

Mature cystic teratoma of the pancreas with 30 years of clinical course

A case report

Zhi Li, MS, Nengwen Ke, MD, PhD, Xubao Liu, MD, PhD, Shu Gong, MS*

Abstract

Rationale: Mature cystic teratoma of the pancreas is an extremely rare condition that is challenging to manage due to inadequate preoperative histological diagnosis.

Patient concerns: A 36-year-old female was admitted to our hospital because of a complaint of worsening paroxysmal abdominal pain for over a year. She was diagnosed with pancreatic pseudocyst and underwent five surgical procedures (i.e., four external drainage and one internal drainage procedures) in the last 30 years.

Diagnoses: Intraoperative frozen section analysis identified the tumor as a cystic teratoma, and pathological evaluation confirmed it to be a mature cystic teratoma.

Interventions: Explorative laparotomy was performed, and the tumor was then resected completely through with partial pancreatectomy, left colon resection, partial gastrectomy, duodenectomy, as well as inferior vena cava and renal vascular repair.

Outcomes: The patient exhibited a generally good wellbeing without any recurrence during the 6 months of follow up.

Lessons: This case demonstrated the significant impacts of misdiagnosis and inappropriate treatments for benign disease on the patient's quality of life and highlighted the importance of early surgical resection with a definitive pathological diagnosis for pancreatic cystic teratoma.

Abbreviations: CT = computed tomography, EUS = endoscopic ultrasound, FNA = fine needle aspiration, MRI = magnetic resonance imaging, QoL = quality of life.

Keywords: pancreatic cyst, pancreatic pseudocyst, teratoma

1. Introduction

Pancreatic pseudocyst is a common gastrointestinal disorder that must be distinguished from other cystic diseases, such as cystadenoma and cystadenocarcinoma. Primary mature cystic teratoma is a rare neoplasm characterized by the inclusion of well-differentiated parenchymal tissues.^[1] Mature cystic teratoma is commonly found in the ovary and testes, but it is extremely rare in the pancreas.^[2] In the present study, we reported a unique case of mature cystic teratoma of the pancreas that had been misdiagnosed and mistreated as pancreatic pseudocyst for 30 years.

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Department of Pancreatic Surgery, West China Hospital of Sichuan University, Chengdu, China.

* Correspondence: Shu Gong, Department of Pancreatic Surgery, West China Hospital of Sichuan University, No. 37, Guo Xue Land, Wuhou District, Chengdu 610041, China (e-mail: gdy-29@163.com).

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2. Case presentation

A 36-year-old female was admitted to our hospital because of a complaint of worsening paroxysmal abdominal pain for over a year. Thirty years ago, she was admitted to a local hospital because of abdominal pain. She was diagnosed with pancreatic pseudocyst through ultrasonography and subjected to an external drainage procedure. During the last 30 years, the symptoms recurred, and the patient underwent another 4 surgical procedures (i.e., 3 external drainage and 1 internal drainage procedures) after the first episode. One year ago, the patient complained of worsening abdominal pain and was admitted to our hospital.

Upon admission, the patient presented a generally poor condition. She complained of loss of sleep and poor appetite, and her weight reduced by 10 kg within a year. Her vital signs were as follows: heart rate, 78/min; blood pressure, 124/78 mm Hg; body temperature, 36.8°C; and respiratory rate, 18/min. Her body mass index was 16.2 kg/m². Physical examination revealed distension and tenderness in the upper abdomen. Assessed by using the European Organization for Research and Treatment of Cancer Quality of Life Questionnaire (EORTC QLQ-C30), the overall quality of life (QoL) score of the patient was only 16.7. Laboratory analyses indicated the following results: hemoglobin, 92 g/L (115–150 g/L); albumin, 34.7 g/L (40–55 g/L); glucose, 3.06 mmol/L (3.9–5.9 mmol/L); C-reactive protein, 74.8 mg/L (<5 mg/L); interleukin-6, 33.45 pg/mL (0–7 pg/mL); procalcitonin, 0.06 ng/mL (<0.046 ng/mL); CA19-9, 45.88 U/mL (<22 U/mL); and CA-125, 38.45 U/mL (<35 U/mL). Computed

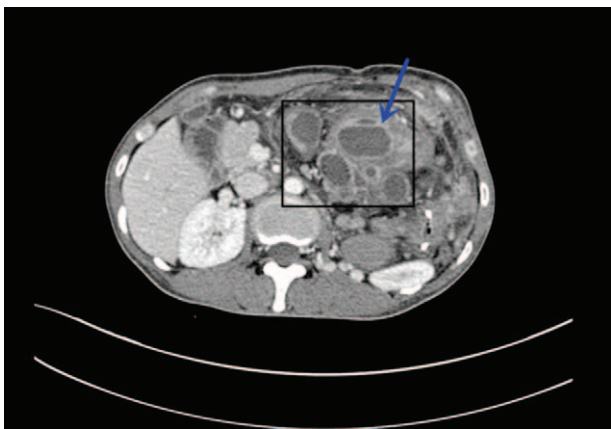


Figure 1. Computed tomographic scan showing several irregular cystic masses mainly surrounding the pancreatic body and tail (blue arrow).

tomography (CT) scan of the abdomen showed several irregular cystic lesions that mainly surrounded the pancreatic body and tail. The lesions included various sizes of soft tissue nodules, of which the largest one measured 3.6 cm × 2.5 cm and displayed a defined margin and partial obvious enhancement (Fig. 1). Magnetic resonance imaging (MRI) scan exhibited an area of mixed signals and patchy and nodular calcifications were identified (Fig. 2). The imaging features failed to define a diagnosis, but chronic inflammation changes with several pseudocysts or neoplastic disease were suggested.

In view of the long course and recurrence of the disease, the poor general condition, and the recently elevated levels of CA19-9 and CA-125, a diagnosis of pancreatic cancer was strongly suspected. Thus, an explorative laparotomy with possible distal pancreatectomy was performed. A huge mass with well-defined borders measuring 20 cm × 15 cm was intraoperatively found to have originated from the pancreatic tail, and severe adhesions were separated from the duodenum, the left colon, the uncinate process of the pancreas, and the gastric wall of the greater curvature. The cyst cavity contained many abscesses and nodules (Fig. 3). The tumor was resected completely through partial pancreatectomy, left colon resection, partial gastrectomy, duodenectomy, as well as inferior vena cava and renal vascular repair. Frozen section analysis identified the specimen as a cystic teratoma, and pathological evaluation confirmed it to be a mature cystic teratoma (Fig. 4). Histologically, the mass contained fibroadipose tissue, nervous tissue, enterogenous

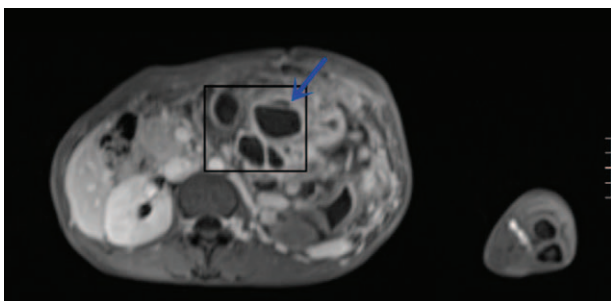


Figure 2. Magnetic resonance imaging exhibiting an area of mixed signals (blue arrow).

glandular epithelium, fibrous columnar epithelium, mucous gland, bronchial cartilage, bone, and bone marrow.

The patient recovered uneventfully and was discharged at day 11 postoperation. During the 6-month follow-up, the patient was asymptomatic and exhibited no signs of recurrence. At present, she has secured a job and gradually returned to normal life. The overall QoL score was 83.3.

The patient permitted for this case to be reported and informed consent had been obtained.

3. Discussion

The pancreas is one of the rarest primary sites for the development of cystic teratomas. We searched the *MEDLINE* and *EMBASE* databases and found only 50 reported cases of pancreatic cystic teratomas. Of the 50 cases, 27 were male and 23 were female. The mean age of the reported patients was 37.7 years old (range: 4 months to 74 years old). Thirty-four (68.0%) patients presented with symptoms. Most of the symptomatic patients complained of nonspecific gastrointestinal symptoms, such as abdominal pain, nausea, vomiting, anorexia, abdominal distension, back pain, and weight loss.^[3-6] The most common physical examination findings were a palpable abdominal mass with/without tenderness. Forty-five reports indicated the tumor location, and the most common location was in the body (16, 35.6%), followed by the head (15, 33.3%), tail (7, 15.5%), body/tail (4, 8.9%), and head/body (3, 6.7%). The sizes of the tumors ranged from 2.2 cm^[7] to 25 cm,^[8] and the median size was 8 cm.^[9] The laboratory values were normal except for the increased CA 19-9 levels in a few cases.^[3,10] In the presented case, the patient had a fair presentation in terms of age, sex, complaints, and tumor location. The patient had suffered a long duration of the disease, manifesting a relatively large mass and a poor general condition due to recurrent infections and insufficient nutrition.

Given that the symptoms and signs are nonspecific, imaging techniques, such as abdominal ultrasound, CT, or MRI, could aid the diagnosis of pancreatic cystic teratoma. The presence of fat, fat-fluid levels, and calcifications are considered highly suggestive indicators of a mature cystic teratoma^[11]; however, these conditions occurred in only a minority of cases. Most imaging findings were nonspecific and had sufficient overlap with other pancreatic cystic lesions, such as pseudocysts, solid pseudopapillary tumors, serous or mucinous cystadenomas, or intraductal papillary mucinous neoplasm.^[10,12]

The preoperative diagnosis of pancreatic cystic teratoma remains difficult. In our case, the patient said that she was first diagnosed with pancreatic pseudocyst through ultrasound. However, the images cannot be obtained as 30 years had passed. Recent CT and MRI features were still nonspecific, thereby preventing radiologists from performing an accurate consultation. Instead, in view of the long courses, recurrent infections, poor nutrition status, and newly elevated levels of tumor makers, a preoperative diagnosis of pancreatic cancer was strongly suspected this time. Recently, fine needle aspiration (FNA) guided by endoscopic ultrasound (EUS) has been shown to be a safe and cost-effective technique to detect pancreatic masses, including solid tumors, cystic neoplasm, pseudocyst, and reactive change. It seems a promising tool to lead to definite preoperative diagnosis and may avoid unnecessary surgery for benign neoplasm.^[10,11] However, FNA is not conclusive for defining the nature of the pancreatic lesions, especially in presence of larger ones (>3 cm).^[13] Moreover, there is still controversy regarding the risk of cancer seeding outweigh the potential benefits of this method.^[3]

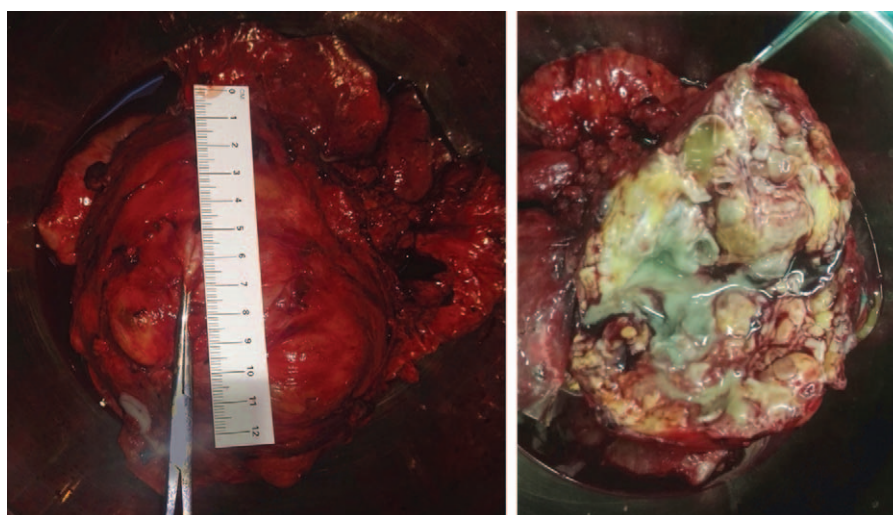


Figure 3. Macroscopic appearance of a resection specimen.

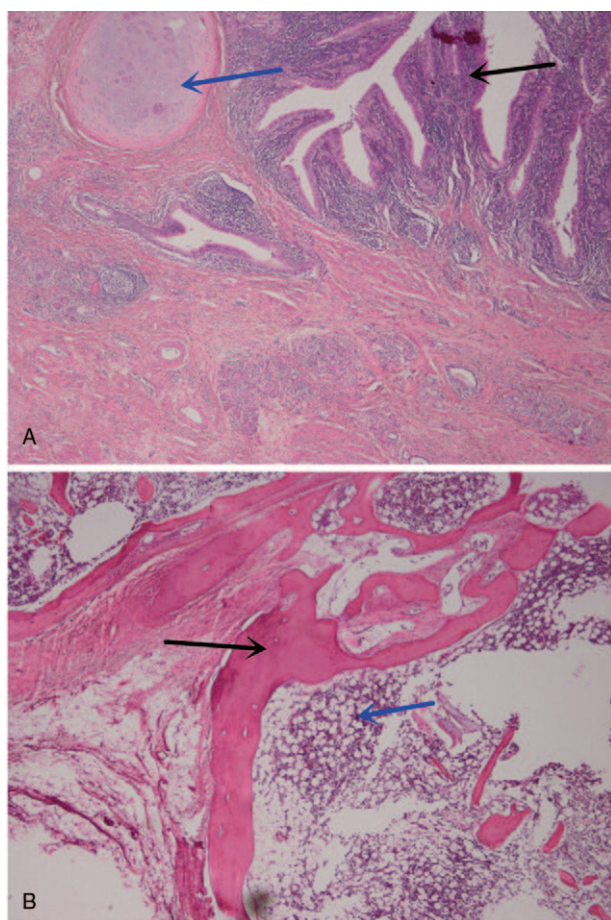


Figure 4. Pathological findings of the cystic lesion. (A) Pseudostratified ciliated columnar epithelium (black arrow) and bronchial cartilage (blue arrow; hematoxylin–eosin stain; original magnification, $\times 40$). (B) Bone (black arrow) and bone marrow hematopoiesis (blue arrow; hematoxylin–eosin stain; original magnification).

We did not perform EUS-FNA in this case because the patient had clear operative indications assessed by our surgeon, and we chose to avoid the potential risk of cancer seeding along the biopsy tract because malignancy appeared to be more likely than a benign cystic tumor in our case.

To treat the suspected pancreatic pseudocyst, the patient underwent 5 times of external/internal drainages in the last 30 years. However, none of these procedures solved the problem. In contrast to pancreatic pseudocysts, pancreatic cystic teratomas are mainly treated by surgical resection. A simple excision of the lesion is possible because of its benign nature. Thus, this patient lived a poor QoL in the last 30 years due to the recurrent seizure of abdominal pain and inappropriate treatments. Although the preoperative diagnosis of pancreatic teratomas is difficult, an early surgical intervention with a definitive pathological diagnosis is very important.

It is noteworthy that in our case, the patient had a long course of disease and extremely poor QoL despite its benign nature. We considered several reasons. First, imaging techniques were limited 30 years ago, which might account for the initial misdiagnosis and inappropriate treatment. Second, the symptoms and the imaging features were nonspecific and could have changed after repeated external and internal drainage procedures, thereby contributing to the difficulties for definite diagnosis. Third, the history of multiple operations increased the risk for reoperation, which might delay the effective treatment again.

4. Conclusion

We reported a unique case of pancreatic mature cystic teratoma with long disease duration and high operation risks. This case demonstrated the significant impacts of misdiagnosis and inappropriate treatments for benign disease on the patient's quality of life and highlighted the importance of early surgical resection for pancreatic cystic teratoma. We hope clinicians could increase the awareness of the possibility of cystic teratoma when performing a differential diagnosis of pancreatic cystic lesions and avoid misdiagnosis through our case.

Author contributions

Supervision: Xubao Liu.

Writing – original draft: Zhi Li.

Writing – review and editing: Nengwen Ke, Shu Gong.

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