful assay for the confirmation of CSF accumulation from fluid drained directly from the perihepatic collection.

6. Conclusion

Peri-hepatic CSF collection as a result of VPS migration is a rare but important consideration in patients presenting with abdominal distention and fluid accumulation in the peri-hepatic region. Fluid beta-2 transferrin is a useful assay for confirmation of CSF accumulation. Our patient highlights the importance of keeping a broad differential diagnosis in a diagnostic dilemma, especially in a patient with complex medical and surgical history.

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