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Neonatal congenital pancreatic cyst: diagnosis and management

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Abstract Congenital pancreatic cysts are extremely rare in newborns. The case of a neonatal congenital pancreatic cyst with pancreatitis is reported. The rapid increase in cyst size concomitantly with clinical symptoms prompted surgical management. Intraoperatively, the pancreatic head showed signs of chronic pancreatitis, and in attempts to preserve most of the functional pancreatic tissue, a Roux-en-Y cystojejunostomy was performed. Histology demonstrated a true pancreatic cyst with degenerated epithelial lining.

Pancreatic cysts are rare lesions in newborns, with inflammatory or traumatic pseudocysts contributing to most of the reported cases [1]. A 2140-g full-term female neonate was diagnosed by prenatal ultrasounds to have an upper abdominal cyst. The cyst persisted in the postnatal ultrasounds, but the origin could still not be predicted. Because the newborn showed no clinical symptoms, she was discharged and scheduled for routine follow-ups. At the age of 10 days, the newborn presented with abdominal distension, pain, and anorexia. Because of the presence of clinical symptoms, an ultrasound-guided puncture of the cyst was performed, and 380 mL of clear yellow fluid was aspirated. Enzymatic analysis showed increased amylase (2771 U/L) and lipase (>6000 U/L) in the aspirate. A marked reduction in pain was observed after drainage. Two days later, abdominal distension recurred, and ultrasounds confirmed refilling of the cyst, which was again aspirated (130 mL). At this stage, the patient was referred to our center.

At the time of admission, a mild abdominal distension was observed, and the laboratory parameters showed an increase in white blood cells (17.8×10^9/L; n = 4.4-11.3 $\times 10^9$/L), normal C-reactive protein, elevated gamma-glutamyl-transferase (GGT) (259 U/L; n < 216 U/L), and elevated lipase (671 U/L; n < 200 U/L). Alkaline phosphatase and amylase were normal. Ultrasounds demonstrated a distended cyst (Fig. 1). At laparotomy, a solitary large retroperitoneal cyst originating from the middle of the pancreatic body was identified. The pancreatic head showed areas of calcification, which correlated with signs of chronic pancreatitis. The cyst wall contained islands of pancreatic tissue (with a diameter of approximately 5 mm) with the pancreatic tail appearing to be normal. At this time, the cyst was punctured, and a clear

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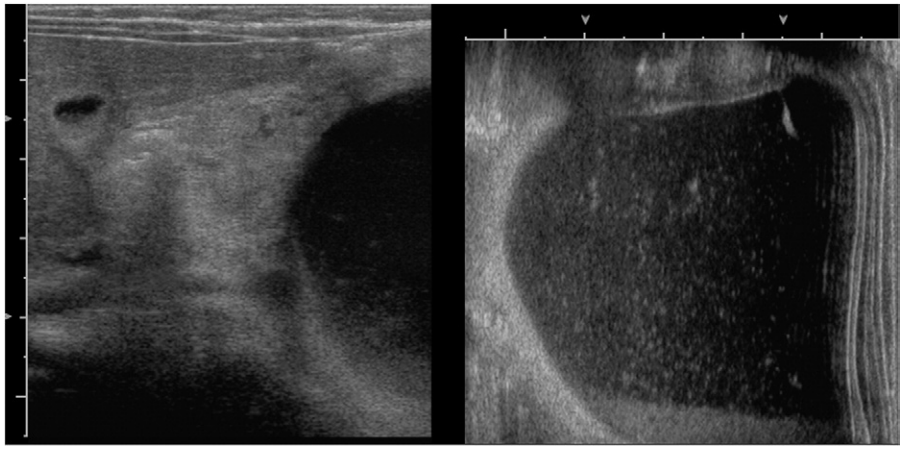

Fig. 1 Abdominal ultrasound after referral of the patient. Cyst with sedimentation located just below the liver.

yellow cystic fluid (65 mL) was aspirated. Contrast medium was instilled into the cyst, whereby no communication of the cyst with the rest of the pancreas could be established. Because the cyst originated from a broad pancreatic base (Fig. 2A), a resection was not possible. Therefore, internal drainage was opted through a side-to-side Roux-en-Y cystojejunostomy (Fig. 2B) (with a Roux limb of 10 cm).

On the first postoperative day, signs of abdominal distension were observed once again, and ultrasound examinations confirmed refilling of the cyst. A revisions laparotomy had to be performed, and the filled cyst was punctured and aspirated. The aspirated fluids were bile tinged, indicating biliary reflux from the Roux limb. The short Roux limb of 10 cm was responsible for the refilling, and to overcome this, the Roux limb was elongated to 45 cm. At this time, the cyst was partially resected, and a drain was placed in the bursa omentalis. Histology of the cyst wall is shown in Fig. 3.

Postoperatively, serum lipase was elevated (7683 U/L), but amylase remained normal. The cystic drain fluid had high levels of lipase (255.170 U/L), but again low amylase (962 U/L). These parameters returned to normal within the next 3 weeks. An magnetic resonance cholangio pancreaticography (MR-CP) showed normal diameters of choledochal and pancreatic duct and no signs of the cyst refilling. The patient was well over the further course and could be discharged after 30 days of hospital stay.

1. Discussion

Clinically, the patient in this report initially presented with signs of pancreatitis, which is rarely reported in cases with pancreas cysts; frequently because normal levels of amylase and lipase have been described [2], and only some exceptions reported [3-5]. Although it is unclear, it is speculated that pancreatitis in these cases results either from congenital anomalies of the duct system [6] or from visceral compression by the cyst [4].

In the postoperative course of the patient reported, the symptoms ceased albeit elevated serum lipase. In an almost similar fashion, an asymptomatic patient with high enzymes and discrepancies between amylase and lipase

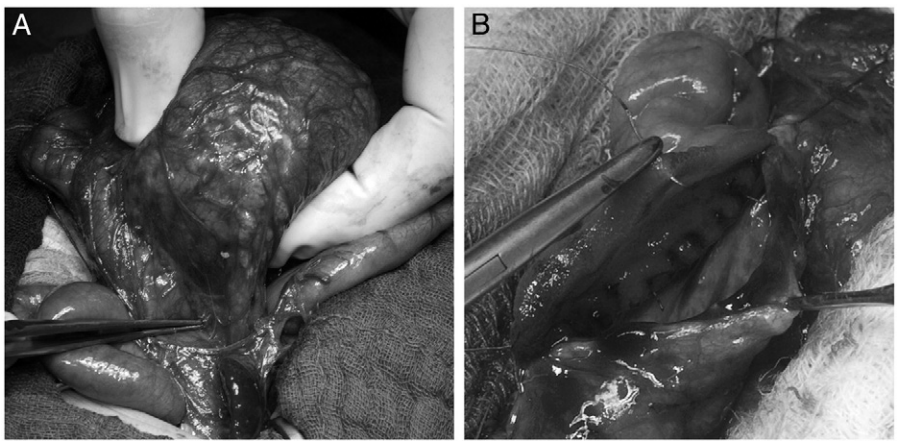

Fig. 2 Operative view of the pancreatic cyst. A, The broad based origin of the cyst from the pancreatic body is seen. The tip of the forceps points to the beginning of the pancreatic tail. B, Side-to-side Roux-en-Y cystojejunostomy. The cyst wall is seen held by the forceps.