Solid pseudopapillary tumor of the pancreas as a cause of recurrent pancreatitis

Y. Ozturk[1], O. Bekem Soylu[1], B. Gurcu[2], R. Ortac[3], H. Cakmakci[4], A. Coker[2]

(1) Dokuz Eylul University Faculty of Medicine, Department of Pediatrics, Inciralti 35340, Izmir, Turkey ; (2) Ege University Faculty of Medicine, Division of General Surgery, Bornova, Izmir, Turkey ; (3) Dr. Behcet Uz Children's Hospital, Division of Pathology, Alsancak, Izmir, Turkey ; (4) Dokuz Eylul University School of Medicine, Division of Radiology, Inciralti-35340 Izmir, Turkey.

Abstract

Solid pseudopapillary tumor (SPT) of the pancreas is a rare pancreatic tumor mostly seen in young women. We here report a twelve-year-old girl presenting with recurrent attacks of pancreatitis. No history of a systemic disease, trauma, drug usage or infection was present. All other etiologic factors like familial, hypertriglyceridemia, hypercalcemia, cystic fibrosis, medications were excluded. On abdominal ultrasound a heterogeneous mass was noticed at the tail of pancreas. Computerized tomography and magnetic resonance imaging proved that the mass was cystic. The mass was surgically removed. The diagnosis was pancreatic solid cystic papillary epithelial neoplasm. Although acute pancreatitis due to SPT was exceptionally reported, this is the first description of SPT leading to recurrent pancreatitis especially in children. (Acta gastroenterol. belg., **2008**, 71, **390-392**).

Key words : recurrent pancreatitis, solid pseudopapillary tumor, children, acute pancreatitis.

Introduction

Acute pancreatitis is the inflammation of pancreas characterized with sudden onset of abdominal pain and arise in digestive enzymes. Even though most of the pancreatitis is idiopathic (22.2%), there are many other etiologic factors like severe systemic illnesses (20.8%), trauma (18.6%), structural abnormalities (10.6%), medications (10.2%), infections (7.7%), gallstone (3.1%), familial (2.4%), post-ERCP (1.2%), diabetic ketoacidosis (0.9%), hypercalcemia (0.9%), hypertriglyceridemia (0.8%), cystic fibrosis (0.6%) and others (2.4%). From these factors structural abnormalities, idiopathic or familial pancreatitis may lead to recurrent attacks of pancreatitis in 10% of the patients (1).

Solid pseudopapillary tumor (SPT) of the pancreas is a rare pancreatic tumor mostly seen in young women (2). The patients may present with abdominal pain, palpable abdominal mass or SPT may be found incidentally (3). Only one case is reported presenting with acute pancreatitis (4). We here report a twelve-year-old girl presenting with recurrent attacks of pancreatitis and solid pseudopapillary neoplasm was the only etiologic factor found.

Case report

A twelve-year-old girl presented with abdominal pain, nausea and emesis. Abdominal pain was sharp, tense, localized to epigastrium and periumblical area, did not relieve with analgesic drugs and was accompanied by nausea and vomiting. No history of a systemic disease, trauma, drug usage, infection or a similar disease in the family was present. She had experienced similar symptoms three and eleven months ago and amylase levels were found elevated during these attacks in other hospitals. The amylase levels, that were between 400 and 450 U/L during attacks, decreased to < 100 U/L afterwards, and abdominal ultrasound revealed edematous pancreas at these attacks. Only intravenous fluid therapy was given to the patient. At the last attack amylase level was found to be 308 U/L (reference range : 35-127 U/L) and leukocytes 10200/mm^3 (reference range : 4500-13500/mm^3) at another hospital. On abdominal ultrasound (US) a 4 cm heterogeneous mass was noticed in the tail of pancreas. The next day she was admitted to our clinic. Her weight was between 75th-90th percentiles, height was above 95th percentile. Physical examination was normal except tenderness on epigastric and periumblical area. Laboratory tests revealed leukocytosis (15000/mm^3) and slightly elevated amylase levels (171 and 158 U/L consecutively). Erythrocyte sedimentation rate, C-reactive protein, serum glucose, electrolyte, calcium, transaminase, bilirubin and triglyceride levels was normal. Chloride concentration in sweat test was within normal ranges. CFTR, SPINK1 and PRSS1 mutations were not present. Pancreatic rest, analgesia and intravenous fluid therapy were provided. Computerized tomography (CT) (Fig. 1) and magnetic resonance imaging (MRI) (Fig. 2) proved that the mass was cystic. The mass was surgically removed. Grossly the tumor was 8 cm in greatest diameter and was surrounded by a well developed capsule from the pancreas. The cut surface contained cystic and hemorrhagic areas. Microscopically the most distinctive feature was the presence of pseudopapillary structures covered by several layers of epithelial cells. The fibrovascular cores showed prominent mucinous changes. The epithelial cells nuclei were

Correspondence to : Ozlem Bekem Soylu, M.D., Camlicay Mahallesi 5182 sokak No : 12/A, Urla, Izmir, Turkey. E-mail : obekem@yahoo.com

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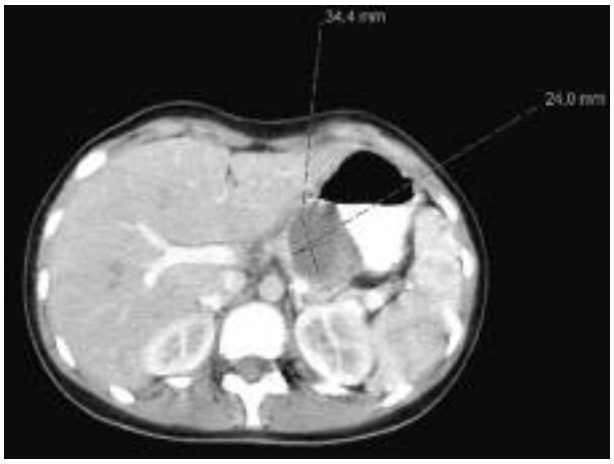

Fig. 1. — Abdominal CT shows encapsulated hypodense mass in tail of the pancreas.

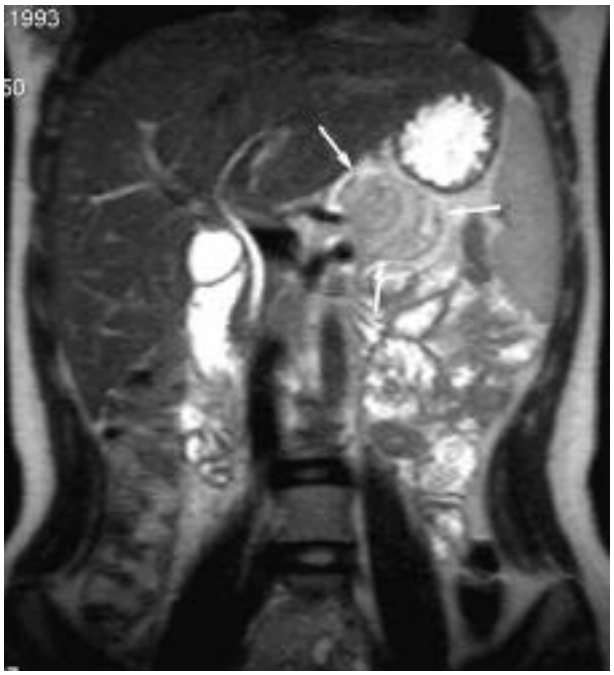

Fig. 2. — T2-weighted coronal image (a) and axial section (b) MRI of the abdomen shows mass with heterogeneous signal in tail of the pancreas.

oval or round shape. Nuclear pleomorphism was inconspicuous. Mitotic figures were not seen (Fig. 3). A few small, eosinophilic, PAS positive cytoplasmic globules were obtained (Fig. 4). The tumor cells showed strong cytoplasmic alpha-1 antitrypsin immunreactivity. The diagnosis was pancreatic solid cystic papillary epithelial neoplasm based on these pathological findings. After surgery she made an uneventful recovery in seven-month long follow-up.

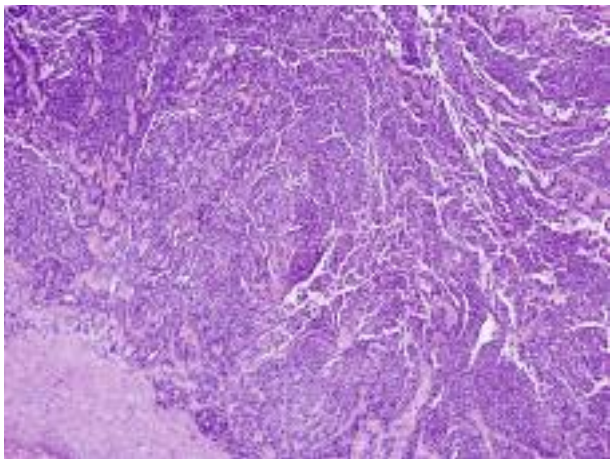

Fig. 3. — Pseudopapillary structures and solid components of the solid pseudopapillary tumor (HE, ×10).

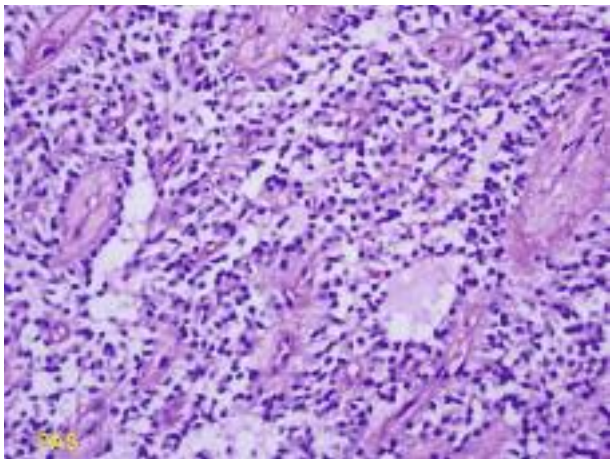

Fig. 4. — Eosinophilic, periodic acid–Schiff (PAS) positive cytoplasmic globules in solid pseudopapillary tumor (PAS, ×40).

Discussion

Solid pseudopapillary tumor of the pancreas is an uncommon childhood malignancy (2,3). This tumor especially affects young women and forms the 1-3% of the pediatric pancreatic tumors (2,4-6). Of the reported patients with SPT 626/690 were women (9.78:1) and the age of 710 patients was between 2-85 years with a mean age of 21.97 years (7). It is known that SPT has a low malignant potential and even though metastasis is reported, the prognosis is good (3,6,8).

Even though it is proposed that presentation of SPT vary, predominant presenting complaints are vague abdominal pain and abdominal mass. Some of the cases are asymptomatic and found incidentally (3,8,9). Backache, digestive symptoms including swelling, nausea and vomiting, fever, jaundice, weight loss, anorexia are the other symptoms reported (7,10). Presentation of SPT may be due to bleeding within the tumor, rupture

and hemoperitoneum (4,5,11). In only one article SPT was reported as the etiologic factor of acute pancreatitis (4). In this report 21-year-old female had presented with nausea and upper abdominal pain. Imaging and histological findings were concomitant with acute pancreatitis and pancreatic enzymes were elevated (4). Only etiological finding found was SPT in the pancreatic tail (4). Other than this case, elevated amylase levels were reported in 18% of 23 pediatric patients by Choi *et al.* (12). In our case presenting symptoms were abdominal pain, nausea and vomiting similar to literature. We also found slightly elevated serum amylase levels. Additionally, the amylase level studied in the other hospital before she was referred to our hospital was higher. The only difference was that it was the third attack of pancreatitis. Even though the previous attacks were evaluated in another hospital, it was learned from the records that amylase levels were between 400 and 450 U/L during attacks, which decreased below 100 U/L after. Additionally abdominal ultrasound had revealed edematous pancreas.

Solid pseudopapillary tumor may be diagnosed by US, CT or MRI (8). The US findings are well-defined solid masses, cystic masses or both cystic and solid masses (7). Abdominal CT or MRI reveals a well-circumscribed cystic mass with internal septa and solid portions (13). Hemorrhage may also be defined in MRI findings (8).

The localization of tumor might be tail (35.9%), head (34%), body (14.8%), body and tail (10.3%) (12). Median tumor size is 6 cm (range 1.2-6 cm) (3,7,13). In our case the tumor was located in the tail and 4 cm in size. Complete resection of the tumor was performed and the histological findings were consistent with solid pseudopapillary tumor being 1-antitrypsin positive which is a common positive marker (83%) for this tumor (2). Complete surgical resection is suggested to be curative and recurrence is rare (2-4). However because metastasis is reported, patients should be followed-up with imaging procedures (8). Metastasis may occur in liver, peritoneum, portal vein, spleen, duodenum, omentum, colon, lung, peritoneum, vessels and regional lymph nodes (7,8).

In this report, we presented a twelve-year-old girl with recurrent pancreatitis. All other etiologic factors like familial, hypertriglyceridemia, hypercalcemia, cystic fibrosis, medications were excluded. Solid pseudopapillary tumor of the pancreas was the only predisposing factor found. Although acute pancreatitis due to SPT was exceptionally reported (2), this is the first description of SPT leading to recurrent pancreatitis especially in children. The mechanism of SPT causing acute pancreatitis is not clear. It was proposed that non-traumatic internal bleeding may be leading to rapid expansion of the tumor and this may cause acute pancreatitis by ischemia, distension and duct obstruction (4). This could be the reason of recurrent attacks in our patient.

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