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Congenital Cyst of Pancreas in a Newborn: A Rare Presentation

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Authors' contributions

This work was carried out in collaboration between both authors. Both authors read and approved the final manuscript.

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Case Report

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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.

1. 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

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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. Total cystectomy was carried out without incident on the cystic mass, which was found in the left upper region of the abdomen. With significant levels of pancreatic enzymes in the cystic fluid, histology demonstrated a congenital genuine pancreatic cyst.

2. CASE REPORT

A 2.5 kgs full-term female neonate with history of antenatally diagnosed upper abdominal cyst in third trimester ultrasound. Antenatal period of pregnancy was uneventful, with a single prenatal ultrasound in last trimester, which was showing cystic lesion measuring ~ 7.2 x 6.4 cms in upper abdomen. There was no history of delayed passage of meconium or abdominal symptoms at birth. The baby appeared active and was feeding fine. The cyst persisted in the postnatal period as suggested by ultrasound on day-4 of life, till that time baby was in newborn nursery. A mass was palpable in upper abdomen during a routine physical examination after birth. 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. 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 left kidney was squeezed and displaced posteriorly, while the stomach was moved medially. Although the pancreas's body and tail were squeezed, tissue contacts between the cystic mass were visible. Pancreatic, mesenteric, and enteric cysts were tentatively identified.

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

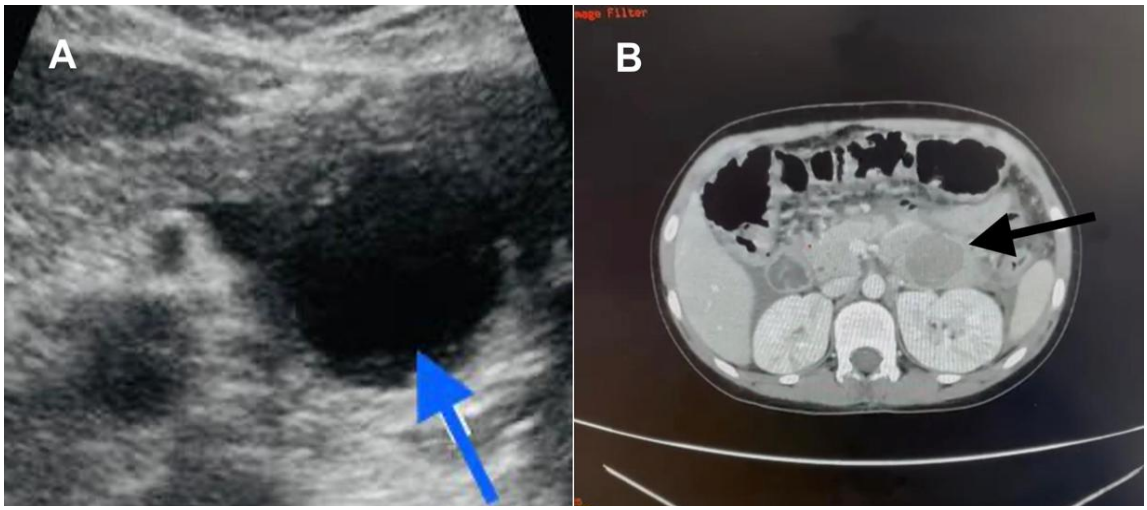

Fig. 1A. Ultrasound revealed a large unilocular non-septated distended cystic swelling seen in the midline anterior to the spine and extending to the left side, with few internal echoes (blue arrow)

Fig. 1B. 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

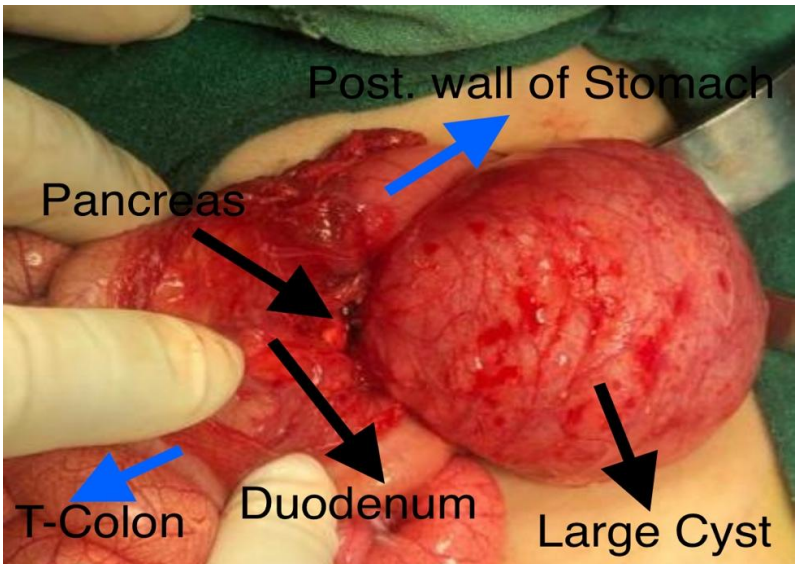

Fig. 2. Intra-operative findings. A huge, tense, cystic lesion attached to the body of pancreas and occupied most of the abdominal cavity. Note the displaced colon and stretched duodenum

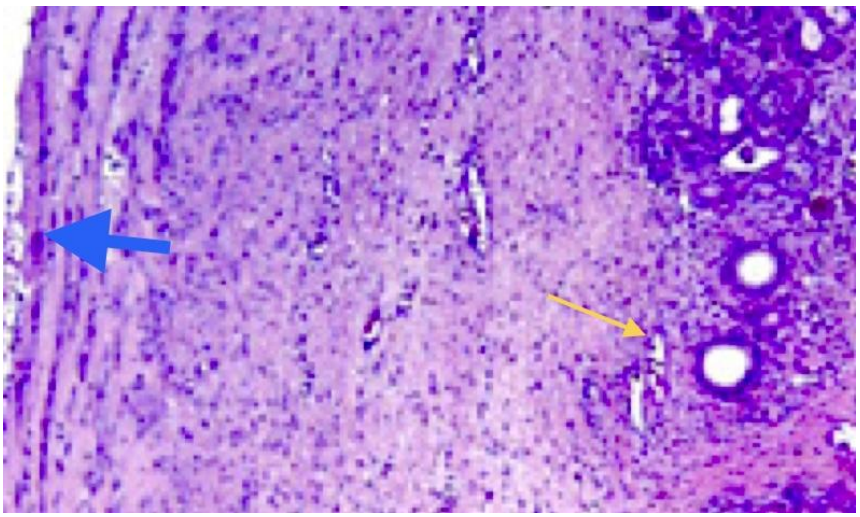

Fig. 3. Thin yellow arrow depicting Cyst wall with embedded pancreatic tissue (H&E stain). Solid blue arrow showing prominent signs of chronic pancreatitis

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.

3. 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]. Pancreatic congenital cysts can develop in a fetus, newborn, 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 [8,9,10]. 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]. Elevated amylase levels and an epithelial lining may or may not be present in congenital pancreatic cysts, but their presence confirms the diagnosis. Pancreatic acini or ductules may be visible under a microscope 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 [11] Pre-operative analysis of the cyst fluid contents may be helpful in defining the nature of the cyst. The prognosis of these cysts is generally good and all attempts should be made to preserve most of the functional pancreatic tissue [12].

4. CONCLUSION

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. When cystic lesions are seen during a regular prenatal ultrasound, this unusual diagnosis should be taken into account. A planned surgical approach in the neonate with the goal of entire excision of the cyst with preservation of the pancreas is justified by the benign nature of these cysts as well as a certain risk for problems when left in place.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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