

Pancreatic panniculitis as the initial presentation of intrahepatic cholangiocarcinoma



Mary Katherine Montes de Oca, BS,^{a,b} and Richard Andrew Jamison, MD^{a,c}
Greenville, South Carolina and Birmingham, Alabama

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INTRODUCTION

Pancreatic panniculitis is caused by the release of pancreatic enzymes into the bloodstream resulting in subcutaneous fat necrosis. Clinically, this condition manifests as painful or painless subcutaneous nodules on the legs. The lesions are similar to erythema nodosum and most commonly are associated with an underlying pancreatic condition. Histopathologically, pancreatic panniculitis lesions appear as subcutaneous focal steatonecrosis, and there are pathognomonic “ghost cells,” which are anucleate necrotic adipocytes with thick, obscure walls. One previous case reported pancreatic panniculitis as the presenting symptom of hepatic carcinoma.

CASE REPORT

A well-appearing 53-year-old woman presented with indurated, erythematous, draining nodules on her right and left calves (Figs 1 and 2). The rash was moderately painful and pruritic and had been present for 2 months. Before the initial eruption on her legs, about 2 months before presentation to the dermatologist, she had an isolated episode of vomiting and intermittent right upper quadrant abdominal pain. She also complained of chronic gastroesophageal reflux, which had become more problematic and was being treated with a proton pump inhibitor for 3 months at the time of presentation. She had a history of a spontaneous pneumothorax 1 year before presentation. The patient denied any weight loss or fevers. The initial workup included a punch biopsy and laboratory tests including liver function tests and amylase and lipase

values. Her laboratory values showed a lipase of 8,880 U/L, amylase within normal limits, and aspartate aminotransferase and alanine aminotransferase within normal limits. Punch biopsy results showed a lobular panniculitis with mixed infiltrate and basophilic deposits and enzymatic fat necrosis with the characteristic ghost cells (Figs 3 and 4). Given the severity of her presentation, she was started on a 40-mg prednisone taper. She was referred to gastrointestinal specialists who found 3 liver masses on computed tomography that were suspicious for malignancy. Colonoscopy and esophagogastroduodenoscopy findings were normal. Two lesions measured up to 10 cm in size, and another lesion was 2 cm in size. There was no evidence of cirrhosis on examination. There was some noted atrophy of the tail of the pancreas, but there was no obvious mass in the pancreas. She underwent ultrasound-guided liver biopsy, which found a moderately differentiated malignant tumor that was CK7⁺ and CA19-9⁺. All other markers, including those for hepatocellular carcinoma, were negative. Based on the pattern of staining, it was thought that this was most likely a metastatic adenocarcinoma from an upper gastrointestinal site or possible intrahepatic bile duct carcinoma. The patient was referred to an oncologist where the decision was made to start chemotherapy including cisplatin and gemcitabine to assess for a response and allow for possible declaration of the site of primary tumor. After 5 rounds of chemotherapy, the size of the liver lesions was unchanged. Next, a right hepatectomy was performed which found

From the University of South Carolina School of Medicine Greenville^a; Samford University, Birmingham^b; and the Division of Dermatology, Greenville Health System.^c

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Correspondence to: Mary Katherine Montes de Oca, BS, University of South Carolina School of Medicine Greenville, 607 Grove Rd, Greenville, SC, 29605. E-mail: maryk.montesdeoca@gmail.com.

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Fig 1. Pancreatic panniculitis presenting as right lower extremity indurated, erythematous, draining nodules.



Fig 2. Pancreatic panniculitis presenting as left lower extremity indurated, erythematous, draining nodules.

intrahepatic cholangiocarcinoma. The patient has recovered well and was able to return to work.

DISCUSSION

Although pancreatic panniculitis may be the presenting symptom for 2% to 3% of pancreatic diseases including chronic pancreatitis, posttraumatic pancreatitis, pancreatic pseudocysts, and pancreatic carcinoma, it may also signal nonpancreatic malignancies such as hepatic carcinoma and cholangiocarcinoma.^{1,2} When the pancreas is damaged, amylase and lipase are released into the bloodstream and deposited in adipose tissue where fat necrosis occurs.³ This manifests clinically as painful, erythematous, edematous nodules, which may drain an oily brown-yellow fluid or ulcerate.^{4,5} These nodules are most commonly found on the legs, arms, buttocks, and trunk.^{4,6} Patients with pancreatic panniculitis may have increased serum levels of pancreatic lipase.⁶⁻⁸ In addition to the cutaneous manifestations, patients may present with abdominal symptoms and signs of arthritis in the ankles, hands, and knees.^{6,8} One previous case

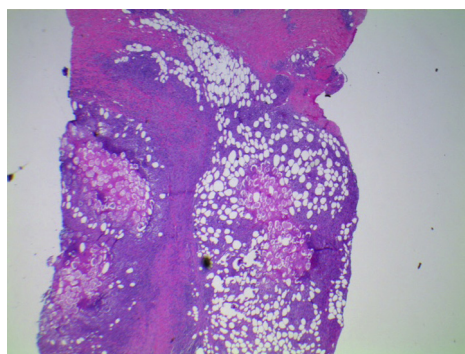


Fig 3. Histopathologic examination of punch biopsy on high power shows lobular panniculitis with mixed infiltrate and basophilic deposits.

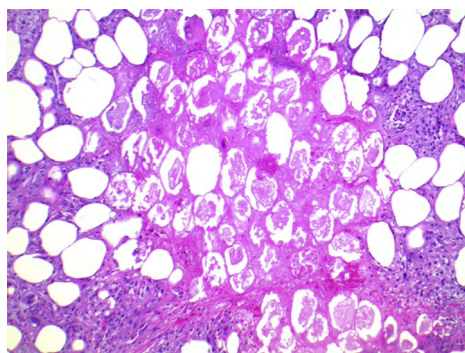


Fig 4. Histopathologic examination of punch biopsy on high power shows enzymatic fat necrosis with ghost cells, anucleate necrotic adipocytes with thick, obscure walls.

reported pancreatic panniculitis as the initial symptom of hepatic carcinoma, and another case presents the first patient with pancreatic panniculitis caused by pancreatic-type acinar cell carcinoma of the liver with no underlying pancreatic disease.^{8,9} Here, we discuss a patient with intrahepatic cholangiocarcinoma with no obvious pancreatic condition.

In this case, the lesion was thought to be a panniculitis at the time, and the differential diagnosis included α -1 antitrypsin deficiency panniculitis, erythema nodosum, pancreatic panniculitis, and lupus panniculitis. Alpha-1 antitrypsin was entertained because of her history of a spontaneous pneumothorax, but the patient had no history of chronic obstructive pulmonary disease. The patient initially had abdominal pain and vomiting before the eruption on her legs, which could represent gallstones or pancreatitis, which led to a draining panniculitis.

The diagnostic workup includes the clinical presentation of skin and joint manifestations, serum levels of pancreatic enzymes, cutaneous punch biopsies, and abdominal imaging.^{1,8} The histopathologic findings in the subcutaneous tissue consist of

focal areas of fat necrosis and anucleate, necrotic adipocytes known as *ghost cells*.⁶ Several treatment strategies have been used and include addressing the underlying condition, systemic steroids, octreotide, nonsteroidal anti-inflammatory drugs, and immunosuppressive drugs.⁴

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