Ciliated Enteric Duplication Cyst Presenting as a Pancreatic Cystic Neoplasm:
Report of a Case with Cyst Fluid Analysis

Michael R. Pins, Carolyn C. Compton, James F. Southern, David W. Rattner, and Kent B. Lewandrowski

Pancreatic cysts include inflammatory pseudocysts, cystic tumors (serous and mucinous), and various rare cystic lesions. We report a case of a ciliated enteric duplication cyst that presented on computed tomographic scan as a pancreatic cystic neoplasm. Cyst fluid analysis revealed markedly increased concentrations of carcinoembryonic antigen and CA 125 and increased fluid viscosity. These features are typical for a mucinous cystic neoplasm of the pancreas and demonstrate a potential pitfall associated with the diagnosis of pancreatic cysts by chemical analyses. Enteric duplications involving the pancreas are rare, usually of gastric origin, and usually communicate with their enteric source. Morphological and histochemical analysis suggest that this cyst was of caudal foregut origin. This is the first reported case of a ciliated foregut cyst involving the pancreas. The chemical characteristics of the cyst fluid of these lesions have not been described previously.

Additional Keyphrases: cancer · carcinoembryonic antigen · CA 125 · amylase

Most cystic lesions of the pancreas are inflammatory pseudocysts; 10% are cystic neoplasms. Cystic neoplasms are classified as serous (or microcystic) cystadenomas, mucinous cystic neoplasms, and mucinous cystadenocarcinomas (1, 2). Rare pancreatic cystic lesions include papillary cystic tumor (3), mucinous ductal ectasia (4), cystic islet cell tumor, ductal adenocarcinoma with cystic degeneration, lymphoepithelial cysts, peripancreatic cholesterol cysts, and cystic teratomas (5). Enteric duplication cysts involving the pancreas are rare and usually of gastric origin (6).

There are no reliable radiological or clinical criteria to permit preoperative differentiation of the various types of pancreatic cysts (7). In a review of cystic tumors from our institution, fully one-third of the cystic neoplasms had been previously misdiagnosed as pseudocysts, and many were inappropriately treated as a result. In this series, 40% of mucinous tumors were misdiagnosed as pseudocysts on the basis of clinical and radiological criteria (7). Chemical analysis of cyst fluid for carcino-

embryonic antigen (CEA) (8-11), CA 125 (12), amylase (7), and viscosity (12) has been proposed as a useful method for differentiating the different types of pancreatic cysts. Thus, cyst fluid obtained by percutaneous computed tomographic (CT) or ultrasound-guided needle aspiration may facilitate preoperative diagnosis. Among the various chemical analyses performed on pancreatic cyst fluids, only CEA and amylase contents have been consistently shown to be of value in differentiating among the various cyst types. CA 125 and cyst fluid viscosity may be useful diagnostic tests, but experience with these analyses is limited. CEA concentrations are typically high in mucinous cystic neoplasms and cystadenocarcinomas, low in serous cystadenomas and pseudocysts. The amylase content of pseudocysts is usually high but is variable (often low) in cystic tumors (7, 8). The chemical characteristics of rare types of pancreatic cysts have not been described. Therefore, these lesions may present problems in interpreting the results of cyst fluid aspirates.

We report a case of an enteric duplication cyst of probable foregut origin that presented clinically as a pancreatic cystic neoplasm. Analysis of the cyst fluid revealed marked increases in CEA, CA 125, amylase, and viscosity. This is the first report of a ciliated enteric duplication cyst involving the pancreas and the first description of cyst fluid contents in these lesions. The chemical features of this case were typical for a mucinous tumor, illustrating that results for cyst fluid aspirates from pancreatic lesions should be interpreted with caution.

Case Report

A 71-year-old man with a history of adenocarcinoma of the prostate presented with multiple trauma after a motor vehicle accident. During his evaluation, an abdominal CT scan demonstrated a 3.5-cm peripancreatic mass that was thought to represent a cystic pancreatic neoplasm. There was no history of pancreatitis. Angiograms of the celiac trunk, splenic, and superior mesenteric arteries failed to demonstrate the vascular supply to the mass. A distal pancreatectomy was performed.

Gross examination of the resection specimen revealed a 5.8 × 3.4 × 2.8 cm unilocular pancreatic cyst enclosed by a 0.1-cm-thick capsule. The viscous, yellow-tan fluid in the cyst was collected immediately after surgical resection. The lining of the cyst was smooth, and no
mucinous cystic neoplasms (benign or malignant) \(2\), serous (microcystic) cystadenoma \(1\), and various rare types of neoplastic, inflammatory, and congenital cysts. Although there is general agreement that mucinous tumors should be resected, some authors favor observation of asymptomatic serous cystadenomas (reviewed in 7). Asymptomatic pseudocysts are often treated expectantly; internal drainage is usually favored for those requiring therapy. The clinical and radiological features of pancreatic cystic lesions do not reliably discriminate among these possibilities. Chemical analysis of percutaneous cyst fluid aspirates for CEA has been advocated as a potentially useful preoperative test to differentiate mucinous tumors (high concentrations, CEA >25 \(\mu g/l\)) from pseudocysts and serous cystadenomas (low CEA concentrations) \(8-11\). Amylase concentrations may also help to differentiate pseudocysts (high, mean 13 132 U/L) from cystic tumors (variable, usually low) \(12,13\). CA 125 and relative viscosity of cyst fluid have been described, but the number of cases in the literature is limited \(13\). In our experience, CA 125 concentrations are high in malignant cysts (mean 10 064 units/L), low in pseudocysts (mean 18 units/L), and variable in benign mucinous and serous cystadenomas \(13\). Measurement of the relative viscosity of cyst fluid is fairly reliable (90% sensitive, 100% specific) in distinguishing mucinous tumors (high, >1.63) from pseudocysts and serous cystadenomas (low \(13\)).

One pitfall in evaluating pancreatic cyst fluid measurements is the lack of data concerning some of the rarer types of cystic lesions (e.g., cystic islet cell tumors, solid and cystic tumors, lymphoepithelial cysts, congenital cysts, and, in our case, enteric duplication cysts). Lack of familiarity with such lesions may lead to misinterpretation of the chemical findings for the cyst fluid. The cyst fluid in this case had markedly increased concentrations of CEA and CA 125 and high relative viscosity. Preoperative cyst fluid analysis, therefore, would have incorrectly indicated that the cyst was a mucinous cystic neoplasm (probably malignant). Although the high amylase concentration might suggest a pseudocyst, high values are occasionally seen in cystic tumors as well \(7,8,13\).

Enteric duplications are unusual congenital anomalies found most frequently in the ileocecal region but they may involve the gastrointestinal tract at any level. Enteric duplications involving the pancreas are rare and usually represent gastric alimentary duplications. We use the generic term enteric duplication cyst instead of alimentary duplication, reserving the latter term for congenital malformations that are connected to and share the same blood supply with their enteric origin \(14\). Alimentary duplications involving the pancreas are most likely the result of failure of regression of an enteric diverticulum formed from the pancreatic duct \(15\). Many of the ~90 reported cases of alimentary duplications involving the pancreas communicated with a pancreatic duct, were lined with gastric-type epithelium, and had ectopic pancreatic tissue within their walls. These patients usually presented at an early age.
with symptoms related to gastric acid secretion or pancreatitis (or both). The exclusive presence of respiratory-type epithelium and the lack of acid-producing gastric-type epithelium in our case may explain the absence of symptoms and the late presentation.

Three ciliated enteric duplication cysts involving the liver were recently described (16). Ours is the second reported case of an enteric duplication cyst isolated from its enteric source involving the pancreas and the first with respiratory-type epithelium. The previous case was a duplication cyst in a neonate, lined by gastric-type epithelium and separate from the stomach (17). Our case and the three ciliated hepatic foregut cysts reported by Terada et al. (16) were all small (<4 cm) unilocular cysts that had ciliated, pseudostratified, columnar epithelium. The cytoplasm of the epithelial cells in all four cases contained neutral, carboxylated, and sulfated mucin and was immunoreactive for CEA. This profile may be typical of duplication cysts of foregut origin involving the abdomen.

In conclusion, this case of a ciliated enteric duplication cyst presented radiographically as a cystic pancreatic tumor. Cyst fluid chemical analysis revealed a profile typical for a mucinous cystic neoplasm and suggestive of malignancy. Our findings suggest caution when interpreting results of cyst fluid analyses from pancreatic cysts.

References