Tumors derived from all three germ layers are designated as teratomas. They are true neoplasms arising from totipotential cells and are composed therefore of numerous types of tissues. Cystic teratoma of the pancreas—also called a dermoid cyst—is a very rare entity. Only a few cases have been reported in the world literature. Most of these patients were young and had presented with symptoms due to the compressive effect of the tumor on surrounding structures. The imaging findings are not pathognomonic and the complete surgical excision of the tumor is the treatment of choice. We describe a new case of this rare tumor and review the previous cases. Optimal treatment of this tumor is possible only if it is recognized preoperatively.

Case
In December of 1998, a 16-year male was admitted to Sina hospital, Tehran, Iran with a 2-month history of progressive jaundice and non-significant weight loss of about 2 to 3 kg during this period. The physical examination revealed icteric conjunctiva and skin, and the liver and spleen extended 8- and 1.5-cm below costal margins, respectively.

Laboratory investigations showed a microcytic-hypochromic anemia that later proved to be a sign of minor thalassemia. Liver function test results were suggestive of an obstructive jaundice (total bilirubin=21.1 mg/dl, direct bilirubin=5.2 mg/dl, SGOT=152 IU/L, SGPT=129 IU/L, ALKP=1205 IU/L, LDH=350 IU/L, PT=11.9 seconds and APTT=43 seconds. Uralysis was positive for bile (2+) and urobilinogen. ESR, CRP and the indirect Coombs's test were negative and the reticulocyte count was 1.5%. At ultrasonography, the liver span was 18 cm (at midline) with a dilated common bile duct (10-13 mm), and dilated intra/extrahepatic ducts. The spleen was homogenously enlarged with a span of 16 cm and the distended gall bladder was filled with sludge. Percutaneous transhepatic cholangiography confirmed that a pressure effect on the common bile duct had impaired its filling (Figure 1). Endoscopic retrograde cholangiopancreatography (ERCP) showed that pancreatic and choledochal ducts were parted due to a large mass, while being compressed because of a pressure effect (Figure 2). In axial sections of a contrast-enhanced abdominal CT scan, a hypodense cystic mass with sharp borders and containing dense and calcified opacities appeared at the head and the uncinate process of the pancreas (Figure 3).

Pancreatic adenoma, mucinous carcinoma, pseudocyst and finally cystic teratoma of the pancreas were put forward as differential diagnoses so the patient was prepared for an
abdominal operation. At laparotomy, a large cyst at the head of the pancreas with tight adhesions to liver hilum was discovered. During dissection in the porta hepatis, the choledochal duct was disrupted, the cyst ruptured and a large volume of secretions and hairy material, in addition to two teeth were emptied. A choledochoduodenostomy was inevitably performed and the peritoneal cavity was irrigated and drained with a large volume of sterile saline.

Gross pathological examination of the cyst proved the diagnosis of mature teratoma of the pancreas (Figure 4). The tumor contained hair and histological elements of the skin appendages (Figure 5).

**Discussion**

Cystic teratomas are true neoplasms of germ cell origin that occur in preaxial, median, or paramedian positions such as the testes, ovaries, the sacrococcygeal region, mediastinum, retroperitoneum and skull. The pancreas is an extremely rare site for a teratoma with very limited number of cases reported up to now (Table 1). Like other extragonadal teratomas, those in the pancreas probably originate from aberrant germ cells arrested during migration to the gonads early in embryonic life. They are found in any part of the pancreas and are recognized by their content of hair or sebaceous material together with epithelial, mesenchymal or neural tissue within the cyst wall. The tumors are variably described as mobile or fixed, firm or cystic, and smooth or nodular. They are benign; but significant adherence to vital surrounding structures occurs. Calcification of the wall and contents of the cyst may occur, resulting in a mistaken pre-operative diagnosis of an echinococcus cyst.

Both sexes are affected and most patients are young people who present with abdominal mass, backache, dyspepsia or
Table 1. Review of cases with cystic teratoma of pancreas (Reproduced from Ref.4,7).

| Age/SEX | Symptoms | Signs | Location of cyst | Treatment | Follow-up |
|---------|----------|-------|------------------|-----------|-----------|
| 55/F45  | Epigastric swelling | Mass, RUQ | Head of pancreas | Ext. drainage, marsupialization | Partial tumor resection and follow-up 2 months later |
| 33/F45  | Lumbar pain | Mass, LUQ | Tail | Resection | Cholecystectomy after 2 years |
| 40/M4  | Lumbar pain | Mass, RUQ | Head | Ext. drainage, marsupialization | Persistent fistula after 1 year |
| 2/F45  | Vomiting, dehydration | Mass, epigastrium | Body | Resection and drainage | Uneventful |
| 2/F45  | Liver failure | Mass, epigastrium | Head | Resection | Well after 6 months |
| 8/M45  | Abdominal pain, vomiting, fever | Tender mass, LUQ | Body | External drainage | Well after 7 months |
| 6/M45  | Abdominal pain, vomiting | Mass, LUQ | Tail | Distal pancreatectomy | Well after 6 month |
| 34/F4  | Pain | Mass, LUQ | - | Cystogastrostomy | - |
| 4/F4  | Pain, vomiting | Mass, epigastrium | - | Ext. drainage | - |
| 11/F45 | Asymptomatic | Mass, LUQ | Body | Excision | Uneventful |
| 21/M45 | Painless mass, nausea, constipation | Mass, LUQ | - | Superior mesenteric vein ligation, biopsy and drainage | Tumor resection after 3 months |
| 25/F4  | Abdominal pain | RUQ tenderness | Head | Excision | Well after 14 years |
| 74/M4  | Backache | Mass, LUQ | Body | Distal splenopancreatectomy | Asymptomatic without recurrence at 2-year follow-up |
| 44/M6  | Pain | RUQ | Head | - | - |
| 2/M7  | Incidental abdominal mass | Epigastric mass | Head and body | Excision | - |
| 16/M*  | Progressive icterus | Hepatosplenomegaly | Head and body | Excision, choledochoduodenostomy | Well after 4-year follow-up |

obstructive gastrointestinal disturbances like the present case.4 Abdominal x-rays occasionally show calcification or displacement of the interabdominal organs.4 Findings at ultrasonography and computer tomography are particularly helpful, but have no pathognomonic significance. In fact, pathological examination is required for definitive diagnosis.5 Ultra-sound will show the pancreatic cystic teratoma as a dishomogenous mass with a thin capsule4 and solid areas with acoustic shadowing.5 Computed tomography may further define small loculations with sharp interfaces and calcifications.5

The differential diagnosis includes all other cystic lesions of the pancreas, both benign and malignant (serous and mucinous) cystadenomas, papillary cystic neoplasms and pancreatic cysts.4 In pathological examination, teratomas are made up of a variety of parenchymal cell types, representative of more than one germ layer, usually all three. They arise from totipotential cells, which differentiate along various germlines, producing, for example, tissues that can be identified as skin, muscle, fat, gut epithelium, tooth structures, or indeed, any tissue of the body.6 In the recent years, FNA cytology has proven to be a valuable diagnostic adjunct in the preoperative evaluation of a patient with a dermoid cyst of the pancreas.7 Cystic teratomas are not expected to heal completely after external drainage or marsupialization alone, since the
cystic wall is lined by a secreting epithelium. Therefore, whenever a cyst of the pancreas is suspected pre- or intraoperatively to be a teratoma, total resection should be carried out. Only if the patient's condition is unfavorable or total excision is technically impossible, should other surgical procedures, such as partial excision and external drainage, be performed. We emphasize that “coming to think of cystic teratomas of pancreas” is the main clue to the correct and timely diagnosis of these tumors.

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