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CASE REPORT

Pancreatic Mucinous Cystadenocarcinoma Presenting with Splenic Infarction in a Young Female

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Introduction

Cystic neoplasms of the pancreas account for a substantial proportion of pancreatic cystic lesions and about 1% of pancreatic malignancies [1]. Mucinous cystadenoma is a pancreatic neoplasm that is almost exclusively seen in females, with a mean age of presentation of 45 years [2]. Mucinous cystadenoma mostly (>90%) arises in the body and tail of pancreas and maybe unilocular or multilocular. Mucinous cystadenoma carries a risk for malignant transformation, and the incidence of carcinoma in situ (CIS) in resected mucinous cystic neoplasms (MCNs) is 25–50%. Pancreatic pseudocysts constitute a majority of cystic lesions of the pancreas, and it is important to distinguish pseudocysts from cystic neoplasms of the pancreas because of difference in management of these two clinical entities. Pancreatic cystic lesions are usually identified in patients who are asymptomatic or have nonspecific symptoms such as vague abdominal pain and weight loss, on abdominal imaging with computed tomography (CT) scan or magnetic resonance imaging (MRI) [3]. Endoscopic ultrasound (EUS) is useful for providing detailed imaging of cystic pancreatic lesions, because the endoscope can be placed close to the cystic lesion and guide fine-needle aspiration (FNA) of cystic fluid [4]. We report an unusual case of pancreatic mucinous cystadenocarcinoma presenting in a 39-year-old female with a remote history of acute pancreatitis, who presented with left upper quadrant abdominal pain. CT scan of abdomen and pelvis showed a unilocular 5.2×4.8 cm cystic lesion in the tail of pancreas suspected to be a pseudocyst, and an irregular area of low density within the posterior–inferior aspect of spleen consistent with splenic infarct. On endoscopic ultrasound, a large cystic lesion was seen and FNA was performed. Distal pancreatectomy, splenectomy and partial gastrectomy were performed. Histologic study confirmed the diagnosis of pancreatic mucinous cystadenocarcinoma. The patient had no postsurgical complications and was discharged with outpatient oncology follow up. The differentiation between pancreatic pseudocyst and cystic neoplasms of the pancreas is important because the treatment of these entities is quite different.

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Case Report

A 39-year-old Caucasian female presented with complaints of left upper quadrant abdominal pain for 2 months, with increasing intensity over the last 2 weeks prior to admission. The pain was characterized as dull initially, later becoming sharp, constant, nonradiating, with no aggravating or relieving factors. Abdominal pain was associated with nausea, vomiting, early satiety and a sensation of fullness in her abdomen. Her medical history was significant for acute pancreatitis in 2007 with formation of a pseudocyst, which resolved spontaneously, and peptic ulcer disease. She reported smoking a half-pack of cigarettes per day for 15 years, and had a history of drinking three to four glasses of alcohol daily which she quit 3 years ago. On physical examination, she had moderate tenderness to palpation in left upper quadrant, without rebound tenderness, guarding or organomegaly. The admission laboratory tests showed normal electrolytes, white cell count and normal liver function tests. Serum amylase was 132 U/l (normal range 36–128 U/l) and serum lipase was 220 U/l (normal range 8–50 U/l). CT of the abdomen and pelvis with contrast showed a low density cystic structure in the pancreatic tail measuring approximately 5.2×4.8 cm in the greatest

axial dimensions (Fig. 1). The lesion extended superiorly and posteriorly towards the splenic hilum and produced mass effect upon the splenic vein. There was an irregular area of low density within the posterior–inferior aspect of the spleen, most consistent with splenic infarction given the finding of mass effect on splenic vein and no history of trauma (Fig. 1). A transthoracic echocardiogram (TTE) was performed and ruled out a cardiac source of emboli that could cause splenic infarction. Hypercoagulable workup was negative for thrombophilia. The differential diagnosis included a pancreatic pseudocyst vs. a cystic neoplasm of the pancreas. Serum carcinoembryonic antigen (CEA) and CA 19-9 were elevated at 957 ng/ml and 792 U/ml, respectively. EUS with FNA of cyst fluid was performed. EUS revealed an anechoic, hypoechoic and distally enhancing lesion in the pancreatic tail, measuring 42×47 mm in maximum cross-sectional diameter. There was a single compartment without septae and the outer wall of the lesion was thin. This was in close proximity to the splenic hilum. There was internal debris within the fluid filled cavity. Parenchymal abnormalities consisting of hyperechoic, hypoechoic foci, lobularity and shadowing foci were noted in the pancreatic body and tail. Pancreatic duct measured up to 2 mm in diameter. There was no evidence of communication between the main pancreatic duct and the cyst. There was no sign of significant pathology in the common bile duct. The cyst was completely drained and sample was sent for cytology, tumor markers and amylase. Fluid analysis showed a CEA level of 154,300 ng/ml and amylase of 13,588 U/l. Cytology revealed highly atypical glandular cells consistent with adenocarcinoma in a background of proteinaceous and cystic debris. In the context of cystic mass in the tail of pancreas, the findings were compatible with either a cystadenocarcinoma or adenocarcinoma with cystic necrosis. Patient had distal pancreatectomy, splenectomy and partial gastrectomy. Pathology report showed moderately differentiated pancreatic mucinous cystadenocarcinoma, the tumor extended into the muscular layer of stomach, and parenchyma of spleen with vascular wall permeation (Fig. 2). Perineural invasion was also identified. Three of the 13 lymph nodes were positive for metastasis. Patient was later discharged under stable condition and followed up as outpatient with Oncology.

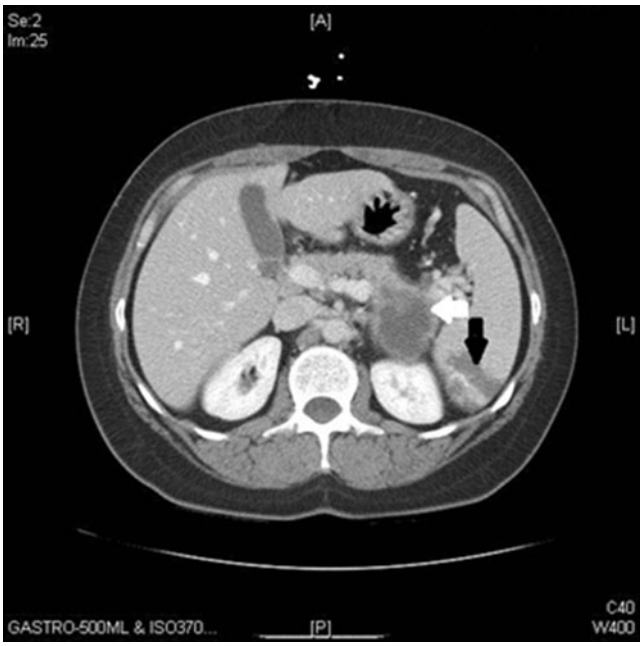

Fig. 1 Unilocular cystic mass in the tail of pancreas and splenic hilum (*white arrow*), and wedge-shaped area of infarction in the lower pole of spleen (*black arrow*)

Discussion

Pancreatic cystic lesions include inflammatory pseudocysts, benign, premalignant and malignant lesions. Pancreatic pseudocysts constitute 80–90% of cystic lesions of the pancreas [5]. Cystic neoplasms of the pancreas account for 10–15% of pancreatic cystic lesion and represent less than 10% of pancreatic neoplasms and about 1% of pancreatic malignancies [6]. The histopathological features of MCNs and intraductal papillary mucinous neoplasms (IPMNs) are almost identical, except for a dense mesenchymal ovarian-like stroma, which is a requisite feature of MCNs. Characteristically, MCNs lack a communication with the pancreatic ductal system, whereas communication is a key feature of IPMNs [7]. MCNs are usually thick-walled, septated cysts. The dense ovarian-like tissue simulates an ovarian hamartoma and, at times, a sarcoma. MCNs mostly (>90%) arise in the body and tail of pancreas and maybe unilocular or multilocular. MCNs are almost exclusively seen in females, with a mean age of presentation of 45 years. MCNs carry a risk for malignant transformation, and the incidence of CIS in resected MCNs is 25–50%.

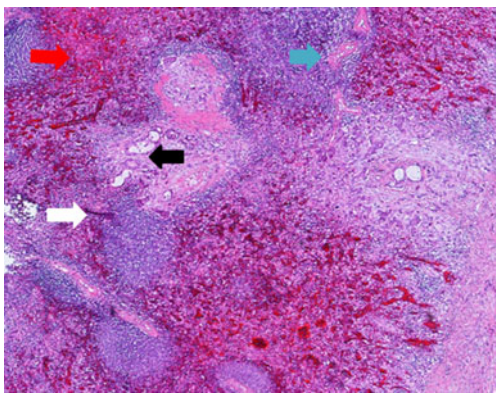

Fig. 2 Pancreatic tumor (*black arrow*) invading red pulp (*red arrow*) and white pulp (*white arrow*) of spleen parenchyma with vascular wall permeation (*blue arrow*)

Differentiation among mucinous cystadenomas, serous cystadenomas and pseudocysts is often difficult on the basis of clinical presentation. Cystic neoplasms of the pancreas may be confused with pseudocysts due to similarities in clinical presentation and visualization on imaging studies. Imaging tests, FNA, and/or cyst fluid analysis have been utilized to provide a diagnosis in patients with cystic lesions of pancreas [8]. Many imaging modalities can be used for differentiating between cystic neoplasms and pseudocysts. CT scan is a useful tool for initial detection of cystic lesions of pancreas as well as for characterization of cystic lesions by visualization of calcification of the cyst wall, septa, mural nodules, and findings suggestive of pancreatitis [9]. Pseudocysts appear on CT scan as low attenuation lesions, which are usually unilocular, with accompanying signs of acute or chronic pancreatitis [10]. MCNs are usually macrocystic. The presence of peripheral eggshell calcification, wall thickening and thick septations on CT scan can be suggestive of malignant MCNs [11]. EUS is an excellent imaging modality, as it allows for high frequency imaging of the pancreas and provides additional morphologic details of cystic lesions. EUS can provide information about presence or absence of a cyst wall, septations, mural nodules, debris, and an associated mass. In a study by Ahmad et al. [12], the accuracy for differentiating neoplastic lesions from nonneoplastic lesions ranged from 40% to 93%. EUS-guided FNA provides cyst fluid for cytologic examination and analysis of various biochemical and tumor markers, which may assist in diagnosis [13]. Mucin and mucinous cells are characteristic of MCNs, glycogen-staining cells are seen in serous cystadenomas, and inflammatory cells and histiocytes are seen in pseudocysts. Amylase-rich fluid is found in pseudocysts and in cysts associated with IPMNs, whereas low concentrations of amylase are found in fluid of serous cystadenomas as well as vast majority of MCNs. Most commonly, CEA and CA 72-4 have been used to differentiate between mucinous and serous cystadenomas [14].The concentration of CEA and CA 72-4 in aspirated cyst fluid tends to be higher in mucinous cystadenomas than serous cystadenomas and pseudocysts.

The differentiation between pancreatic pseudocyst and cystic neoplasms of pancreas is important because the treatment of these entities is quite different. Cystic neoplasms of pancreas, mucinous as well as serous, have malignant potential, and surgical resection is recommended [15], whereas pancreatic pseudocyst can often be managed conservatively, depending on the clinical setting.

It is extremely rare to find pancreatic mucinous cystadenocarcinoma in females younger than 40 years. Splenic infarction is frequently seen in patients with atrial fibrillation, significant hematologic diseases and thromboembolism. Because of the close proximity of the pancreas to the splenic vessels, splenic involvement including infarction, abscess, intrasplenic pseudocysts, and hemorrhage is frequently associated with pancreatitis [16]; however, splenic infarction caused by pancreatic mucinous cystadenocarcinoma has been rarely reported. Shiroma et al. [17] reported a case of splenic infarction caused by pancreatic cancer in Japan. Wong et al. [18] reported a case of advanced anaplastic carcinoma presenting with splenic infarction in Singapore. Görg et al. [19] reported a case series of ten patients with splenic infarction secondary to cancer, including two patients with pancreatic carcinoma and one patient with pancreatic neuroendocrine tumor. In the case of splenic infarction associated with pancreatic cancer, infarction of the splenic artery may be caused by torsion, direct invasion of the tumor, compression, or thromboembolism. It is also a known fact that complex factors associated with cancer contribute to the hypercoagulable and thrombophilic state in cancer patients.

Conclusion

Malignant MCNs are uncommon pancreatic tumors. Mucinous cystadenocarcinoma is extremely rare in young women (<40 years). Splenic infarction is a rare presentation of mucinous cystadenocarcinoma of the pancreas. A precise diagnosis is critical to help guide further management. Pancreatic pseudocysts can be managed operatively as well as non-operatively, but surgical resection is recommended for cystic neoplasms of the pancreas, benign as well as malignant. Different imaging modalities, including CT scan, MRI and EUS, can be used for differentiating between pancreatic pseudocysts and cystic neoplasms. EUS-guided FNA is highly accurate in differentiating pancreatic pseudocysts from cystic neoplasms and diagnosing malignancy in patients with focal pancreatic lesions detected by CT scan. EUS, as compared to CT scan and MRI, provides further details of the morphologic features of the cystic lesions as well as an opportunity to obtain the cystic fluid, which are useful in differentiating pancreatic mucinous lesions, non-mucinous lesions and pseudocysts.

References

1. Horvath KD, Chabot JA. An aggressive resectional approach to cystic neoplasms of the pancreas. Am J Surg. 1999;178(4):269–74.
2. Verbesey JE, Munson JL. Pancreatic cystic neoplasms. Surg Clin North Am. 2010;90(2):411–25.
3. Krishna NB, LaBundy JL, Saripalli S, et al. Diagnostic value of EUS-FNA in patients suspected of having pancreatic cancer with a focal lesion on CT scan/MRI but without obstructive jaundice. Pancreas. 2009;38(6):625–30.
4. Brugge WR. The role of EUS in the diagnosis of cystic lesions of the pancreas. Gastrointest Endosc. 2000;52:S18–22.

5. Sedlack R, Affi A, Vazquez-Sequeiros E, et al. Utility of EUS in the evaluation of cystic pancreatic lesions. Gastrointest Endosc. 2002;56:543–7.
6. Moparty B, Brugge WR. Approach to pancreatic cystic lesions. Curr Gastroenterol Rep. 2007;9(2):130–5.
7. Brugge WR, Lauwers GY, Sahani D, et al. Cystic neoplasms of the pancreas and their management. N Engl J Med. 2004;351:1218–26.
8. Brugge WR, Lewandrowski K, Lee-Lewandrowski E, et al. Diagnosis of pancreatic cystic neoplasms: a report of the cooperative pancreatic cyst study. Gastroenterology. 2004;126:1330–6.
9. Curry CA, Eng J, Horton KM, et al. CT of primary cystic pancreatic neoplasms: can CT be used for patient triage and treatment? Am J Roentgenol. 2000;175:99–103.
10. Morgan DE, Baron TH, Smith JK. Pancreatic fluid collections prior to intervention: evaluation with MR imaging compared with CT and US. Radiology. 1997;203:773–8.
11. Procacci C, Carbognin G, Accordini S, et al. CT features of malignant mucinous cystic tumors of the pancreas. Eur Radiol. 2001;11:1626–30.
12. Ahmad NA, Kochman ML, Brensinger C, et al. Interobserver agreement among endosonographers for the diagnosis of neoplastic versus non-neoplastic pancreatic cystic lesions. Gastrointest Endosc. 2003;58:59–64.
13. Vignesh S, Brugge WR. Endoscopic diagnosis and treatment of pancreatic cysts. J Clin Gastroenterol. 2008;42(5):493–506.
14. Hammel P, Voitot H, Vilgrain V, et al. Diagnostic value of CA 72-4 and carcinoembryonic antigen determination in the fluid of pancreatic cystic lesions. Eur J Gastroenterol Hepatol. 1998;10:345–8.
15. Khalid A, Brugge W. ACG practice guidelines for the diagnosis and management of neoplastic pancreatic cysts. Am J Gastroenterol. 2007;102(10):2339–49.
16. Fishman EK, Soyer P, Bliss DF, et al. Splenic involvement in pancreatitis: spectrum of CT findings. Am J Roentgenol. 1995;164:631–5.
17. Shiroma J, et al. Splenic infarction in pancreatic cancer. J Pancreas. 2009;10:699–700.
18. Wong M, See JY, Sufyan W, Diddapur RK. Splenic infarction. A rare presentation of anaplastic pancreatic carcinoma and a review of the literature. J Pancreas. 2008;9:493–8.
19. Görg C, Seifart U, Görg K. Acute, complete splenic infarction in cancer patient is associated with a fatal outcome. Abdom Imaging. 2004;29:224–7.