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Congenital true pancreatic cyst: a rare case

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ABSTRACT
Congenital true pancreatic cysts are very rarely seen in children. We report magnetic resonance imaging features of a case of congenital true pancreatic cyst with a high level of enzymatic activity which, to the best of our knowledge, have not previously been reported. A 4-month-old boy was admitted to our clinic with a history of abdominal swelling for one month. A mobile, smooth, non-tender mass was palpated on the left side of the abdomen during physical examination. Ultrasonography and MR imaging revealed a bilocular cystic mass sized 9.5 x 8 x 6 cm. The spleen was displaced superiorly, whereas the left kidney was displaced posteriorly. Obliteration of the peripancreatic fat planes between the cystic mass and tail of the pancreas was observed. During abdominal surgical exploration, the pancreatic tail was larger than normal, and a pancreatic cyst arising from the tail of the pancreas was observed. Total cystectomy was performed with distal pancreatectomy. Although it is extremely rare in children, congenital true pancreatic cysts should be diagnostically considered in cases involving a cystic mass neighboring the pancreas. MR imaging is not helpful in differential diagnosis of other cysts originating from neighboring organs. Total excision with distal pancreatectomy and splenic preservation are advised for distal pancreatic cysts.

Key words: • pancreas • cyst • child • magnetic resonance imaging

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Congenital true pancreatic cysts, which are not the result of trauma, are rarely seen in childhood, and less than 1% of pancreatic cysts seen in children are true pancreatic cysts. In most cases, symptoms are seen under the age of two years (1-3). To the best of our knowledge, only 25 cases were reported as congenital true pancreatic cysts in the literature none of which included magnetic resonance (MR) imaging findings (4). In true cysts, enzymatic activity of cystic fluid is at normal levels. Herein, we report such a rare case of congenital true pancreatic cyst with high enzyme activity of cystic fluid.

Case report

A 4-month-old boy (6,800 g) was admitted to our clinic with a history of abdominal swelling for one month. His birth was a normal vaginal delivery at 38 weeks of gestation and the postnatal period was unremarkable. A mobile, smooth, non-tender mass was palpated on the left side of the abdomen during physical examination. Routine blood and urine biochemistry profiles were at normal levels. Abdominal ultrasonography demonstrated a bilocular cystic lesion sized 9.5 x 8 x 6 cm and located medial to the spleen, originating from the left midline lying towards the pelvis, nearby the left kidney and spleen. Abdominal MR images revealed that the left kidney and the spleen were displaced towards the posteriolateral and superior, respectively, by a bilocular cystic mass. The body and tail of the pancreas and tissue interfaces could not be observed. The mass was evaluated as hypointense in T1-weighted and hyperintense on T2-weighted MR images (Figure 1). A preliminary diagnosis of duplication cyst, mesenteric cyst, or pancreatic cyst was made.

Surgical exploration of the patient revealed two major cystic masses adjacent to each other and originating from the tail of the pancreas. These masses were filled with a light yellow-colored cystic fluid and were not attached to adjacent structures. Additionally, tiny cystic masses were found in the tail of the pancreas (Figure 2). The tail of the pancreas was observed to be wider than usual. Distal pancreatectomy with splenic preservation and total cystectomy was performed. The postoperative period was complication-free and the patient was discharged on the sixth postoperative day. No complications occurred during the six-month follow up period.

Histopathological examination revealed cystic cavities in various shapes and sizes in the tail of the pancreas. These cavities were embedded in the pancreatic tissue and coated with cuboidal epithelial tissue (Figure 3). Biochemical analysis of the cystic fluid revealed an amylase activity of 300 U/L.

Discussion

Pancreatic cysts are classified into six types as congenital-developmental cysts, retention cysts, duplication cysts, pseudocysts, neoplastic cysts,

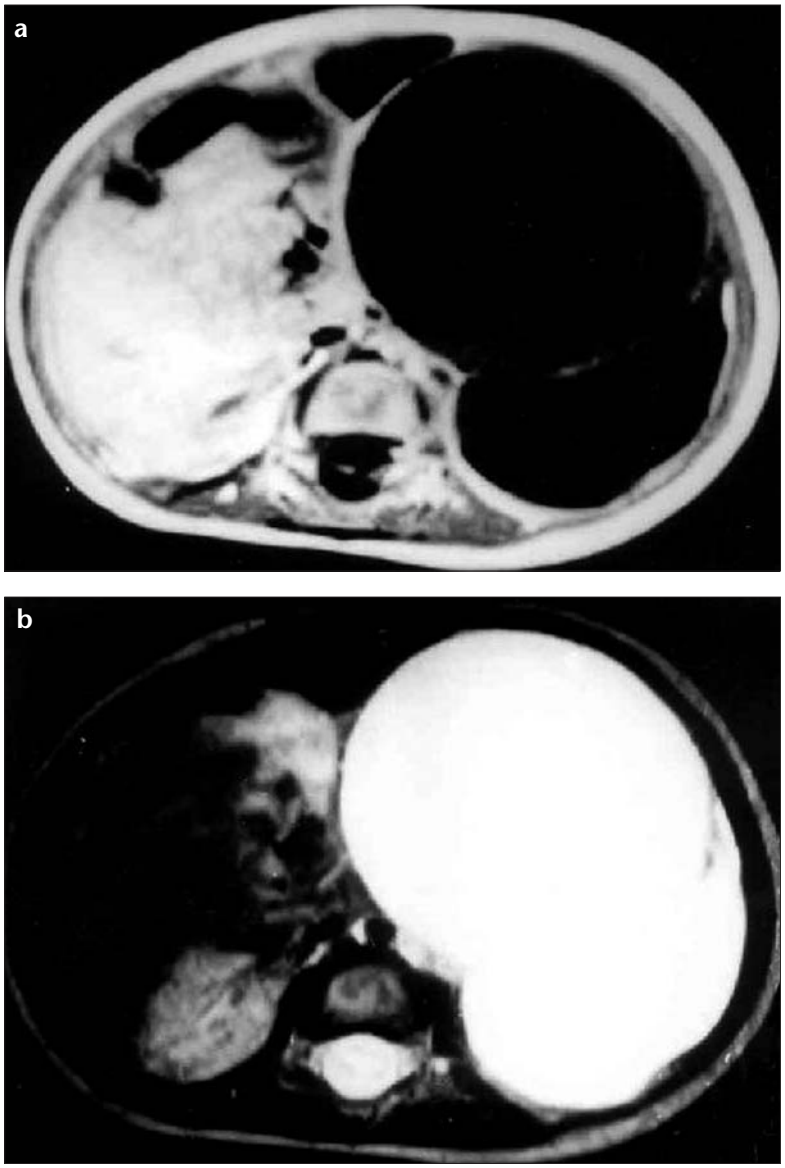

Figure 1. a, b. Transverse T1- (**a**) and T2-weighted (**b**) MR images show a lobulated mass in the left upper quadrant of the abdomen, hypointense on T1- and hyperintense on T2-weighted sequences.

and parasitic cysts (3). The first three types are also known as true cysts and these cysts have true epithelial components on their walls (5). In the most frequently seen posttraumatic pancreatic pseudocysts, the walls of the cysts do not contain epithelial components (1-3).

Generally, symptoms of congenital pancreatic cysts are seen in patients under the age of two years. To the best of our knowledge, only few cases have been reported in the literature and most of these cases were diagnosed under the age of two years (4). It is accepted that true cysts occur as a result of developmental anomalies related to the sequestration of primitive pancreatic ducts (6). In our case, the presence of tiny cysts, in addition to two major cysts, showing continuity towards the tail of the pancreas were evaluated as indicators of ductal developmental anomaly. Furthermore, the epithelial component on the cystic wall was indicative of a true cyst.

Congenital pancreatic cysts are generally asymptomatic, although abdominal distention, vomiting, jaundice, or pancreatitis can be observed (1, 2). In our patient, evident symptoms related to the cystic mass had not been observed. True cysts are frequently localized in the tail or neck of the pancreas (62%). Localization in the head of the pancreas was reported in 32% of cases (1). In our case, it was determined that the cyst had originated from the tail of the pancreas.

In congenital cystic fluid low enzymatic activity is observed generally (3), whereas enzymatic activity of retention cysts is considerably high (1,000-3,000 U/L). Retention cysts secondary to chronic obstruction of the ductal system are generally diagnosed in adulthood. It is difficult to differentiate them from congenital true cysts by histopathological features. In our patient, cystic fluid had an amylase activity of 300 U/L, which is greater than the amylase activity of serum. Although amylase activity of cystic fluid was higher than the normal limits, this activity was lower than the value expected for a retention cyst. When we also took the age of the patient into consideration, we concluded that the cyst was a true congenital cyst.

For evaluation of the abdominal masses, ultrasonography is a rapid and reliable technique used for cystic-solid differentiation. However, for an accurate localization of the abdominal masses, there is a frequent necessity for computed tomography (CT) or MR imaging. MR imaging may be insufficient for evaluating the origin of huge cysts. However, the relationship between a cyst and surrounding tissues, and extensions of a cyst can be demonstrated better with MR imaging (due to its multiplanar capacity) than with CT. In the present case, MR imaging was performed and hypo- and hyperintense signals were obtained in T1-weighted, and T2-weighted images, respectively. Although these signals cannot be distinguished from other cysts localized in this region, we believe that for evaluating extensions and the nature of huge cysts in pediatric patients which are observed by ultrasonography, MR imaging is a good choice.

Total excision should be the preferred treatment for cysts. If it is not possible, internal drainage methods are advisable (2, 7). In our patient, total excision with distal pancreatectomy and splenic preservation was performed for the distal pancreatic cysts.

In conclusion, with true pancreatic cysts, enzymatic activity of cystic fluid can be higher than would be expected. Although for lesions of this region MR examination is not completely effective for the determination of the origin of the cysts, it is preferable to CT. For surgical treatment, total excision with

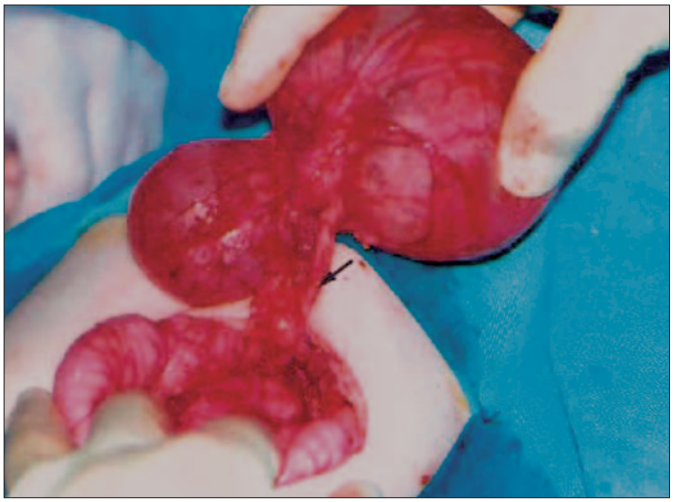

Figure 2. During the surgery, a bilocular cystic mass showing continuity towards the tail of the pancreas was observed *(arrow: tail of the pancreas)*.

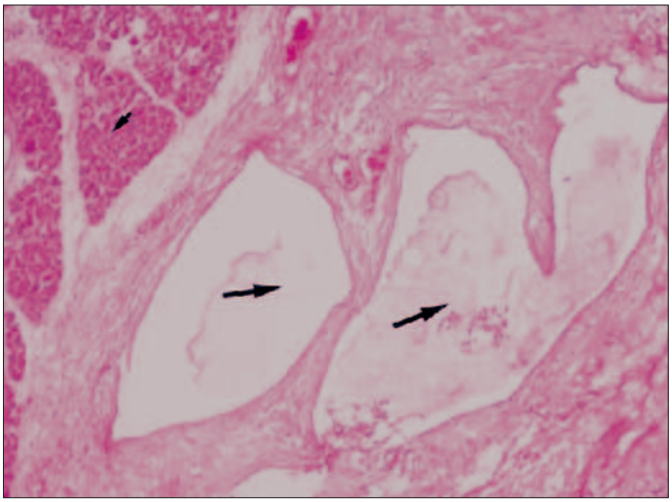

Figure 3. Normal pancreatic tissue *(small arrow)* and cystic areas *(large arrows)* coated with cuboidal epithelial tissue in various sizes were observed during histopathological examination (HE x40).

distal pancreatectomy and splenic preservation is preferred.

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