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# Congenital pancreatic cyst with Ivemark II syndrome: a rare case

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**Abstract** An infant with congenital pancreatic cyst with Ivemark II syndrome is reported because it is a rare association. The infant had associated situs inversus, asplenia, and complex congenital heart disease. The pancreatic cyst was successfully managed by cystoduodenostomy because of connection to the biliary tract. The infant succumbed as a result of heart failure at age 2 months. Prognosis depends on the presence of life-threatening malformations.


Congenital pancreatic cyst (CPC) is a rare condition in infants and can be isolated or associated with other malformations. We report the case of a CPC with Ivemark II syndrome, which was associated with situs inversus, asplenia, and complex heart disease.

## 1. Case report

A 45-day-old full-term male twin was born after normal pregnancy and labor. The birth weight was 2.3 kg. Apgar score at 1 and 5 minutes was 7 and 9, respectively. The mother was G4P3A0 with no significant family history. The second twin was a healthy male. The patient was hospitalized in the pediatric department at day 13 of life for vomiting, hypothermia, altered general health (weight loss and hypotonia) with leukocytosis (white blood cell count, 33 000/mm$^3$), and a high C-reactive protein level (32 mg/L). A postnatal infection was suspected and treated with antibiotics (a combination of cefotaxime and aminoside).

The onset of illness was marked by abdominal distension with bilious vomiting during oral feeding. On physical examination, a mobile smooth mass was palpated on the left upper abdomen.

A plain thoracoabdominal radiograph showed dextrocardia and a left abdominal opacity without calcifications (Fig. 1). An abdominal ultrasonography study showed an anechoic cystic mass measuring 7 × 6 × 6 cm, occupying the left abdomen, against the anterior wall of the stomach, which was displaced to the right side of the abdomen along with the left kidney and liver. An abdominal computed tomographic (CT) scan revealed the presence of this cystic mass and asplenia (Fig. 2). Echocardiography

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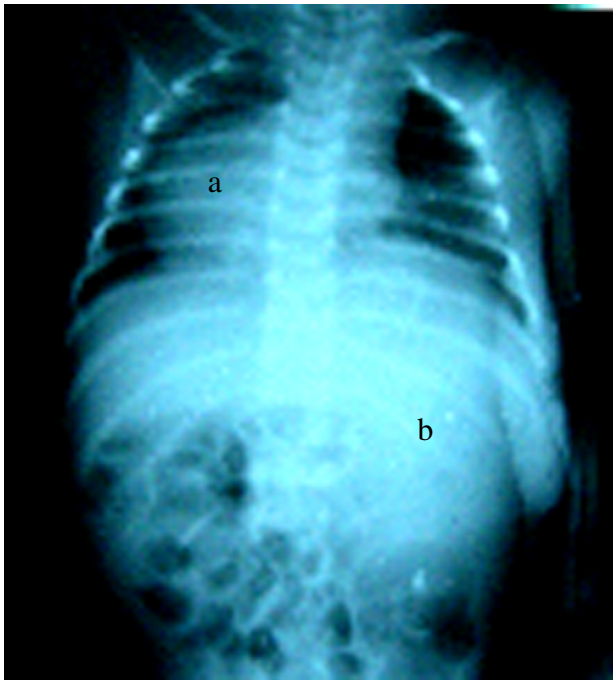


**Fig. 1** Thoracoabdominal x-ray. a, Dextrocardia. b, Left abdominal opacity.

showed a complex congenital heart disease with single ventricle and pulmonary atresia.

The differential diagnosis included digestive tract duplication, choledochal cyst, and pancreatic cyst. At laparotomy, a large cyst with a vascularized wall in contact with the gastric antrum, pylorus, duodenum, biliary ducts, and the pancreas was identified (Fig. 3). Aspiration of the cyst returned bilious content suggesting cystic communication with the biliary tract. Intestinal malrotation with an ectopic development of the pancreas, asplenia, and a dysplastic liver was also observed during the surgical procedure. A total

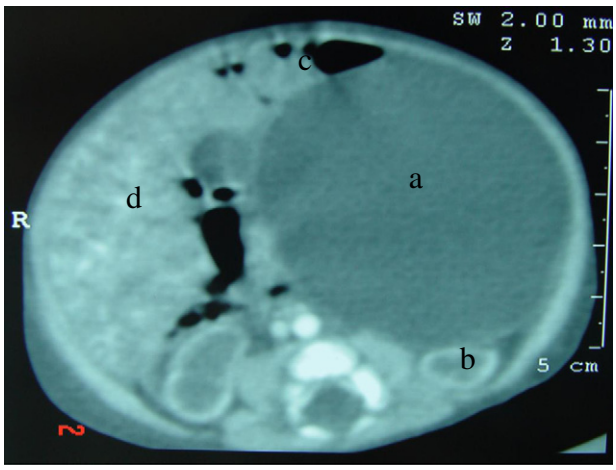


**Fig. 2** Abdominal CT scan. a, Cystic mass. b, Left kidney. c, Intestine. d, Liver.

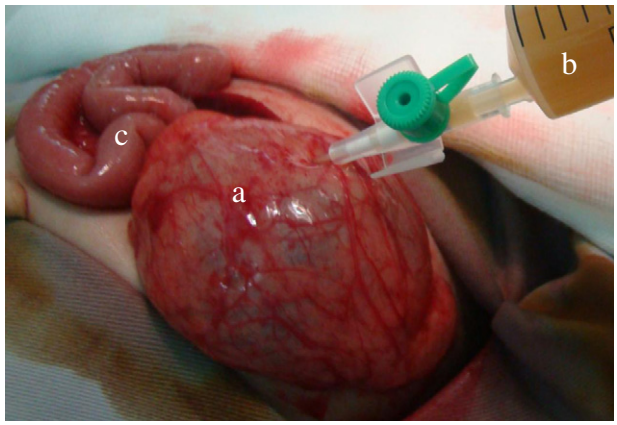


**Fig. 3** Surgical exploration of the cystic mass (cyst aspiration). a, Pancreatic cyst. b, Aspirated liquid. c, Intestine.

cystectomy was not possible because of the intimate contact with the biliary ducts, pancreas, duodenoantropyloric structures, and the right kidney. Thus, cyst aspiration and an internal drainage procedure (side-to-side cystoduodenostomy) were performed.

On microscopic examination, the cystic wall was fibrous and contained focal pancreatic acini and ducts. The wall was lined with a completely abraded epithelium (Fig. 4). The biochemical analysis of the cystic fluid revealed a bilirubin level of 132 $\mu$mol/L (normal serum value, 5-17 $\mu$mol/L) and a lactic dehydrogenase level of 4998 UI/L (normal serum value, 180-640 UI/L). Amylase and lipase levels were not performed because of technical problems.

The association of hepaticopancreatic dysplasia with asplenia and complex heart disease suggested an associated Ivemark II syndrome with this CPC.

The initial postoperative course was uneventful, and enteral feedings was started 6 days after surgery. The

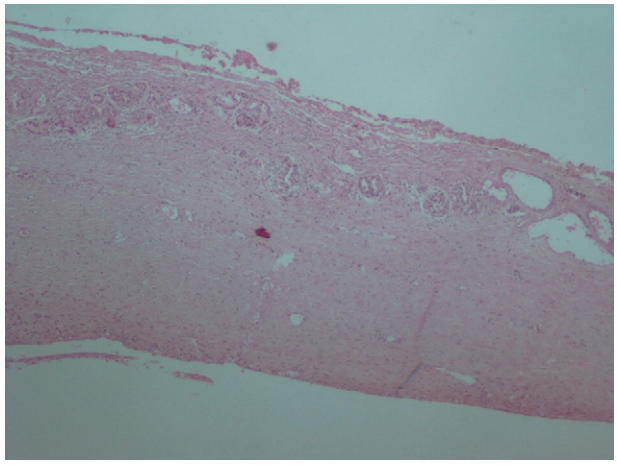

**Fig. 4** Histology: cystic wall made of a fibrous tissue containing pancreatic acini grouped in lobules (H&E, original magnification ×40).