Intrahepatic cerebral spinal fluid pseudocyst: A case report

Emad R. Al-Sadoon,*
*Ahmed S. Tawfeeq***
**Dept. of Padiatric, College of Medicine, Tikrit University*
***Dept. of Surgery, College of Medicine, Tikrit University*

Case Report

A 5 year old boy presented with shortness of breath, chest pain, and fever for the last few days. He had history of hydrocephalus secondary to aqueduct stenosis diagnosed after birth and ventriculo-peritoneal (V-P) shunt was done for him at age of 6 month. The pregnancy was unremarkable and he delivered by caesarian section at term because of obstructed labor due to large head.

The physical examination revealed a toxic child, ill looking, pale, with large head and he was dyspnic with severe chest retraction but no cyanosis. Vital signs: Temperature is 40 C, Respiratory rate is 55 breath / minute, and blood pressure is 75 / 50. Chest examination revealed limitation of movement at the right side of chest, vocal fremitus is diminished, and dullness at right upper and lower chest with diminished air entry at the right side of chest, the trachea was centrally positioned.

Auscultation of chest shows diminished breath sounds at right side of chest with fine crackles in both lungs. First and second heart sound were normal. Local examination of the valve of the shunt was poorly functioning with indentation lasting more than 5 minutes.

The child was admitted to Tikrit Teaching Hospital (TTH) and send for blood tests, chest X – ray, abdominal ultrasound and CT-scan investigations. The blood investigations was as follow: Hb 10 g/dl ., WBC 15000 / mm^3 . Platelets 150000/l. Blood film: hypochromic microcytic anemia with no abnormal cells.

Liver function test was normal, BUN & Serum creatinine was normal.

Cerebrospinal fluid (CSF) aspiration reveals clear fluid under normal pressure

And shows the following results: cells = 0 cell / hpf , protein 15 mg /dl, sugar 66 mg /dl (blood sugar was 110 mg /dl), and latex agglutination test is normal finding.

Chest X-Ray findings & CT-scan findings as following:

Chest X-ray PA and lateral views showing homogenous opacity occupying middle and lower zones of right hemithorax ,with suspicious elevation of right hemidiaphragm on lateral view raising the possibility of subphrenic collection or hydatid cyst(fig 1). Abdominal ultrasound exam was done, confirming intrahepatic fluid collection (picture not available), later on CT scan exam of upper abdomen was done with coronal and saggital reformatting, revealing intrahepatic localized fluid collection with end of V-P shunt catheter seen at the periphry.(fig2).

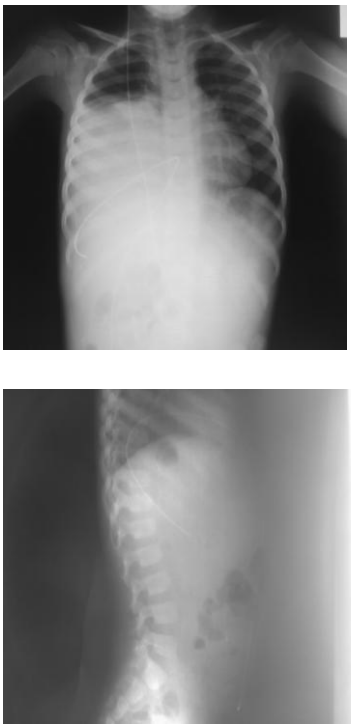
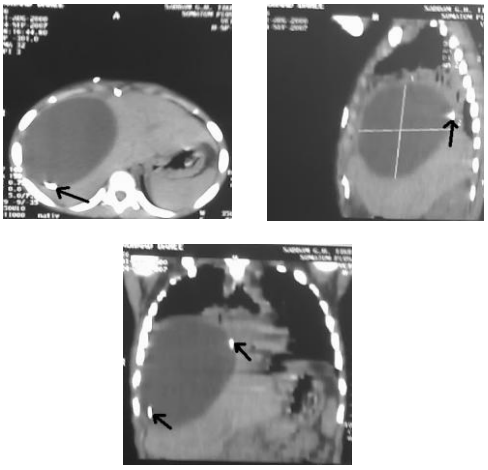


FIG1: (Intrahepatic CSF Pseudocyst)-PA chest X-ray & lateral abdominal films

showing homogenous opacity occupying middle and lower zones of right hemithorax ,with elevation of right hemidiaphragm,with V-P shunt catheter end at the right upper abdomen.



Fig(2) : Multiple axial,sagittal&coronal reformatted CT sections through the liver, showing intrahepatic localized fluid collection(CSF Pseudocyst)with V-P shunt end within(black arrows).

The patient received oxygen therapy, IV fluid, and IV antibiotic for 3 days and after that the diagnosis of this case is ***intrahepatic CSF pseudo cyst*** and we referred the child to Al- Mousl for surgical removal of the cyst and change the VP shunt.

Discussion

Abdominal cerebrospinal fluid (CSF) pseudocyst is an uncommon complication of ventriculo-peritoneal shunts .We report a case of a large intrahepatic CSF pseudocyst in a 5-years-old male operated for congenital aqueduct stenosis 5 years earlier. During the initial surgery a ventriculo-peritoneal catheter was inserted.

Loculated intra-abdominal collections of cerebrospinal fluid are termed CSF pseudocysts or CSF-omas. They may be diagnosed with plain radiographs when bowel loops are displaced or a soft tissue mass is present (1). A shunt normally drains up to 500 ml. of cerebrospinal fluid per day, so a small amount of fluid in the peritoneal cavity may be normal. However, if adhesions develop around the tip of the shunt tube, eventual encapsulation and obstruction will occur (2). Sonography can confirm the presence of these collections. When plain-film signs of fluid loculation are not identified, sonography is very sensitive for the detection of smaller but significant collections. The shunt tube has a characteristic sonographic appearance, typically several parallel echogenic lines. When the shunt tube can be seen within or leading to a localized fluid collection, the diagnosis of a CSF pseudocyst can be made with confidence (3).

Peritoneal shunt tips may migrate to regions outside the peritoneal cavity, such as the thorax via a diaphragmatic hiatus or into the scrotum through a hernia sac. The shunt may also perforate the bowel or other viscera including gallbladder, liver, vagina, bladder and uterus (4,5) .

Shunt complications are reported to occur at a rate of approximately 26 % (1) Diverse complications associated with the peritoneal end of a VP shunt have been described. These include CSF loculation and cyst formation, perforation of the viscera, migration of the shunt, bowel obstruction secondary to adhesions and

metastatic spread via the shunt (2). CSF loculation may present as recurrent ascites, peritoneal cyst, omental cyst subphrenic loculation or intrahepatic loculation as in our case. Abdominal CSF pseudocysts are an uncommon complication and the incidence varies from less than 1% to 4.5% of VP shunts (3,4).

The exact cause of abdominal CSF pseudocysts is still debated. Predisposing factors for pseudocyst formation are multiple shunt revisions, silicon allergy and prior.

Shunt / CNS infection. In a series of 12 patients, Gaskill et al reported that inflammatory process is a frequent predisposing factor.(1) In their series 16% of the patient had acute infection while 41.6% had past history of CNS infection. In a review by Rainov N et al, Microbiologically proven infection was present in 30%

of their cases (4,5).

However, our patient had no objective/subjective evidence of acute infection or past history of CNS infection. Thus infection, while an important factor, is not likely to account for all cases of pseudocyst. Sometimes a pseudocyst may mimic appendicitis. However, the diagnosis

is clearly evident on radiology. The most common presentation of the pediatric patients is with symptoms of elevated intracranial pressure and abdominal pain, whereas the adults have predominantly local abdominal signs.

Improved shunt devices and careful surgical techniques may lead to a decrease but not elimination of the incidence of these complications. A high degree of suspicion and careful clinical and radiological examination could help diagnose and treat CSF Pseudocyst, minimizing major complications.

Ultrasonographic evidence of a larger, localized or loculated collection is abnormal and suggests CSF pseudocyst. CT scanning, which provides accurate localization, has now replaced other imaging modalities as the modality of choice.

The treatment involves exploratory laparotomy followed by surgical removal of the catheter with or without excision of the pseudocyst wall and placement of a new catheter intraperitoneally in a different quadrant or an intra-atrial shunt.

From gaining access to the rest of the peritoneal cavity. Thus a loculated collection of CSF develops. The etiology of this condition is usually low grade sepsis or infection.(6,7,8,9) No satisfactory explanation has been given, to date, for this condition.(10) Infection has been proposed as a causative factor (11).

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