Management Dilemma of a Massive Intra-Abdominal CSF-oma

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ABSTRACT

CSF-oma is a rare complication of ventriculo-peritoneal shunt. Progressive abdominal symptoms are often a common feature at presentation. The authors report the first case of CSF-oma in Saudi Arabia for a patient who presented with a progressive abdominal distention, acute urinary retention and a leaking gastrostomy tube. Computerized tomography (CT) scan showed a massive CSF-oma pushing on the liver with secondary compression on the inferior vena cava and the right atrium, centralization of the small bowel loops, displacement of the large bowel to the anterior abdominal wall, as well as bilateral obstructive uropathy. The CSF-oma was managed using a laparoscopic approach without the need for shunt externalization nor a diversion to the vascular system. There were no signs of recurrence, clinically or radiologically at 6 month of follow-up.

Keywords: CSF-oma; Obstructive Uropathy; Ventriculo-peritoneal shunt

INTRODUCTION

Ventriculo-peritoneal (VP) shunt is a well-recognized therapeutic modality for obstructive hydrocephalus. CSF-oma or peritoneal cerebrospinal fluid (CSF) pseudo-cyst is a rare but potentially fatal complication of VP shunt with an incidence of 1% to 4.5% [1, 2]. It was first described by Harsh [3] in 1954, and thereafter various techniques have been utilized to correct this complication [4]. Initial symptoms often involve the gastrointestinal system making them of the commonest mode of presentation [5, 6].

We are reporting the first case of CSF-oma in Saudi Arabia presenting with obstructive symptoms and signs of gastrointestinal, urinary and cardiovascular systems.

CASE REPORT

A 12-year-old boy presented to the emergency department with abdominal distention and urinary retention. The patient had a history of cerebral palsy with developmental delay, obstructive hydrocephalus on VP shunt, and complex type seizure disorder on treatment following neonatal meningitis. He had a gastrostomy tube inserted in early childhood due to feeding difficulties. The patient had a two month history of progressive abdominal distention treated with diuretics (spironolactone and furosemide). He was admitted with progressive abdominal distention, not responding to diuretics, fever, decreased urine output, and gastrostomy tube leakage for further investigations.

Initial blood investigations and blood cultures identified no signs of blood borne infection, with normal renal and hepatic profile. Urine culture grew Klebsiella pneuomoniae. Gastrostomy tube site culture was positive for yeast. Accordingly, antibiotic therapy was initiated. Plain x-ray of the abdomen revealed a distended abdomen with diffuse haziness suggestive of ascites with significant displacement of internal organs [Figure 1]. Diagnostic ascitic tap illustrated a serum-ascites albumin gradient (SAAG) of > 1.11g/dL raising the suspicion of a heart failure or portal hypertension. Ultrasound showed normal liver and hepatic vessel with normal biliary tree. Transthoracic echocardiogram (TTE) revealed normal systolic heart function with no major structural defects except mild compression of inferior vena cava and right atrium by the liver secondary to the massive ascites.

Computerized tomography scan (CT scan) with contrast revealed massive ascites (CSF-oma) with mass effect causing central gathering of small bowel loops, lateral displacement of the

large bowel [Figure 2a and Figure 2b] and anterior compression of the stomach with resultant displacement of the gastrostomy tube. In addition, moderate bilateral hydrouretronephrosis was also noted [Figure 2a]. Clinical and radiological examination demonstrated no signs of shunt malfunction. Laparoscopic exploration was performed. Under general anaesthesia and endotracheal intubation, the patient was prepped and draped in supine position. Using an angio-cath, the fluid was aspirated to have a safe insertion site of a 5mm step trocar. This was confirmed endoscopically to be within the CSF- oma cavity and around 5.5L of fluid was suctioned out. With the aid of a 5mm 30° scope, additional 3-5mm trocars (2 of each) were inserted under direct vision. The transverse colon and flexures were taken down from their adhesions to the anterior abdominal; the cyst wall was excised until a free common peritoneal cavity was established with an apparently functioning peritoneal catheter of the VP shunt, without the need for externalization nor conversion to ventriculo-venous or ventriculo-atrial shunt. Post-op period was unremarkable and there were no signs of CSF-oma recurrence, clinically or radiologically, at 6 month of follow-up.

DISCUSSION

Obstructive uropathy resulting from a direct effect of a CSF-oma in a patient with no established urological anomaly is extremely rare. On literature review, only one case with similar presentation has been reported [7]. In our case, the uropathy occurred secondary to the mass effect of the massive intra-abdominal CSF-oma with resultant compression on the ureters and the bladder causing outflow obstruction. The atypical presentation, lead to a delay in diagnosis and initiation of appropriate management.

Several etiologies of VP shunt malfunction have been recognized. Along with infections, proximal shunt obstruction, and distal catheter obstruction, CSF-oma has also been recognized as a cause of VP shunt malfunction [8, 9]. Increased intra-abdominal pressure may increase resistance to CSF flow in the VP shunt leading to shunt malfunction.

Chronic inflammatory nature of the ascitic fluid, as illustrated by the analysis of the diagnostic ascitic tap, may suggest low grade chronic infection which may have had a role in the formation of the pseudocyst. Although no organisms were able to be cultured from the aspirated fluid, this may be attributed to early initiation of antibiotic therapy.

Additional techniques have been described to manage CSF-oma in pediatric population including conservative management (antimicrobials, diuretics) and surgical (laparotomy, cyst aspiration, shunt externalization, and conversion to ventriculo-atrial or ventriculo-venous shunt) [10-12].

The diagnosis of CSF-oma can be challenging especially in children with developmental delay and other co-morbidities. In such scenarios, symptoms are often vague and abdominal distention may go un-noticed. It is important to consider CSF-oma in the differential diagnosis of patients with VP shunt who present with progressive abdominal distention and secondary obstructive symptoms and signs of other systems. Although rare, early diagnosis and management can prevent more serious complications. Overall recurrence rate after primary management is 19.4% in children [4]. A laparoscopic approach may prevent the need for shunt externalization or diversion to the vascular system, decreasing associated procedure complications and proving to be more cost-effective, and in addition decreasing the hospital stay.

Diagnosis of a CSF-oma may be complicated with obstructive uropathy, presenting atypically to the health care provider. Laparoscopic approach for managing a CSF-oma may prevent the need for shunt externalization or a diversion to the vascular system, improving cost effectiveness and decreasing hospital stay. In a patient with a VP shunt and progressive abdominal distention, CSF-oma should be considered in the differential diagnosis to ensure early and effective treatment.

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Figure 1: (Pre) Abdominal x-ray of the patient 1 year ago, (Current) x-ray at presentation showing diffuse haziness with centralization of small bowel with pressure effect on the gastrostomy tube

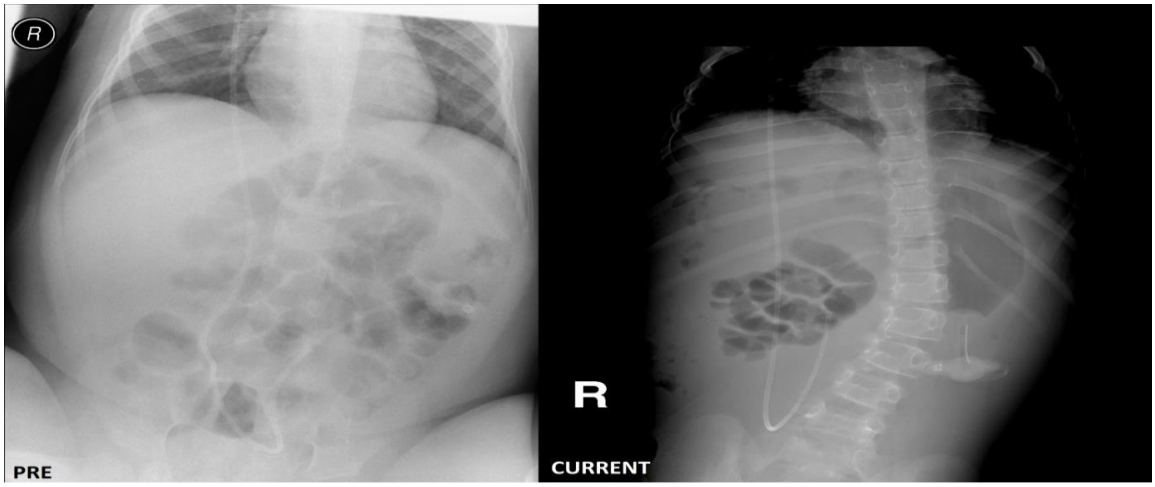
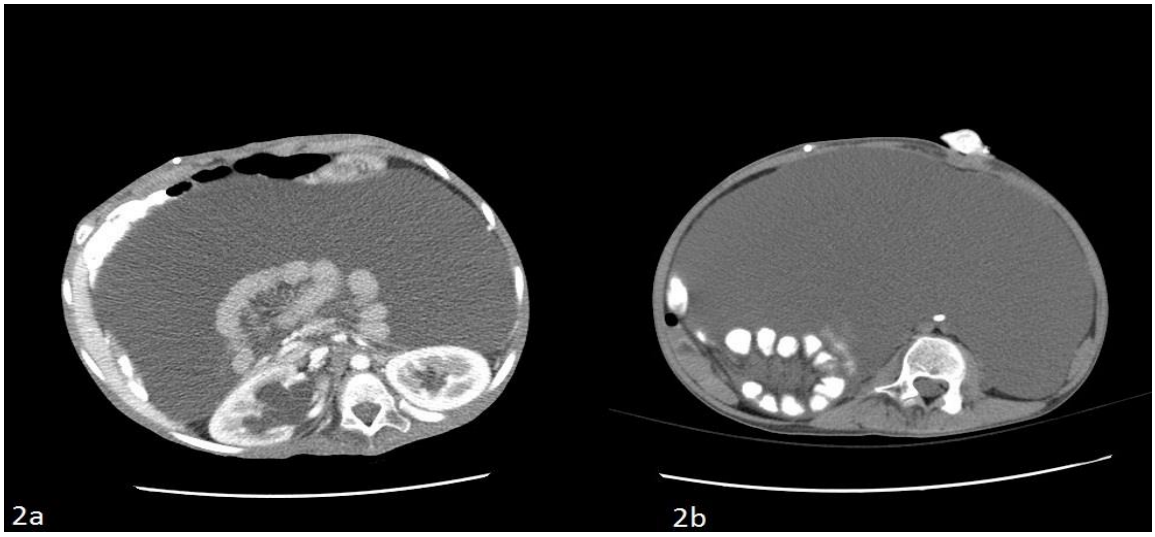


Figure 2: CT scan with contrast demonstrates (a) centralization of the small bowel with bilateral hydronephrosis and (b) displacement of the large bowel to the anterior abdominal wall



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