



Pancreatic panniculitis preceding acute pancreatitis and subsequent detection of an intraductal papillary mucinous neoplasm: A case report

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INTRODUCTION

Pancreatic panniculitis is a rare skin manifestation of pancreatic disease. It is believed to result from seepage of pancreatic enzymes into the systemic circulation, which leads to skin and occasionally joint manifestations. It is reported to occur in up to 3% of patients with pancreatic disease.¹ Pancreatic panniculitis can develop before, concomitantly with, or after the diagnosis of clinically detectable pancreatic conditions. The condition is most frequently associated with acute pancreatitis, chronic pancreatitis, and pancreatic carcinoma. The most common carcinoma is acinar cell type.

Here we report an unusual association of pancreatic panniculitis preceding a single episode of acute pancreatitis and subsequent development of an intraductal papillary mucinous neoplasm (IPMN).

CASE REPORT

A 61-year-old female nondrinker and nonsmoker was first seen by the dermatology service for a putative diagnosis of recurrent bilateral lower limb cellulitis. She was previously admitted on 3 occasions over 2 months with presumed lower limb cellulitis treated with oral and intravenous antibiotics. She had a medical history of schizophrenia for which she was taking risperidone. Examination found multiple tender erythematous nodules on both lower limbs extending to the thighs. The nodules were nonsuppurative throughout (Fig 1). Skin biopsies results showed striking fat necrosis in the dermis and subcutis with focal neutrophilic

Abbreviations used:

CT:	computed tomography
EUS:	endoscopic ultrasound scan
IPMN:	intraductal papillary mucinous neoplasm
MRCP:	magnetic resonance cholangiopancreatography
MPD:	main pancreatic duct

infiltrate. Ghost outlines of fat cells were seen with focal basophilia, raising the possibility of pancreatic fat necrosis (Fig 2). Her amylase was 148 IU/L (reference range, 0–100 IU/L).

The patient attended the gastroenterology outpatient clinic 4 weeks later, and on the day of consultation presented with her first attack of acute pancreatitis (sudden-onset epigastric pain with serum amylase 5,500 IU/L). Liver function tests, serum calcium, and triglycerides were normal. A computed tomography (CT) scan of the pancreas and magnetic resonance cholangiopancreatography (MRCP) confirmed grade 2 nonnecrotizing pancreatitis. This condition was treated successfully with intravenous fluids and analgesia. There was a small thrombus in the proximal splenic vein, close to the portal-splenic confluence. No direct communication with the pancreatic ductal system was visualized. The main pancreatic duct (MPD) was normal, and there were no dilated branch ducts. Endoscopic ultrasound scan (EUS) at that time found the main pancreatic duct to be in contact with the portal/splenic confluence, but no direct communication was seen. In addition, there was a small cystic 6-mm

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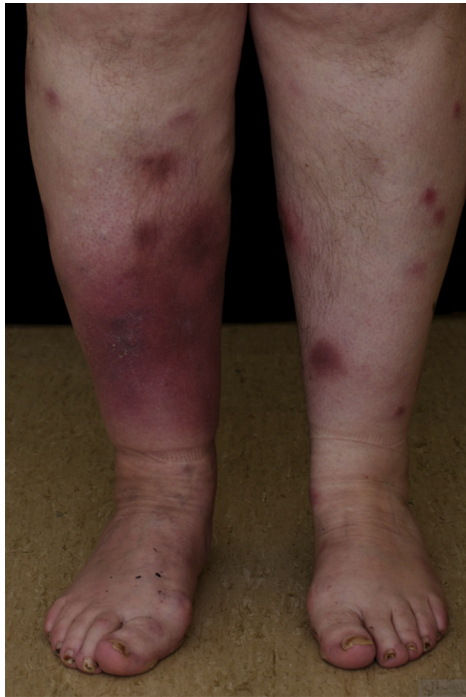


Fig 1. Pancreatic panniculitis. Erythematous indurated nodules, bilateral lower limbs.

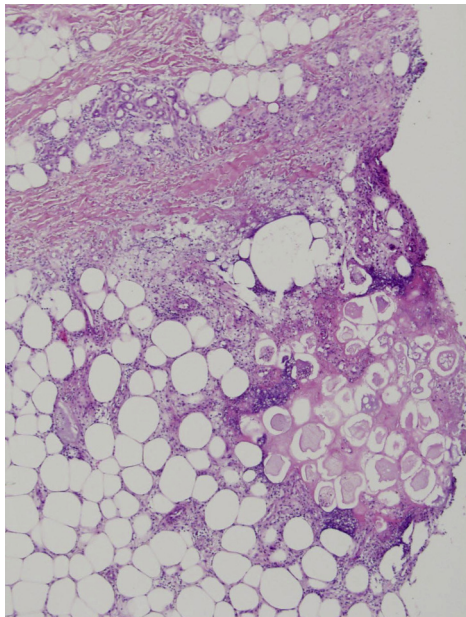


Fig 2. Pancreatic panniculitis. Skin biopsy results (high power) show necrotic ghostlike fat cells.

lesion in the upper isthmus/body junction but at a distance from the MPD and vessels. The pancreatic parenchyma was normal without stigmata of chronic pancreatitis. The biliary system was normal and without biliary lithiasis. The thrombus was treated successfully with anticoagulants. Her pain rapidly

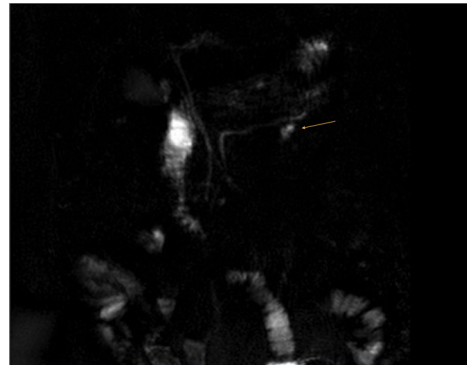


Fig 3. MRCP (thick slice) dilated branch duct at the body/tail of the pancreas.

settled, and pancreatic enzymes returned to normal within a few days. Follow-up EUS 1 year later found 2 small elongated cysts in contact with the MPD at the level of isthmus (11 × 4 mm) and midbody (6 mm). This finding was consistent with small dilated accessory ducts (ie, branch duct IPMNs), which revealed minimal enlargement on yearly MRCP (Fig 3). The panniculitis resolved spontaneously within 2 to 3 months.

She was subsequently lost to follow-up but had been well up to July 2015 when she presented with a 1-week history of painless obstructive jaundice. She had no associated weight loss. Serum amylase was normal. A CT scan of the abdomen and EUS identified a 4.7-cm ill-defined solid and cystic mass in the pancreatic head emanating from the dilated MPD (maximum diameter, 13 mm). This mass was felt to be consistent with mixed main duct and branch duct IPMN with malignant transformation close to upper head and isthmus. Involvement of the mesenteric fat and the superior mesenteric vein was also noted. EUS-guided fine-needle aspiration biopsy confirmed a pancreatic adenocarcinoma. She was referred for a surgical opinion, but her tumor was deemed unsuitable for resection. She is currently undergoing systemic chemotherapy.

DISCUSSION

Pancreatic panniculitis generally affects the lower limbs and buttocks but has been reported to occur on the trunk and upper extremities.² The differential diagnosis includes erythema nodosum, erythema induratum, alpha-1 antitrypsin deficiency–associated panniculitis, trauma-induced panniculitis, and cutaneous metastases. Histology from a skin biopsy of pancreatic panniculitis typically shows lobular panniculitis with fat necrosis, basophilic debris, and anucleated adipocytes with a partially digested shadowy cell membrane, also known as *ghostlike cells*.

Lipase and amylase have been detected in areas of subcutaneous fat necrosis associated with pancreatic disease, suggesting that these enzymes have a role in the development of pancreatic panniculitis.^{3,4} Notably, pancreatic panniculitis has been reported to occur in patients with low or normal serum pancreatic enzymes, suggesting a multifactorial process.⁵ Zellman⁶ suggested that additional factors are required for the development of pancreatic panniculitis, such as increased vessel permeability, allowing the pancreatic enzymes to leak out of the circulation and into subcutaneous fat. Other case reports in the literature support this theory of vascular permeability, for example, pancreatic panniculitis occurring after femoral vein catheterization⁷ and paracentesis.⁸

There are 2 reports in the literature of patients with IPMN having pancreatic panniculitis. One patient presented with weight loss and was subsequently found to have a pancreatic mass on abdominal CT scan. The cutaneous lesions occurred 2 days after pancreaticoduodenectomy.⁹ The second patient presented with a 10-week history of multiple tender subcutaneous nodules; IPMN was detected following diagnosis of pancreatic panniculitis.¹⁰

We highlight this case of pancreatic panniculitis given the unusual sequence of events: the initial dermatologic condition possibly resulted from a small dilated branch duct communicating with the portal venous system. As a consequence, there was seepage of pancreatic enzymes into the systemic circulation, leading to the development of pancreatic

panniculitis. IPMN was the cause of pancreatic panniculitis in this case, and malignant transformation was discovered 3 years later. It is intriguing that the patient's skin lesions resolved despite progression of the pancreatic disease.

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