CASE REPORT

Pancreatic Sarcoma Mimicking Pseudocyst After Pancreatitis: A Case Report and Review of the Literature

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A 45-year-old female presented with abdominal pain for 3 days without trauma history. The pain was located over the epigastric area with radiation to the back. A pancreatic pseudocyst was impressed initially according to clinical symptoms, laboratory data, and image studies. However, rapid progression of the lesions was noted later after discharge. Following image studies showed circumscribed tumors in the head, body, and tail regions of the pancreas. Surgery with the Whipple operation was performed during the second admission. The post operation course was uneventful. Pathologic results showed sarcoma of the pancreas. Primary pancreatic sarcomas are extremely rare lesions accounting for < 0.1% of all pancreatic malignancies. Pancreatic sarcomas tend to be aggressive and have a poor prognosis. In addition, < 5% of pancreatic tumors are cystic; the rarest cystic neoplasm is also the primary pancreatic sarcoma, with only a few cases having been documented. However, it should be considered in the differential diagnosis of pancreatic cystic lesions.

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Introduction

Sarcomas represent a relatively rare malignant entity. Primary sarcomas of the pancreas are even rarer [1]. They comprise < 0.1% of malignant pancreatic neoplasms, with only a few cases previously documented in English literature [2]. Fewer than 5% of pancreatic tumors are cystic;
microcystic cystadenoma, and macrocystic cystadenocarcinoma are the most common. The rarest cystic neoplasm is the primary pancreatic sarcoma [3]. These tend to be more aggressive and have a poor prognosis. We present a patient with such a rare condition with pancreatic sarcoma initially mimicking a pseudocyst.

Case report

A 45-year-old female suffered from epigastric pain with radiation to the back for several days. When she came to our outpatient department for help, elevated amylase and lipase levels were noted (476 U/L and 1239 U/L, respectively). She was then admitted to a ward in Cheng-Ching General Hospital, Taichung, Taiwan for further evaluation and management under the impression of acute pancreatitis.

After admission, an abdominal sonogram revealed: moderate fatty liver, a cystic lesion 2 cm in the pancreatic head, and another lesion ~2.2 cm in the body of the pancreas (see Fig. 1). Differential diagnosis included: pseudocyst, cystic tumor or hypoechoic tumor of pancreas. Computed tomography (CT) of the abdomen was done and it showed: two cystic lesions and two suspected pseudocysts in the pancreatic head and body. Tumor markers were as following: Carcino Embryonic Antigen (CEA) 0.77 ng/mL (< 5.0) and CA19-9 17.65 U/mL (< 37). After medical treatment, the abdominal pain improved gradually. She was discharged 5 days later. After discharge, she was followed regularly at our outpatient department. Unfortunately, an abdominal sonogram 2 months later showed increased sizes of previous pancreatic lesions. Abdominal CT followed and the results showed: four circumscribed tumors with irregular wall thickening 3.4 cm in the head, 3.2 cm and 3 cm in the body, and 1.2 cm in the tail region of pancreas. The patient was then admitted again for further evaluation and management.

The patient denied a history of systemic disease or major operations. She did not have a habit of cigarette smoking or alcohol drinking. No previous family history of malignancy was noted. After admission, the patient appeared ill looking. She was conscious, lucid, and cooperative. Vital signs were as follows: blood pressure 128/76 mmHg, pulse rate 88/minute, respiratory rate 18/minute, and temperature 36.2°C. Her sclera was not icteric and the conjunctiva was not pale. Chest examination showed clear breathing sounds over both lung fields. Observation of the heart showed a regular heartbeat without murmur.

Distension of the abdomen was present and mild abdominal tenderness over the epigastric area was noted.

Laboratory data showed that the white blood cell count was 7180 per microliter without left shifting of segment 70%, hematocrit 38.9% (34–44%), hemoglobin 13.2 g/dL (12–16 g/dL), sodium 138 mmol/L (135–148 mmol/L), potassium 3.6 mmol/L (3.5–5.0 mmol/L), blood urea nitrogen 8 mg/dL (7–20 mg/dL), creatinine 0.8 mg/dL (0.5–1.5 mg/dL), Aspartate Aminotransferase (AST) 19 U/L (5–40 U/L), Alanine Aminotransferase (ALT) 28 U/L (3–35 U/L), alkaline phosphatase 124 U/L (35–104 U/L), total bilirubin 0.3 mg/dL (0.2–1.2 mg/dL). Tumor marker levels examined after admission including alpha-fetoprotein (AFP) 3.75 ng/mL (< 8.78), CEA 0.69 ng/mL (< 5.0), and CA19-9 9.32 U/mL (< 37) were within normal limits. The chest roentgenogram showed normal lung fields. The electrocardiogram showed normal sinus rhythm. On the 2nd day of admission, an operation was performed after discussion with the patient and her family. The Whipple operation was performed smoothly. Partial pancreatectomy, partial gastrectomy, and splenectomy were done (see Fig. 2).

The operative findings showed many endurated mass lesions locating over the pancreatic head with extension to neck, and body. The pathologic report showed sarcoma involved all of the pancreatic lesions and pancreatitis of the pancreas. The microscopic finding was: French Federation of Cancer Centers Sarcoma Group (FNCLCC) Grade II sarcoma composed of hyperchromatic spindle cell with mitosis (10-19/10HPF), pancreatitis with lymphocytic infiltration, focal fibrosis, and fat necrosis. Immunohistochemical stain: CD117 (–), smooth muscle actin (+), Cytokeratin (CK) (+), S100 (–), desmin (–), and vimentin (+). The post operation course was uneventful and she was discharged 15 days after the operation. Her condition remains stable after 12 months of follow up from the time she was discharged (see Fig. 3).

**Fig. 1** Abdominal ultrasonogram during the first admission. The sonogram shows a cystic lesion 2 cm in the body of the pancreas.

**Fig. 2** Abdominal sonogram after the first admission. The sonogram shows increased sizes of cystic lesions with irregular wall thickening of the pancreas.
Discussion

Sarcomas represent a relatively rare malignant entity. Primary sarcomas of the pancreas are even rarer. Amongst pancreatic sarcomas, leiomyosarcomas tend to occur relatively frequently [1]. Other sarcomas of the pancreas reported include: fibrosarcoma, carcinosarcoma, rhabdomyosarcoma, liposarcoma, hemangioendothelioma, malignant neurilemmoma, malignant hemangiopericytoma, and malignant fibrous histiocytoma [3]. Yet, they comprise only 0.1% of malignant pancreatic neoplasms [2]. Pancreatic sarcomas occur frequently in younger individuals. The pancreatic caput is most commonly involved followed by the tail and the body [1]. Patients present clinically with epigastric abdominal pain, nausea, and vomiting. Some may have body weight loss and the presence of an abdominal mass. In the present case, the patient had only the same symptoms as pancreatitis but she did not have significant weight loss.

The diagnosis of pancreatic cancer and pancreatitis, especially chronic pancreatitis, is notoriously problematic, in that both diseases may have unspecific clinical symptoms [4]. Ultrasonography (US)-based diagnosis of small pancreatic tumors was based on the identification of a poorly reflecting and attenuating pancreatic mass. The larger tumors were more heterogeneous echogenically and exhibited well-defined irregular or lobulated margins [5]. US usually reveals a well-circumscribed hypoechoic mass with heterogeneous inner echoes. A CT scan may show a solid tumor with heterogeneous density with or without a cystic component. Some reports on the radiological aspects also describe central areas of low attenuation on CT that present cystic degeneration and necrosis of the tumor [6,7]. Our findings in the present case are consistent with some of the above descriptions such as cystic degeneration of the tumor.

The differential diagnosis of a cystic pancreatic lesion includes pseudocyst, retention cyst, microcystic adenoma, mucinous cystic neoplasm, papillary cystic tumor, cystic islet tumor, nontumoral cyst in association with polycystic disease, vascular tumors, cystic lymph node metastasis (from primary cancer of the lung, ovary, or melanoma), and primary hydatid disease of the pancreas [3]. The pancreas is a rare location for a sarcoma and tumors arising from other sites, such as retroperitoneum, female genital tract, and soft tissues, may involve the pancreas, simulating a primary pancreatic lesion [2]. In the present case, both imaging studies and intraoperative impression ruled out an extrapancreatic origin. Some suggest markedly shaggy and thickened wall without definite cysts on CT should exclude a pancreatic pseudocyst and other cystic tumors [3].

A correct preoperative diagnosis of cystic masses, arising from the pancreatic or peri-pancreatic region, is important for appropriate treatment. Imaging modalities including CT, magnetic resonance imaging, and endoscopic US have proven to be useful in assessing resectability. Surgical resection is the only possible cure for pancreatic malignancies. With respect to a pancreatic sarcoma, a definite therapy recommendation is hard to make. Nonetheless, radical resection seems to offer the best possibility for cure [2]. Therefore, we performed aggressive surgical intervention after explanation to the families. Fortunately, the postoperative recovery was uneventful.

Pancreatic cancers usually have a poor prognosis because they tend to be diagnosed in an advanced stage. Pancreatic sarcomas tend to grow much more rapidly and are believed to be associated with an even worse prognosis [1]. Prognosis is influenced by patient age, tumor size, presence of tumoral necrosis, and vascular invasion [2]. In the present case, the prognosis seems not bad. This may be due to the following reasons: resectability of the tumors, less tumor necrosis, and no vascular invasion. In addition, the patient was not old.

Special attention should be directed to the predominant solid component of the mass to avoid misdiagnosing these lesions as pseudocysts. In addition, the presence of a pseudocyst does not exclude the possibility of a coexisting malignant tumor of the pancreas. Some suggest positron emission tomography scans may be useful in tumors with liquefying necrosis that mimic pseudocysts or abscesses on a CT scan [4].

In conclusion, pancreatic sarcomas are extremely rare. They appear to be more aggressive and are associated with a worse prognosis [1]. We should consider a pancreatic
tumor including sarcoma in the differential diagnosis of a pancreatic cystic lesion in a patient with pancreatitis. Awareness of the possibility may permit earlier diagnosis, improve the management, and enhance the chance for cure.

References


