Acinar Cell Cystadenocarcinoma of the Pancreas in a 4-year-old Child

To the Editor:

Cystic neoplasms of the pancreas are rare, constituting only 10% of all cystic lesions of the pancreas in the adult population. These neoplastic pancreatic cysts can be divided into serous adenomas, mucinous cystadenomas, and cystadenocarcinomas.

Acinar cell cystadenocarcinomas (ACCCs) are a very rare variant of acinar cell adenocarcinoma of the pancreas, of which only 7 cases have been reported. All these cases occurred in adults. No child was reported to have this disease. We now describe the clinicopathologic features of a 4-year-old child with ACCC and review the relevant literature.

CASE REPORT

A previously healthy 4-year-old boy presented with a 6-month history of abdominal pain, which was accompanied by weight loss. Physical examination showed one mass in right hypochondrium. An abdominal ultrasound scan showed a low echo mass cystic mass. Abdominal computed tomography scan demonstrated a soft tissue imaging arising from the pancreatic head and no para-aortic lymph node swelling.

He underwent aggressive surgery. A 16 × 16 × 18-cm pancreatic multilocular cystic mass was found replacing the head of the pancreas. Duodenum and perigastric lymph nodes were not involved. Resection of the mass required a pylorus-preserving pancreaticoduodenectomy (PPPD). Acinar cell cystadenocarcinoma was confirmed by postoperative pathological examinations (Fig. 1).

The child was discharged 15 days after operation. In 1 year, the gastrointestinal barium-meal examination showed normal gastric storage and emptying. No symptom of bile regurgitation gastritis was found, the weight was regained, no metastasis and recurrence occurred, and normal growth was achieved.

DISCUSSION

Acinar cell adenocarcinomas are rare tumors of the exocrine pancreas, accounting for 1% to 2% of all pancreatic tumors. They are defined as carcinomas exhibiting evidence of pancreatic enzyme production by neoplastic cells, although the histopathological criteria are highly specific of this entity.

Cystic changes in acinar cell carcinomas are uncommon; and to the best of our knowledge, ours is only the eighth case reported to date and is the first case that occurred in a child in the literature. The clinical data of these 8 patients are summarized in Table 1.

Diagnosis

According to Table 1, the age at onset, sex, and tumor site of ACCCs have no distinctiveness. The main presenting symptoms in all the cases were an abdominal mass, epigastric pain, and weight loss. The patients of Ishizaki et al had high serum lipase, trypsin, and alpha-fetoprotein levels, without any specific clinical symptoms. The patients of Beltraminelli et al had painful erythematous nodules on the lower legs. It was easily found by ultrasound and computed tomography. Large tumor can suppress and infiltrate surrounding tissues, and distant metastasis occurred in the liver, pelvic, omentum, peritoneum, mesenteric tissue, etc.

The macroscopic characteristics of the neoplasm were similar in all cases: large multilocular cystic masses replacing the head, body, or tail of the pancreas. Microscopically, our case and all of the previously reported cases had similar features: multiple cysts and tubules lined by a single layer of cuboid or columnar cells with basal nuclei, and the cytoplasmic characteristics of acinar cells as in the solid tumors. Histochemistry and the immunohistochemical phenotype confirmed the acinar origin of the tumor, and documented enzyme production is often helpful in confirming the diagnosis. Periodic Acid Schiff reaction after diastase digestion revealed granules within the cytoplasm; however, a more sensitive way to detect acinar differentiation is immunohistochemical staining for specific enzymes. Confirmed diagnosis is based on the method of pathology or immunohistochemistry.

Surgical Treatment

Klimstra et al estimated that the overall survival of patients with acinar cell carcinoma lies between that of patients with ductal adenocarcinoma of the pancreas. A, × 40. B, × 100.
TABLE 1. Summary of Cases

<table>
<thead>
<tr>
<th>Author</th>
<th>Reference Year Age, yr</th>
<th>Sex</th>
<th>Site</th>
<th>Size, cm</th>
<th>Metastasis</th>
<th>Outcome (m)</th>
<th>Symptoms at Presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cantrell</td>
<td>1 1981 64 Male</td>
<td>Body and tail</td>
<td>39</td>
<td>Liver, pelvic</td>
<td>Dead (13)</td>
<td>Epigastric pain, abdominal mass</td>
<td></td>
</tr>
<tr>
<td>Stamm</td>
<td>2 1987 42 Male</td>
<td>Body and tail</td>
<td>17</td>
<td>Liver</td>
<td>Alive (18)</td>
<td>Weight loss (6 kg), abdominal mass</td>
<td></td>
</tr>
<tr>
<td>Hoorens</td>
<td>3 1993 64 Male</td>
<td>Body</td>
<td>17</td>
<td>Not reported</td>
<td>Not reported</td>
<td>Not reported</td>
<td></td>
</tr>
<tr>
<td>Ishizaki</td>
<td>4 1995 57 Male</td>
<td>Tail</td>
<td>Not reported</td>
<td>Liver</td>
<td>Alive (37)</td>
<td>Epigastric pain with vomiting</td>
<td></td>
</tr>
<tr>
<td>Joubert</td>
<td>5 1998 32 Female</td>
<td>Head</td>
<td>13</td>
<td>Liver</td>
<td>Alive (13)</td>
<td>Diffuse abdominal pain</td>
<td></td>
</tr>
<tr>
<td>Colombo</td>
<td>6 2003 69 Male</td>
<td>Body and tail</td>
<td>25</td>
<td>Peritoneum, mesenteric tissue</td>
<td>Alive (6)</td>
<td>Acute abdominal pain</td>
<td></td>
</tr>
<tr>
<td>Beltraminelli</td>
<td>7 2003 60 Male</td>
<td>Not reported</td>
<td>10</td>
<td>Liver, pelvic, etc.</td>
<td>Dead (2)</td>
<td>Painful erythematous nodules</td>
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<tr>
<td>Current case</td>
<td>7 2005 4 Female</td>
<td>Head</td>
<td>Not reported</td>
<td>Surrounding soft tissue</td>
<td>Alive (12)</td>
<td>Abdominal pain, weight loss</td>
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Surgical management is curative in cases without distant metastasis or with organ-confined diseases. No recurrence was found in our case in 1 year. It may have a good prognosis. For those patients with metastatic disease, treatment should concentrate on alleviating pain and improving the quality of life.

However, we believed active operative treatment is critical, especially for those patients without distant metastasis or with organ-confined diseases. In our case, PPPD was performed after considering the pediatric anatomical and physiological features and the conditions of the case, as follows:

1. If the tumor of the pancreatic head has not invaded the first part of the duodenum, metastasis will not occur in the perigastric lymph nodes of No. 5 (suprapyloric lymph nodes) and No. 6 (infrapyloric lymph nodes). Under these conditions, PPPD can prevent complications after gastrectomy, keep stomach storage and other physical functions, maintain the normal level of gastrointestinal hormone, and help in improving nutrition status and regaining lost weight.

2. During choledochojejunostomy and pancreaticojejunostomy, stent drainage was impossible because the pancreatic and bile ducts were very small. End-to-end pancreaticojejunostomy invagination can reduce the incidence of complications such as pancreatic leakages and is easy to perform. The diameter of the bile duct in our case is 0.2 to 0.3 cm. Two symmetrical incisions in the bile duct can enlarge the inner diameter, making choledochojejunostomy possible.

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**REFERENCES**


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Mucinous Cystic Tumor of the Pancreas in a Man

A Rare Case

**To the Editor:**

In 1996 and 1997, the World Health Organization and the Armed Forces Institute of Pathology clearly defined and differentiated mucinous cystic tumor (MCT) and intraductal papillary-mucinous tumor; that is, MCT involves “ovarian-type stroma” that consists of densely packed, spindle-shaped cells, with round...
or elongated nuclei and sparse cytoplasm, whereas intraductal papillary-mucinous tumor does not. In the Japanese criteria, the ovarian-type stroma is not essential but usually present in MCT. Interestingly, regardless of the difference in these criteria in the presence of ovarian-type stroma, MCTs of the pancreas have been reported to be predominantly located in the distal pancreas of middle-aged women in the United States, Europe, and Japan since then. Therefore, there are several issues to be clarified concerning the clinicopathological features of MCT of the pancreas occurring in men.

**CASE REPORT**

A 45-year-old man was referred to our hospital because of a pancreatic cystic mass found in abdominal ultrasound at a physical checkup. His medical history was not significant for pancreatitis or abdominal injury. Computed tomography scan revealed a multilocular cystic tumor in the tail of the pancreas. Endoscopic retrograde cholangiopancreatography showed no communication between the pancreatic duct and the tumor. Because malignancy could not be completely ruled out and the patient complained of abdominal discomfort, distal pancreatectomy with preservation of the spleen was performed. The sectioned tumor showed multilocular cysts measuring 3.5 cm in greatest dimension, with smooth, glistening cavities separated by thin, transparent septations. The tumor consisted of various-sized cysts lined by the epithelium of mucin-producing columnar cells that were positive for periodic acid Schiff alcian blue staining. Immediately subtending the epithelium was a dense, cellular, fusiform to spindle cell stroma, reminiscent of ovarian stroma. This ovarian-type stroma immunohistochemically reacted with monoclonal antibodies to estrogen receptor and progesterone receptor (ER and PgR, respectively) and inhibin-α (Figs. 1A–C). Serum levels of estradiol and progesterone were normal, and the patient did not show any external signs of feminization. The present tumor sufficiently fulfilled the World Health Organization and Armed Forces Institute of Pathology criteria, so it is considered to be a male case of MCT of the pancreas.

**DISCUSSION**

To date, the exact pathogenesis of MCT of the pancreas has not been fully understood. Zamboni et al hypothesized that MCT of the pancreas originates from remnant primordial gonadal cells that migrated to the dorsal pancreatic
anlage that forms the distal pancreas at an early stage of development (fourth to fifth week). This unique hypothesis is based on the finding that the left primordial gonad and the dorsal pancreatic anlage lie side by side during that period, and is very convincing because it could well explain the unique nature of MCT of the pancreas.

Immunohistochemical staining revealed that the ovarian-type stroma of the present case reacted with monoclonal antibodies to ER, PgR, and inhibin-α. The possible role of the female sex hormone receptors has not been well clarified; however, the frequent occurrence of ER and/or PgR positivity in the cells of the ovarian-type stroma suggests that the interaction of female sex hormones with their receptors may promote or sustain tumor growth. In contrast, inhibins are secreted by granulosa cells of the ovary and their receptors may promote or sustain tumor growth. In contrast, inhibins are secreted by granulosa cells of the ovary and Sertoli cells of the testis. In pancreas and Sertoli cells of the testis are incorporated into the dorsal pancreatic anlage; or the left primordial gonad of men and the dorsal pancreatic anlage usually may not lie side by side during this period, leading to the rarity of male cases of MCT of the pancreas.

In any event, the present case indicates that MCT of the pancreas may also develop in middle-aged men through the same mechanism as that for women; and additional male cases would be required to investigate the pathogenesis of this distinctive tumor.

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**REFERENCES**


**Shingles-associated Pancreatitis**

**To the Editor:**

Pancreatitis is an almost unusual complication of shingles, and only a few cases of pancreatitis caused by varicella-zoster virus (VZV) have been described among immunocompetent patients. We report a case of VZV-induced pancreatitis in an apparently immunocompetent patient.

An 82-year-old man presented with a 4-day history of nausea, vomiting, and constant pain in the epigastrium that radiated to the flanks; 5 days before admission, he had noted a painful and itchy rash in the periumbilical zone. His previous history was otherwise unremarkable; he did not report any acute or chronic pancreatic disease and denied alcohol use, toxic habits, taking any medications, including over-the-counter medications, or herbal remedies, or exposure to ill persons.

Upon admission, the patient was fully alert and oriented, aperistalsis, and had normal vital parameters. Physical examination yielded normal findings apart from a severely tender abdomen...
with no bowel sounds; we also observed several vesicles, some of which were coalescing and others were scabbing, in his left periumbilical region with a dermatomal distribution. Laboratory data showed leukocytosis (13.5 × 10^9/L), with a normal differential and increased blood levels of amylase (791 IU/L; reference range, 0–115 IU/L), lipase (1129 IU/L; reference range, 0–190 IU/L), aspartate aminotransferase (176 U/L; reference range, 10–36), and alanine aminotransferase (161 U/L; reference range, 10–36); total bilirubin was 1.6 mg/dL (reference range, 0.2–1), and γ-glutamyltransferase and alkaline phosphatase were within the reference range. Findings on electrolytes, hematological variables, cholesterol level, and triglycerides and kidney function test results were normal. Results of serologic tests for Mycoplasma and Chlamydia species, viral hepatitis, and a wide range of viral infections were also negative, except for elevated VZV-specific antibody titers (1:640). Abdominal ultrasonography and computed tomography showed a moderately enlarged and edematous pancreas, with normal findings on gallstone and biliary ducts and without evidence of biliary stones or sludge or any other abnormal findings.

The patient was treated with acyclovir (800 mg for every 8 hours), intravenous fluids, and bowel rest. Five days after admission, blood levels of amylase, lipase, and aminotransferases returned to normal, and he was free of symptoms. We continued acyclovir therapy until day 11 when the patient was discharged home; at this time, several healed lesions were present in the left periumbilical region with no vesicles or pustule. At a follow-up visit 1 month later, he was doing well, with no clinical or laboratory evidence of active pancreatic or hepatic disease or VZV infection.

Many findings implicate VZV as the cause of acute pancreatitis in this patient, and the elevated blood levels of aminotransferases seem to indicate that also the liver, in addition to pancreas, was involved in VZV dissemination and replication. As a matter of fact, he had no history of acute or chronic pancreatic disorder, and the temporal relationship between the development of shingles in the left periumbilical region and presentation with clinical symptoms and laboratory findings of pancreatitis is clear. We found no alternative causes, such as biliary stones or sludge, alcohol abuse, hypertriglyceridemia, or hypercalcemia to explain the onset of acute pancreatitis in this case. Furthermore, the patient was not taking any other drugs or natural remedies at the onset of pancreatitis and during the preceding 6 months.

Pancreatitis is a rare complication of VZV infection, which does occur more frequently among immunocompromised patients such as those with acquired immunodeficiency syndrome, leukemias, lymphomas, or other malignancies or transplanted patients. In contrast, our MEDLINE search yielded only 3 cases of pancreatitis caused by VZV reactivation among apparently immunocompetent subjects—two of which were infants or young children.

Varicella-zoster virus may spread to the pancreas during the course of shingles by several paths; however, the onset of abdominal pain along with the finding of elevated blood levels of pancreatic enzymes does usually precede the development of the typical vesicular eruption. Such an interval could delay the diagnosis of VZV-related pancreatitis and preclude the early initiation of the appropriate antiviral therapy and monitoring and preventing the complications of disseminated VZV infection. Abdominal pain is often excruciating, which could suggest a catastrophic event such as a visceral perforation or mesenteric infarction, but physical examination is substantially unrevealing in most patients. Varicella-zoster virus–related pancreatitis usually has a benign course, and outcome is favorable, with a rapid and full recovery particularly among immunocompetent subjects and among patients with shingles rather than those with a disseminated vesicular eruption, when antiviral therapy is timely started. Nonetheless, even in these low-risk settings, a minority of patients could experience a severe form of pancreatitis with a prolonged and complicated course.

One interesting point is that our patient did experience the onset of severe abdominal pain several days after periumbilical shingles was noted, which underscores the important concept that pancreatitis may also be a late complication of shingles with an abdominal dermatomal distribution. This should suggest great caution before simply dismissing any episode of abdominal pain as a reflection of post-herpetic neuralgia rather than the direct involvement of the pancreas or other intra-abdominal viscera with VZV dissemination and replication.

Clinicians should include VZV-related pancreatitis in the differential diagnosis of patients with shingles who present with severe and otherwise unexplained abdominal pain. How often VZV is responsible for idiopathic pancreatitis is unclear.

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REFERENCES
Autoimmune Pancreatitis With Autoimmune Hemolytic Anemia

To the Editor:

We report a case of autoimmune pancreatitis presenting as a mass with obstructive jaundice, hemolytic anemia, diabetes, and hypothyroidism. Results of initial endoscopic ultrasound and fine-needle aspiration were suggestive of pancreatic head adenocarcinoma. The patient had a good response to steroid therapy, with improvement in pancreatic appearance, resolution of biliary obstruction, normalization of hemoglobin levels, and improvement in his diabetes control. To our knowledge, this is the first case report of autoimmune pancreatitis in association with hemolytic anemia.

A 52-year-old man presented with upper abdominal pain, and signs and symptoms of obstructive jaundice. His total bilirubin level was 231 umol/L. An endoscopic retrograde cholangiopancreatography (ERCP) was performed and showed a distal stricture of the common bile duct. A stent was placed with resolution of the patient’s jaundice. Brush cytology did not reveal evidence of malignant or atypical cells. A computed tomography (CT) scan of the abdomen showed a bulky pancreas with blurred margins, but no evidence of mass lesion or pancreatic duct dilatation (Fig. 1A). An endoscopic ultrasound was performed and demonstrated a mass in the pancreatic head and an enlarged lymph node. Fine-needle aspiration from the lymph node was negative. The aspirates from the pancreatic head mass, however, showed atypical cells suggestive of adenocarcinoma.

While investigating this patient for his obstructive jaundice, it was noticed that he had a low hemoglobin level of 64 g/L and severe postural hypotension. He was found to have an elevated reticulocyte count of 27.7% and a positive Coomb test, which were suggestive of acute hemolytic anemia. He had also developed acute-onset diabetes, with marked insulin resistance requiring approximately 100 U/d of insulin and was found to have an elevated thyroid-stimulating hormone level. He was positive for rheumatoid factor, and his serum IgG level was elevated at 19.2 g/L. The patient’s clinical picture led to the suggestion of an autoimmune process that could also include autoimmune pancreatitis. Therefore, he was started on prednisone 100 mg/d and cyclophosphamide 100 mg/d.

A repeat endoscopic ultrasound showed diffuse enlargement of the pancreatic head, body, and tail, but no mass was seen at that time. Multiple repeat fine-needle aspirates were taken from various parts of the pancreas, and no evidence of pancreatic lymphoma was found. He started to improve while on medical treatment, with complete resolution of abdominal pain, elevation of hemoglobin level to 103 g/L, and decreasing bilirubin level to 38 μmol/L. He was discharged home on prednisone 90 mg every other day and cyclophosphamide 100 mg/d. A repeat CT scan of the abdomen 2 months later demonstrated resolution of the changes in the pancreas (Fig. 1B), and a repeat ERCP 4 months later showed complete resolution of the bile duct stricture. Therefore, the stent was removed. A year and a half after diagnosis, he remains pain free with normal bilirubin and hemoglobin level, and insulin requirement is down to 10 U/d. He is off prednisone and takes 25 mg/d of cyclophosphamide.

DISCUSSION

The first reference to autoimmune pancreatitis was made in 1961 by Sarles.
et al\(^1\) and was called chronic inflammatory sclerosis of the pancreas. The term, autoimmune pancreatitis, was adopted in the 1990s. Autoimmune pancreatitis affects the male population more than the female population by a ratio of 2:1. Most patients are older than 50 years.\(^2\) Symptoms range from mild abdominal discomfort to an acute attack of pancreatitis or obstructive jaundice. It is often associated with other autoimmune diseases. There are several autoantibodies reported to be positive in patients with autoimmune pancreatitis that include antinuclear antibodies, serum antibodies to carbonic anhydrase 1 and 2, and antibodies to lactoferrin. They also have elevated levels of IgG\(^3\) and antiphospholipid antibodies.\(^4\)

Imaging studies usually show diffusely enlarged pancreas with no significant peripancreatic inflammation. In one third of the patients, a focal enlargement of the pancreas or even a mass lesion could be identified, with enlarged lymph nodes that could be confused with adenocarcinoma.\(^5\) In half of the patients, ERCP will demonstrate stricture of the distal common bile duct with irregular narrowing, extending into the hepatic duct.\(^6\) In most cases, there are no signs of chronic pancreatitis, but pseudocysts\(^7\) and pancreatic duct stone formation\(^7\) have been reported.

On pathological examination, the gross appearance of autoimmune pancreatitis can be similar to pancreatic adenocarcinoma. The appearance is usually gray to yellow/white induration of the affected portion.\(^8\) On microscopy, there is periductal infiltration with lymphocytes and plasma cells mainly\(^8\) that leads to periductal fibrosis, causing ductal thickening and obstruction.\(^2\) IgG 4-positive plasma cells were observed in the pancreas of patients with autoimmune pancreatitis.\(^9\)

Recently, a diagnostic criterion of autoimmune pancreatitis was proposed by the Japanese Pancreas Society and contains 3 criteria: 1) diffuse enlargement of the pancreas, with diffuse narrowing of the main pancreatic duct with irregular wall; 2) elevated serum IgG with the presence of antibodies; and 3) histological evidence of lymphocyte and plasma cell infiltration with fibrotic changes.\(^10\)

Autoimmune pancreatitis is usually highly responsive to steroid therapy, with normalization of the pancreatic size within 1 month. However, the pancreatic duct irregularity can last longer. Biliary obstruction can persist in approximately 50% of patients and will require endoscopic or surgical intervention.\(^10\) Some patients will need long-term, low-dose steroid therapy. There have been no reports of development of pancreatic cancer.\(^10\)

This case clearly indicates that autoimmune pancreatitis can occasionally present clinically in a picture similar to pancreatic carcinoma. The presence of other autoimmune diseases should raise the suspicion for autoimmune pancreatitis, which needs to be confirmed by further laboratory, imaging, and pathological studies to avoid unnecessary surgical intervention.

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