A congenital cyst of the pancreas

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Cystic lesions of the pancreas are relatively rare and are commonly seen in adults. Among these, pseudocysts, either post-traumatic or post-inflammatory following acute pancreatitis, are the commonest.¹ True congenital pancreatic cysts on the other hand are extremely rare.^{2,3,4,5,6} This report describes a 4-month-old female infant with a large congenital cyst of the pancreas. The literature on the subject is also reviewed.

Case

A 4-month-old female infant, a product of a full-term normal vaginal delivery with a birth weight of 3.2 kg was referred to our hospital because of intermittent vomiting and abdominal distension. The vomiting started immediately after birth and was mostly not bile stained. Abdominal distension was noticed only two weeks prior to presentation. Clinical evaluation revealed a large, smooth, non-tender and slightly mobile abdominal swelling. The remainder of the examination was unremarkable. Laboratory tests found a normal CBC, blood urea, glucose, electrolytes and amylase. A plain abdominal x-ray film showed a large soft-tissue density mass involving most of the abdomen and pushing all bowel loops to the left side. A CT scan of the abdomen showed a large, single cystic mass with well-defined margins involving most of the cross-sectional area of her abdomen and measuring about 13.5 cm in diameter (Figure 1). Both kidneys were normal and no definite pancreas was identified. The possibility of a mesenteric or a duplication cyst was entertained. During laparotomy, there was a large cystic swelling occupying most of the abdomen with the bowel pushed to the left side. The duodenum was found stretched around the right side of the cystic wall (Figure 2). The cyst was found arising from the head and body of the pancreas. Aspiration of the cyst was made and about 1100 ml of clear fluid was aspirated. The amylase content of the fluid was later found to be high (>5000 IU/mL). The cyst was drained by a Roux-in-Y cysto-jejunostomy. Biopsy of the cyst showed a cyst wall lined by cuboidal epithelium with a mild inflammatory infiltrate. Postoperatively, the patient did well and was discharged home 3 weeks later. At 15 months follow-up, she was found well, asymptomatic and gaining weight.

Discussion

True congenital cysts of the pancreas are extremely rare. In 1958, Miles reported a case of congenital pancreatic cyst with a review of the literature that described only eight other cases.² Auringer et al in 1993 reviewed the literature and found only 21 reported cases of congenital pancreatic cyst.³ In a literature review in 2003, Boulanger et al found only 25 cases of congenital pancreatic cyst.⁵ The exact pathogenesis of congenital cyst of the pancreas is not known, but it is believed that these cysts are caused by anomalous development of the pancreatic ductal system wherein a sequestered segment of a primitive secretory

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Figure 1. CT-scan showing a large abdominal cystic swelling.

Figure 2. Intra-operative photograph showing a large cystic swelling arising from the head of the pancreas. Note the loop of the duodenum stretched around the right border of the cyst.

ductal system gives rise to microscopic or macroscopic cystic lesions.³ The cysts more often are single than multiple, but occasionally are seen in association with other diseases such as Von Hippel-Lindau syndrome or polycystic kidney disease, particularly when multiple.^{1,6} They can present at any age from as early as in utero to as late as 62 years, but the majority are diagnosed before the age of 2 years.^{3,7,8,9}

The clinical presentation of these cysts is variable. They may be found on routine physical examination or as an incidental finding during evaluation for other complaints. Occasionally, they may be symp-

tomatic, causing abdominal pain, vomiting, and abdominal distension or may be complicated causing pancreatitis, obstructive jaundice or splenic vein thrombosis.^{2,4,10}

Although there are reports of in utero diagnosis of congenital pancreatic cysts, these cysts are difficult to diagnose preoperatively.^{7,8,9} The differential diagnosis includes duplication and mesenteric cysts, lymphatic cyst, choledochal cyst and renal cyst. In 1977, Mares and Hiresch reported the first accurate preoperative diagnosis of a congenital pancreatic cyst in a 20-month-old female infant by means of a celiac and gastroduodenal angiogram.⁴ A high amylase content of the cyst and cuboidal epithelial lining confirmed the diagnosis of a true congenital pancreatic cyst. This, however, is not always the case as inflammation and infection of the cyst can destroy the epithelial lining and interfere with enzyme secretion.^{3,5}

The treatment of congenital pancreatic cyst is variable depending on its location. The cysts can be unilocular or multilocular and are more frequently located in the pancreatic body or tail than in the pancreatic head.³ In general, whenever feasible, complete surgical excision is the treatment of choice, in particular for cysts located in the pancreatic body or tail. Cysts arising from the pancreatic head are usually not resectable and are best treated by internal drainage in the form of cystogastrostomy or cystojejunostomy.

Because of the epithelial lining of the cyst, the chance of recurrence is likely, which makes a CT-scan guided or ultrasound-guided percutaneous drainage not a good alternative. If a drainage procedure is performed, a biopsy of the cyst wall should be taken to exclude cystadenocarcinoma, a rare but important cystic neoplasm of the pancreas.⁵

In conclusion, although congenital pancreatic cyst is rare, it should be included in the differential diagnosis of children presenting with cystic abdominal swellings. To avoid complications, these cysts should be treated early. Surgical resection is the treatment of choice, but unresectable cysts should be treated by internal drainage in the form of cystogastrostomy or cystojejunostomy.

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