

## CASE REPORT

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# Recurrent Pancreatitis Due to a Cystic Pancreatic Tumor: A Rare Presentation of Acinar Cell Carcinoma

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### ABSTRACT

**Context** Acinar cell carcinoma is an uncommon malignancy of the pancreas. It has characteristic histomorphology, immunohistochemistry profile, and clinicopathological behavior.

**Case report** We report a rare case of recurrent pancreatitis secondary to acinar cell carcinoma of the pancreas. We describe the endoscopic ultrasound characteristic, treatment and the surgical outcome.

**Conclusions** Acinar cell carcinoma should be considered in the differential diagnosis of cystic pancreatic tumors presenting with recurrent pancreatitis.

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### INTRODUCTION

Acinar cell carcinoma of the pancreas is a rare pancreatic tumor, accounting for approximately 1% of pancreatic exocrine tumors [1]. Clinical presentation is usually non-specific unless tumor is associated with syndromes related to excessive lipase production. Unlike ductal adenocarcinoma, this tumor rarely presents with pancreatitis. Data regarding course, treatment, and prognosis of this tumor is generally lacking. Tumor arises from the acinar cells and generally carries a better prognosis than

ductal adenocarcinoma. We report, for the first time, a 68-year-old man with recurrent attacks of pancreatitis with a cystic tumor of the pancreas who was found to have acinar cell carcinoma.

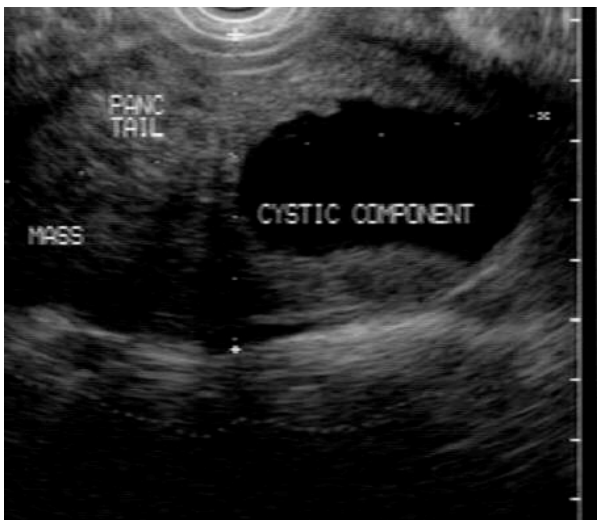
### CASE REPORT

A 68-year-old man was admitted to the general medical service with two days history with epigastric abdominal pain. Pain was moderate in severity, radiated to the back and aggravated by eating. Physical exam showed moderate epigastric tenderness but no active peritoneal irritation signs. Initial laboratory evaluation showed normal hemoglobin, normal white blood cell count, and normal platelets. Serum lipase and amylase were elevated. Computed tomogram (CT) scan of the abdomen confirmed a diagnosis of pancreatitis, in addition to a suspicious mass lesion of the pancreatic tail (Figure 1). Patient was managed conservatively with complete recovery and normalization of pancreatic enzymes. Past medical history was only significant for hypertension, and one attack of pancreatitis two years before this admission. He was admitted to another facility at that time and was treated conservatively for three days with full recovery. Records from that admission included a CT scan showing minor abnormality of the pancreatic tail with possible pseudocyst formation. Serum lipase levels were elevated at time of admission and

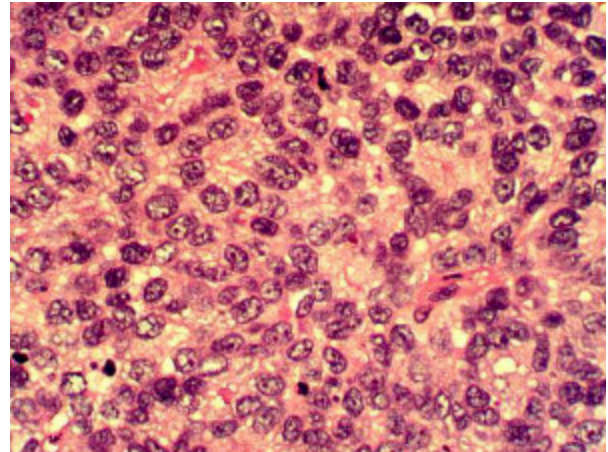


**Figure 1.** Axial contrast-enhanced CT. Yellow arrow indicates central area of fluid density. Green arrows outline the pancreatic mass.

continued to be abnormal through the past two years with no symptoms. He denied any change in appetite or weight loss. There was no history of skin abnormalities, deep vein thrombosis, or unexplained fever. He never drank alcohol and he did not smoke. Family history was negative for any gastrointestinal or pancreatic malignancies. Four weeks after discharge, patient was seen in the clinic for further evaluation of the pancreatic mass. EUS confirmed the presence of a seven cm mixed echogenicity mass without any lymph node enlargement (Figure 2). Patient was taken to the operative room where en bloc resection of the tumor was performed. Histopathology confirmed the diagnosis of acinar cell carcinoma (Figure 3). Regional



**Figure 2.** EUS showed large mass with both cystic and solid components.



**Figure 3.** Acinar cell carcinoma. High magnification showing the acinar architecture (H&E, x400).

lymph nodes were positive for tumor with positive margins in the resected specimen. Because of the extent of the disease, patient received postoperative adjuvant chemotherapy (5-Fluorouracil) and radiation therapy (total 50.4 gray). Patient continues to do well one year after his surgery with no evidence of recurrence.

## DISCUSSION

Acinar cell carcinoma makes up approximately 1% of exocrine pancreatic tumors [1]. It is classically known for the systemic manifestations related to liberation of lipase [2]. There is a very limited data regarding this tumor, clinical behavior, EUS findings and treatment outcome.

Most cases of acinar cell carcinoma are asymptomatic. It predominantly affects white elderly men, with a peak age of incidence in the seventh decade (mean age 62 years) [3]. Few cases are reported in children. Commonest clinical presentation is weight loss (50%), abdominal pain (32%), nausea and vomiting (20%), and elevated lipase levels (16%) [1]. Jaundice is uncommon even with tumors involving the head of the pancreas. Functional acinar cell carcinoma can present with several characteristic clinical syndromes. Patients with elevated lipase levels are at risk of developing the systemic manifestations with subcutaneous fat necrosis (panniculitis) and polyarthritis. Patients with

this hyperlipasemia related syndromes frequently have disseminated disease at the time of presentation. Endocrine manifestations are another unique but uncommon feature of acinar cell carcinoma. Hypoglycemia can occur secondary to tumor secretion of insulin, insulin-like growth factors and glucagon [4]. Also, acinar cell carcinoma can be associated with increased levels of tumor markers such as alpha-fetoprotein and carcinoembryonic antigen, a unique feature among the pancreatic exocrine tumors [5].

Acinar cell carcinomas commonly involve the head or the tail of the pancreas (56 and 36% respectively). These tumors are usually circumscribed, non-calcified fleshy tumors. They are usually large with average diameter of 10.8 cm. Large tumors tend to have multiple areas of hemorrhage and necrosis. Cystic changes can be seen but are not a prominent feature of these tumors [1].

The microscopic features of acinar cell carcinoma include acinar or trabecular arrangement of cells that contain cytoplasmic granules. A capsule often surrounds these tumors. Focal capsular invasion is seen in most of these cases, with finger-like projections extending into adjacent pancreatic parenchyma. Vascular invasion occurs in 2/3 of patients and perineural invasion in 32% of cases. Acinar cell carcinoma is a cellular tumor with minimal desmoplastic reaction. Thick fibrous bands will separate the tumor cells into lobules. The tumor cells grow into four distinct patterns of growth: acinar, cellular, glandular, and trabecular. Most of these tumors will present with the first two patterns.

Acinar cell carcinoma has a characteristic immunohistochemical staining. Acinar cells stain strongly positive for enzymes such as trypsin, chymotrypsin, lipase and amylase. Positive trypsin staining is seen in 100% of cases, positive lipase in 77% of cases, while chymotrypsin and amylase are positive in approximately one third of cases. Other common histochemical stains that can be used include butyrate esterase and d-periodic acid schiff.

There is limited data regarding the imaging features of acinar cell carcinoma. Most of these tumors appear as well circumscribed, homogenous or heterogeneous mass with minimal biliary obstruction. CT scans show an intense enhancement during the arterial-dominant phase of imaging correlating with the hypervascular nature of these tumors [6]. Areas of necrosis and hemorrhage can also be seen. Magnetic resonant imaging (MRI) is another modality that can be used. Mangafodipir trisodium (Mn-DPDP) enhanced MRI may be superior to CT imaging in visualizing these tumors especially they are functional [7]. MRI may also be helpful in showing tumor thrombus in cases with splenic or portal vein thrombosis. EUS is an excellent modality to visualize the pancreas. The features of acinar cell carcinoma on EUS have not been clearly defined [8]. In our case, EUS findings are shown in Figure 2. The tumor showed as a large mass with both cystic and solid components.

Treatment choice depends on the stage of the disease at time of diagnosis. Surgical resection is the best treatment in absence of distant metastasis. The extent of resection depends of the extent of the disease. En bloc resection has been reported in patients with locally advanced disease [9]. Adjuvant chemotherapy and radiotherapy may help to prolong survival but most of the data is coming from case reports [10]. Intraperitoneal chemotherapy with cisplatin has been used for peritoneal metastasis. 5-Fluorouracil is the commonest chemotherapeutic agent to be used. Gemcitabine has been used in few case reports with decent outcomes [10]. In our case, the tumor was locally advanced at time of surgery and was managed with en bloc resection followed by adjuvant chemotherapy (5-Fluorouracil) and radiation therapy with excellent outcome.

Acinar cell carcinoma carries a bad prognosis. The mean survival is approximately 18 months. The 3-year survival is 26% and 5-year survival is only 6%. The prognosis of these tumors is better than ductal carcinomas but worse than islet cell tumors. Half of the

patients will have distant metastasis at time of the diagnosis, and an additional 25% of patients develop metastasis later in their course. Predictors of poor prognosis include: age greater than 60 years, elevated serum lipase, and tumors larger than 10 cm [1].

Our case has some unique features. The patient presented with two attacks of confirmed pancreatitis over a two years period; the persistent elevation of serum lipase was not associated with any of the classical symptoms that accompany hyperlipasemia; and despite the presence of locally advanced disease at time of surgery, the patient continues to be tumor free after one year of follow up, confirming the validity of en bloc resection and adjuvant chemo and radiotherapy in selected cases.

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