Multicystic adenomatoid pancreatic hamartoma in a child: Case report and literature review

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A B S T R A C T

\textbf{INTRODUCTION:} Pancreatic tumor is a rare condition in children, but reasonably common in adults. Histopathology in children also differs from that in adults, with most cases being pancreaticoblastoma, solid pseudopapillary tumor, or pancreatic endocrine tumors.

\textbf{PRESENTATION OF CASE:} A 14-month-old boy was noticed abdominal distension and referred to our hospital. Laboratory findings revealed leukocytosis and elevation of serum level of C-reactive protein and pancreatic enzymes. Radiological findings at admission were the huge abdominal cyst in abdominal computed tomography. As the levels of pancreatic enzyme elevated synchronous to oral feeding, total parenteral nutrition was needed. Besides radiographically the abdominal lesion changed from multiple large cystic type to multiple microcystic lesion including solid component over time. It was considered different diagnosis was pancreatic blastoma, pancreatic pseudo cyst, and lymphangioma and he was performed operation. The huge multicystic and partially solid tumor arising from the tail of pancreas existed from posterior of stomach to pelvic cavity. The tumor was completely resected without pancreatectomy and residual pancreas can be kept without tumor invasion. Histopathological finding was pancreatic hamartoma.

\textbf{DISCUSSION:} Pancreatic hamartoma was extremely rare, and only 17 cases were previously reported in the literature. This is the first case that the change of radiographic findings overtime was shown. We reviewed 17 cases (4 cases in children) of pancreatic hamartoma including our case.

\textbf{CONCLUSION:} Although extremely rare, pancreatic hamartoma should be considered in the differential diagnosis of cystic abdominal mass in children.

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1. Introduction

Pancreatic tumor is a rare condition in children, but reasonably common in adults. Histopathology in children also differs from that in adults, with most cases being pancreaticoblastoma, solid pseudopapillary tumor, or pancreatic endocrine tumors.\textsuperscript{1} We report an extremely rare case of multicystic pancreatic hamartoma in a 14-month-old boy and review the literature.

2. Presentation of case

A 14-month-old boy was noted to have abdominal distension by his parents and was referred to our hospital for assessment. He was delivered at term after a normal pregnancy and growth had been normal. Family history was negative for pancreatic and other gastrointestinal anomalies. On physical examination, the upper abdomen was markedly distended and tender.

Laboratory findings revealed leukocytosis (17,400/mm\textsuperscript{3}) and elevation of serum C-reactive protein (6.2 mg/dL). Pancreatic enzymes were also elevated: serum amylase (Amy) was 242 IU/L; serum lipase (Lip) was 1648 IU/L. Tumor markers (Ca19-9, AFP, HCG-β) were within normal range. Magnetic resonance imaging (MRI) showed a homogenous, multicystic lesion in the abdomen (Fig. 1a).

As serum pancreatic enzyme levels rose with oral feeding, total parenteral nutrition was commenced. Fine needle aspiration of cyst fluid was performed under ultrasonographic (US) control and found to contain elevated pancreatic enzymes (Amy: 14,085 IU/L; Lip: 247,660 IU/L), but no evidence of malignancy. Over 2 months, the radiologic appearance of the abdominal lesion changed from multiple large cysts to multiple microcysts with solid components (Fig. 1b). Provisional diagnoses included pancreatic blastoma and pancreatic pseudocyst and excision was planned. At laparotomy, a huge multicystic and partially solid tumor arising from the tail of the pancreas was found extending from the posterior aspect of the stomach to the pelvic cavity. Macroscopically, the tumor was 19 cm × 17 cm × 7 cm in size, and the multiple cysts contained clear...
fluid. The pancreatic duct was not connected to the tumor. The tumor was resected in toto without pancreatectomy. Postoperatively, Amy and Lip returned to normal. One month later, a cystic lesion developed near the pancreas and was drained of fluid suggestive of an old hemorrhage. Currently 26 months later, he is well and symptom-free.

Histopathology finding on the low magnification was showed that the mass grew toward the outside pancreas, had the cystic lesion with multiple small cysts. On high magnification it was revealed that the cystic lesions were lined by flat columnar epithelial cells and immature acinar cells, ductal cells, and endocrine cells embedded in rich fibrous immature stroma. Exocrine and ductal components were embedded in fibrotic stroma, but there were no spindle cells. Pancreatic ducts were dilated to varying degrees and surrounded by islet cells with normal components (Fig. 2). Based on these findings, a diagnosis of multicystic adenomatoid pancreatic hamartoma was made.

3. Discussion

In the pediatric age group, cystic lesions of the pancreas are unusual, with pseudocyst predominating. Primary neoplasms of the pancreas are rare in to the first two decades of life.² Albrecht⁴ first introduced the term “hamartoma” to describe “tumor like malformations” of the liver, spleen, kidney, and breast that show an abnormal admixture of normal components typical of the organ involved. Presently, hamartoma is defined as a lesion with disarranged component cells which for the pancreas involves presence of three types of disarranged cellular components in varying proportions; acinar, islet, and ductal cells.⁵ Morphologically, there are two types of pancreatic hamartoma; solid with cystic lesions⁶ and solid.⁵ It is an extremely rare condition both in children and adults, and there are only 17 other cases reported in the literature (Table 1).¹⁴-⁸ Age at presentation ranged from 34 weeks to 62 years (mean: 35.6 years); 4 were in the pediatric age group. Although the head of the pancreas was the most common origin, tumor was reported to arise from all parts of the pancreas. Size varied from 1.0 cm to 14 cm, with our case being the largest. Symptoms were nebulous and only one pediatric case had hypoglycemia. Laboratory data were available for only 7 cases and only 2 had elevated pancreatic enzymes. Surgical resection was performed in all except for 2 autopsy cases. Pancreaticoduodenectomy was performed when tumor arose from the head of the pancreas, even in children (n = 2), but local resection was performed when tumor arose from the tail of the pancreas (n = 3, including our case). There is only one other reported case that is similar to ours; a 20-month-old child with multicystic pancreatic hamartoma with solid components.⁷ The tumor was localized, 9 cm in size, but not associated with pancreatitis or change in radiologic appearance.

The most distinguishing feature of our case was the unique change in consistency that progressed over time; from a mass with large multiple cystic lesions to a mass with microcystic lesions with

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Fig. 1. (a) Abdominal MRI T2-weighted image on admission. The abdominal lesion contains multiple large high intensity cysts. Solid components are present. (b) Abdominal MRI T2-weighted image 2 months after admission. The abdominal lesion contains multiple small cysts with solid components.

Fig. 2. Hematoxylin and eosin staining of a cystic lesion. Left panel: immature acinar cells, ductal cells, and endocrine cells embedded in rich fibrous immature stroma. The exocrine and ductal components are embedded in fibrotic stroma. Right panel: the cystic lesion is lined by tall columnar pancreatic epithelial cells (arrows). The stroma has hemorrhage lesion.
Table 1
Review of reported cases of pancreatic hamartoma.

| Case | Ref. | Year | Age | Sex | Symptoms          | CP | Location | Size  | Consistency | Surgery                  | Outcome       |
|------|------|------|-----|-----|-------------------|----|----------|-------|-------------|--------------------------|---------------|
| 1    | 4    | 1977 | 46y | M   | –                  | Head | 1.6 cm   | Solid and cystic | PD           | Died 3 months later      |               |
| 2    | 4    | 1977 | 52y | F   | Abdominal mass     | +   | Tail     | 254 g | –           | Autopsy                  |               |
| 5    | 4    | 1983 | 34w | F   | Hypoglycemia, abd mass | Diffuse | 11.5 cm | Solid and cystic | PD + splenectomy | Died 3 months later      |               |
| 6    | 7    | 1992 | 20m | M   | Abdominal distension | –   | Head     | 9.0 cm | Solid and cystic | Local resection          | Alive 9 months |
| 7    | 4    | 1994 | 25y | M   | Epigastric pain, nausea | –   | Head     | 10.6 cm | Solid and cystic | PD           | Alive 4 years            |
| 8    | 4    | 1998 | 39y | M   | Epigastric pain     | –   | Head     | 8.0 cm | –           | PD                       | Alive 9 months |
| 9    | 4    | 2004 | 29y | M   | Abd pain, weight loss | –   | Head     | 1 cm   | –           | PD                       | Alive 2 years  |
| 10   | 5    | 2004 | 62y | M   | Abd pain, weight loss | –   | Head     | 3.5 cm | –           | PD                       | Alive 3 months |
| 11   | 5    | 2005 | 36y | F   | Epigastric pain     | –   | Head     | 7.0 cm | Solid and cystic | PD + nephrectomy         | Alive 15 months|
| 12   | 5    | 2005 | 55y | F   | Abd pain            | –   | Neck     | 3 cm   | Solid and cystic | Distal pancreatectomy    | Alive 23 months|
| 13   | 6    | 2005 | 51y | M   | –                  | Tail | 3 cm     | Solid | Local resection | Alive 2 years            |               |
| 14   | 6    | 54y  | F   | Abdominal discomfort | –   | Body     | 2 cm   | Solid | Distal pancreatectomy | Alive 4 years            |               |
| 15   | 4    | 2005 | 58y | F   | –                  | Body | 2 cm     | Solid | Distal pancreatectomy | Alive 4 months            |               |
| 16   | 1    | 2008 | 3y  | M   | Epigastric pain     | +   | Head     | 3 cm   | Multicystic | PD                       | Alive 3 months |
| 17   | 8    | 2009 | 46y | M   | Abdominal mass      | –   | Head     | 8 cm   | Solid | PD           | Unknown                  |               |
| 18   | 5    | 2009 | 14m | M   | Abd pain, abd distension | +   | Tail     | 14 cm  | Multicystic | Local resection         | Alive 26 months|

CP: clinical pancreatitis, F: female, M: male, PD: pancreaticoduodenectomy.

Solid components. Another feature of interest was the association of pancreatitis. We speculate pancreatitis was induced by oral feeding because there was elevation in pancreatic enzymes after every oral feed. Because the cyst fluid contained pancreatic enzymes, we considered that communication between the ducts and the cysts might be present but no such communication was confirmed at surgery. Persistent pancreatitis could have been etiologic in remodelining the tumor, causing it to grow so large with solid tissue components.

To the best of our knowledge, this is the first report of morphologic change over time in pancreatic hamartoma. Although extremely rare, pancreatic hamartoma should be considered in the differential diagnosis of cystic abdominal mass in children.

Conflict of interest
None.

Funding
None.

Ethical approval
We hereby state that written consent has been obtained from the patient’s parents and will be provided upon request.

Author contributions
Ryo Sueyoshi has written the entire manuscript for this study. Atsushi Arakawa and Takashi Yao have checked the validity of pathological results. Finally, Tadaharu Okazaki, Geoffrey J. Lane and Atsuyuki Yamataka have reviewed the manuscript for exactness.

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