

## **Case report**

### **Congenital cyst of pancreas in a newborn: A rare presentation**

#### **Abstract**

Pancreatic cysts are rare lesions in newborns. Prenatal diagnosis of true congenital pancreatic cyst is rare and many times cystic lesion on prenatal scans confused with mesenteric cyst, ovarian cyst, and left renal cyst. Here we report a newborn presented with antenatally detected upper abdominal cystic lesion. The cystic mass was located in the left upper side of the abdomen, and total cystectomy was performed without any intra or post-operative complication. Histopathological examination demonstrated presence of pancreatic tissues in the wall of cyst with foci of inflammation and cystically dilated ducts, lined by flattened epithelium. Features suggestive of congenital true pancreatic cyst with pancreatic fluid as its predominant content.

Keywords: Congenital Pancreatic Cyst, True Pancreatic Cyst, Serum Amylase, Serum Lipase, Chronic Inflammation

## **Introduction**

True congenital pancreatic cysts in newborn period are most unusual and rare.[1] Due to their extreme rarity, they are difficult to diagnose antenatally as well as postnatally and may be confused with other types of cystic lesions.[2] The lesion is usually small and lined with true epithelium, and can be solitary or multiple. A complete excision is considered to be the best treatment option for patients with these cysts. In this article we report a newborn girl with histologically proven true pancreatic cyst that was detected by prenatal ultrasonography. The cystic mass was located in the left upper side of the abdomen, and total cystectomy was performed without complication. Histology proved congenital true pancreatic cyst with high levels of pancreatic enzymes in the cystic fluid.

## **Case Report**

A 2.5 kgs full-term female neonate with history of antenatally diagnosed upper abdominal cyst in third trimester ultrasound. The cyst persisted in the postnatal period as suggested by ultrasound on day-4 of life.

Newborn showed no clinical symptoms till the age of day-8 of life after which she developed abdominal distension and infrequent episodes of non-bilious vomiting. A palpable mass in upper abdomen was found during a routine physical examination. Her complete blood cell count, electrolytes, blood urea nitrogen, creatinine, liver function tests, and amylase were normal. Abdominal ultrasound revealed a large unilocular non-septated distended cystic swelling measuring 8 × 6.5 cms seen in the midline anterior to the spine and extending to the left side, with few internal echoes (Fig. 1.A).

Abdominal computed tomography (CT) scan showed unilocular thick wall cystic swelling about 7.8 × 7.7 × 6.7 cm seen extending from the anterior abdominal wall up to the left anterior pararenal space posteriorly.[Fig.1.B] The stomach was displaced medially, whereas the left kidney was compressed and displaced posteriorly by this mass. The body and tail of the pancreas were compressed, but tissue interfaces between the cystic mass were observed. A preliminary diagnosis of enteric cyst, mesenteric cyst, and pancreatic cyst was made.

She underwent exploration laparotomy, which revealed a large cystic swelling occupying most of the left side of the abdomen and extending to the midline displacing the stomach as well as transverse colon over it. It had a thick wall and contained large amount of light yellow fluid. The cyst was dissected until it was completely removed. It was attached closely to

the body of the pancreas with a stalk of tissue that was transfixed, tied, and cut (Fig. 2). Fluid analysis was sent, that revealed very high levels of amylase (435U/L) and lipase (342U/L). Histopathology revealed presence of pancreatic tissues in the wall of cyst with foci of inflammation and cystically dilated ducts, lined by flattened epithelium.(Fig.3) Features suggestive of congenital true pancreatic cyst with pancreatic fluid as its predominant content. Postoperative course was uneventful and she was discharged home on the seventh postoperative day.

## **Discussion**

True pancreatic cysts are rare lesions in pediatric population. Antenatally detected pancreatic cystic lesions are extremely rare. [3-5]. These account for less than 1% of all pancreatic cysts, and they are often difficult to distinguish from one another. Congenital cysts are found preferentially in the neck or tail (62%) and the head (32%) of the pancreas, or can be diffusely involving the whole pancreas (6%) [3]. These growths are typically small in diameter (2–3 cm), but they may grow to great size in some cases. It is believed that these cysts are caused by anomalous development of the pancreatic ductal system wherein sequestered segments of a primitive secretory ductal system

give rise to microscopic or macroscopic cystic lesions.[6] Congenital cysts of the pancreas may be encountered in the fetus, infant, child, or adult.

Clinically, the antenatal cases presented incidentally or with polyhydramnios.[7] Postnatally these congenital cysts may have varied presentation including an asymptomatic abdominal mass, abdominal distension, vomiting caused by gastric mass effect, and jaundice from biliary obstruction.[8,9] Pathologically, congenital pancreatic cysts are more often unilocular than multilocular, are more often single than multiple and are more frequently located in the pancreatic body or tail than in the pancreatic head.[1,6] Congenital pancreatic cysts may or may not possess an epithelial lining or elevated amylase levels; the presence of either supports the diagnosis. Microscopic examination may show pancreatic acini or ductules between the cystic locules.

Surgical treatment consists of total excision for cysts located in the pancreatic body or tail. Internal drainage procedures may be required for the lesions located in the head region. If the tumor is located at the head of the pancreas, drainage of the mass via a cystoduodenostomy or a Roux-en-Y cystojejunostomy may be preferable [10] Pre-operative analysis of the cyst fluid contents may be helpful in defining the nature of the cyst.

Although congenital cysts of the pancreas are extremely rare, they should be considered as a differential diagnosis for a pancreatic cyst, especially in asymptomatic children.

## References

1. Auringer ST, Ulmer JL, Summer TE, Turner CS. Congenital cyst of the pancreas. *J Pediatr Surg* 1993; 28:1570-1571.
2. Agarwala S, Lal A, Bhatnagar V, Dinda AK, Mitra DK. Congenital true pancreatic cyst: presentation and management. *Tropical Gastroenterology* 1999; 20:87-88.
3. Fremond B, Poulain P, Odent S, et al. Prenatal detection of a congenital pancreatic cyst and Beckwith-Wiedemann syndrome. *Prenat Diagn* 2006;17:276-80.
4. Kebapci M, Aslan O, Kaya T, et al. Prenatal diagnosis of giant congenital pancreatic cyst of a neonate. *AJR Am J Roentgenol* 2000; 175:1408-10.
5. Sepulveda W, Carstens E, Sanchez J, et al. Prenatal diagnosis of congenital pancreatic cyst: case report and review of the literature. *J Ultrasound Med* 2000;19:349-52.

6. Cotran RS, Kumar V, Robbins SL: Robbins Pathologic Basis of Disease (ed 4). Philadelphia, PA, Saunders, 1989, p 989
7. Baker LL, Hartman GE, Northway WH: Sonographic detection of congenital pancreatic cysts in the newborn: Report of a case and review of the literature. *Pediatr Radio* 20:488-490.1990
8. Miles RM: Pancreatic cyst in the newborn: A case report. *Ann Surg* 149:576-581,1959
9. Pilot LMJR. Gooselaw JG, Isaacson PG: Obstruction of the common bile duct in the newborn by a pancreatic cyst. *Lancet* 84:204-205,1964
10. Hammel P, Levy P, Voitot H, et al. Preoperative cyst fluid analysis is useful for the differential diagnosis of cystic lesion of the pancreas. *Gastroenterology* 1998;108(4):1230–1235.

Fig.1.A ultrasound revealed a large unilocular non-septated distended cystic swelling seen in the midline anterior to the spine and extending to the left side, with few internal echoes (blue

arrow). Fig.1.B Abdominal computed tomography (CT) scan showed unilocular thick wall cystic swelling seen extending from the anterior abdominal wall up to the left anterior pararenal space.

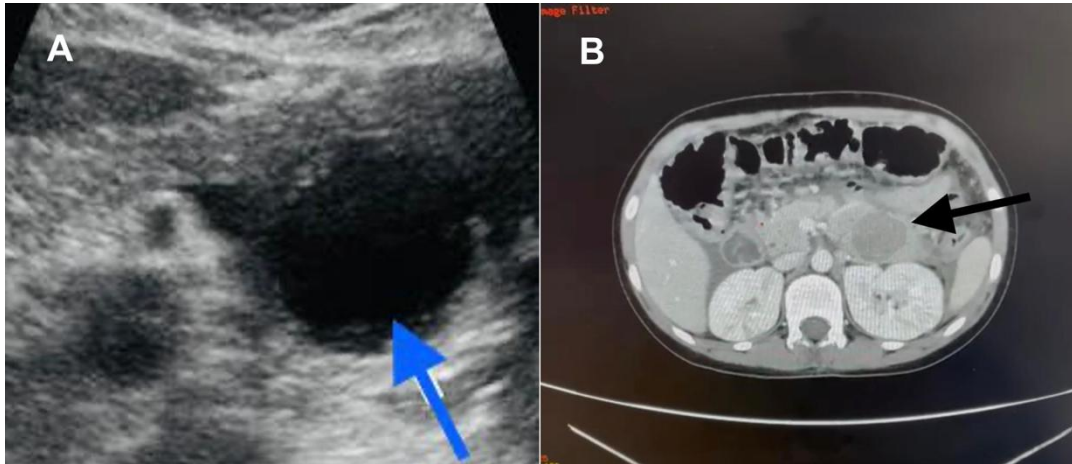


Figure.2 Intra-operative findings. A huge, tense, cystic lesion attached to the body of pancreas and occupied most of the

abdominal cavity. Note the displaced colon and stretched duodenum

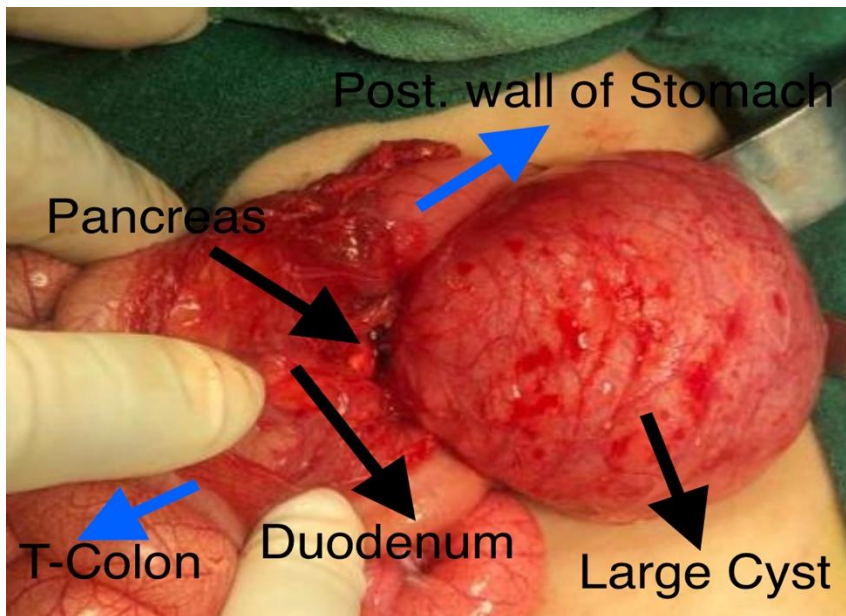
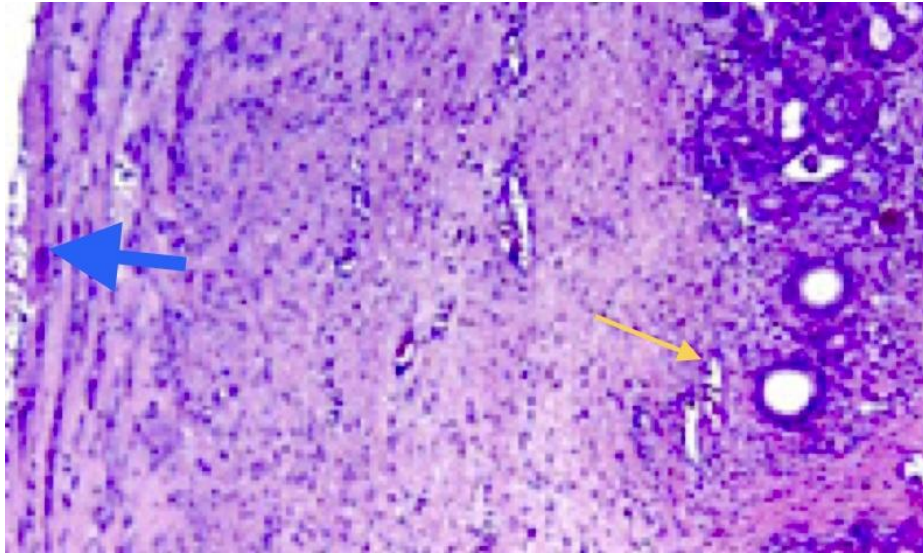


Figure.3 Thin yellow arrow depicting Cyst wall with embedded pancreatic tissue (H&E stain). Solid blue arrow showing prominent signs of chronic pancreatitis



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