Congenital pancreatic pseudocyst presenting as neonatal ascites

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A B S T R A C T

Congenital pancreatic pseudocysts are extremely rare in infants and very few cases have been reported in the literature. We are reporting a case of 2-month-old infant with pancreatic pseudocyst. He presented with progressive abdominal distension and large bilateral hydrocele. The diagnosis was based on the operative findings and further confirmed by histopathological examination of the wall of pseudocyst.

Later on the patient was operated by laparoscopic assisted transgastric cystogastrostomy technique. To the best of our knowledge, this is the first case of pancreatic pseudocyst presenting as neonatal ascites and managed by laparoscopic assisted transgastric cystogastrostomy technique. We are going to discuss the diagnostic dilemma and therapeutic modalities.

1. Case presentation

A 2-month-old male infant admitted to the hospital for the complaint of progressive abdominal distension and large bilateral hydrocele. Regarding the birth history it was an uncomplicated pregnancy, full term and delivered by SVD and with the birth weight of 3.5 kg. The postnatal period was unremarkable and discharged home immediately without any nursery stay. Later on he developed increasing abdominal distension since one month and along with it there was gradual enlargement of the scrotum for the last two weeks. Moreover, there is associated complaint of non-bilious vomiting since birth and usually two to three episodes per day. However, there is no history of constipation or bowel disturbance and fever.

At the time of presentation the patient was stable and with normal vital signs: Temperature 37.5 °C, pulse 129/min, blood pressure 85/48 mm/Hg and the body weight 5.1 kg. The physical examination revealed markedly distended, non-tender abdomen and with a positive shifting dullness. Also a large bilateral communicating hydrocele was noted. Laboratory results showed: Hb 10.2 g/dl, platelets 1179, WBC 6.3 × 10^9/L, neutrophils 40% and lymphocytes 39.7%.

The patient was initially admitted with the diagnosis of ascites and for further investigation.

The abdominal US showed a well-defined cystic mass in the left upper quadrant measuring 4.6 × 4.5 cm and with marked ascites. It was followed by CT scan which demonstrated a well-defined 5.1 × 5.7 cm circular cystic lesion in the left hypochondrium adjacent to the tail of the pancreas. The cyst had a wall calcification without intra-lesional soft tissue component. Also there was a gross abdominopelvic ascites extending through patent processes vagnalis to the scrotum (Fig. 1). Diagnostic ascitic tap done and peritoneal fluid analysis revealed: albumin 21 g/L, amylase 71 u/L, lipase >1200 u/L, triglyceride 0.81 mmol/L, creatinine 25.6 mg/L, and the culture was negative.

Laparoscopic exploration revealed a cystic mass at the anatomical site of the tail of the pancreas, pushing the stomach anteriorly and bulging between the greater gastric curvature and
the transverse colon, which was pushed inferiorly. The surrounding tissue was edematous and erythematous with the tendency to bleed on touch by the instruments (Fig. 2). These intraoperative findings confirmed the diagnosis of pancreatic pseudocyst. The procedure was continued by opening the anterior gastric wall while exposing the posterior wall of the stomach which was bulging due to the mass effect of the cyst. Then mini-laparotomy done at the site of anterior gastric wall incision and the posterior wall of the stomach opened in order to get access to the cystic cavity, which in turn was filled with fluid and lined by a pseudopyogenic membrane. The membrane was removed and transgastric cystogastrostomy was performed. Thereafter, the anterior gastric wall and abdominal wall were closed.

Histopathological examination of the cyst revealed retained secretory material and acute inflammation without epithelial lining, which was consistent with pseudocyst. Furthermore, fragments of gastric wall with ulceration and chronic inflammation were noted.

Post-operative period was uneventful and oral feeding resumed on the 8th post-operative day. Follow up abdominal US done before discharge and showed marked decrease in both the ascites and the size of the cyst as well. At six and twelve months follow up, the child was found well and there was no clinical or laboratory signs of pancreatic insufficiency and CT scan did not demonstrate any residual cystic cavity.

2. Discussion

Pancreatic cysts in children are classified as congenital-developmental cysts, retention cysts, duplication cysts, pseudocysts, neoplastic cysts, and parasitic cysts [2]. Cystic lesions of the pancreas in the pediatric population are rare entities and the true incidence of this clinical entity is not known, 25

![Computed tomographic scan demonstrates the cyst at the bed of the pancreas with severe ascites.](image1)

![Laparoscopic view of the pseudocyst bulging between the greater gastric curvature of the stomach and the transverse colon which is pushed inferiorly.](image2)
cases were collected by Boulanger et al. [3]. In children under the age of 2 years only individual cases were reported [1,4–6].

Congenital pancreatic pseudocyst is an unusual phenomena and only five cases of congenital pancreatic pseudocyst were reported in the literature [7,8].

Khalifa et al reported a series of 19 children with pancreatic pseudocysts. One case in this series was mentioned as congenital pancreatic pseudocyst, who was one-month-old [8].

Congenital pancreatic pseudocysts are usually asymptomatic. The clinical presentation in our patient was due to huge abdominal distension and communicating hydrocele as a result of the severity of ascites.

The diagnosis of pancreatic cyst is usually done by radiological imaging. In our case, both the radiological as well as laboratory findings helped in the diagnosis. As the CT scan demonstrated the cyst lying at the bed of the pancreas and in addition to it the high lipase level in the aspirated ascetic fluid indicated that the cyst has a pancreatic origin. But the final diagnosis of pseudocyst was evident by laparoscopic exploration, and confirmed later on by histopathological examination of the wall of the cyst.

The congenital origin of the pseudocyst in our case was based on the fact that the symptoms started in the neonatal period and without any preceding perinatal or postnatal trauma or other features of pancreatitis or intra-uterine viral infection. However, the exact etiology remains unclear in our case like the cases reported by Kurrer [7].

The pancreatic pseudocysts are unresectable lesions. Therefore, in order to avoid injuries to the surrounding structures or any possible complication, the treatment of these lesions is achieved by internal drainage procedure cystogastrostomy or cystojejunostomy [8,9].

Endoscopic drainage of pseudocysts is becoming the preferred therapeutic approach in adults because it is minimal invasive. However, there is no reported experience found in small infants due to the fact that internal drainage is accomplished with transpapillary approach with ERCP [10], which is difficult to be done in small infants. Also due to rarity of the entity, all the reported cases have been diagnosed peroperatively.

Nowadays laparoscopic cystogastrostomy has been used in adult and elderly children. In a five-institution survey, Yoder SM et al., concluded that a laparoscopic approach toward pancreatic pseudocysts by cystgastrostomy has been proved to be safe and effective [11].

In our small infant patient, the assisted laparoscopic technique proved to be of great help in demonstrating the location of cyst and confirming the diagnosis of pseudocyst. Furthermore, it minimized the laparotomy incision, reduction in the manipulation of abdominal viscera and shortened the post operated period. However, a full laparoscopic transgastric cystogastrostomy is performed in elderly age [11].

In conclusion, to the best of our knowledge this is the first reported case, in which there is unusual presentation of congenital pancreatic pseudocyst in early infant life, with severe ascites and dealt successfully with minimal invasive approach.

Although congenital pancreatic pseudocysts are rarely seen, it should be included in the differential diagnosis of children presenting with cystic abdominal masses with ascitis. And the surgeon should be prepared to perform internal drainage when the diagnosis is made peroperatively.

References