Ulcerated, tender nodules of the lower extremities

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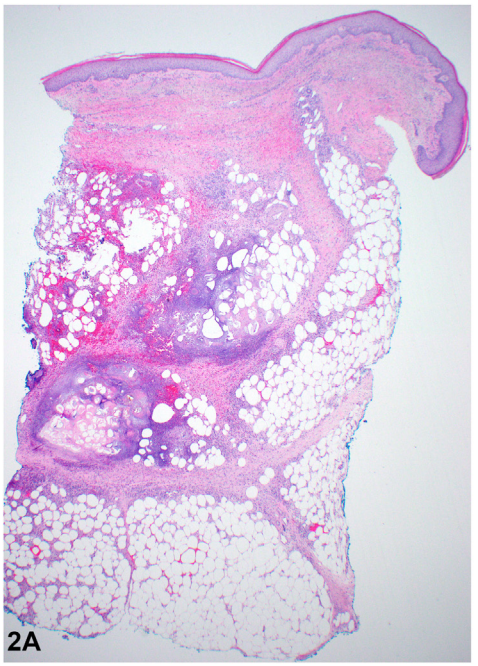

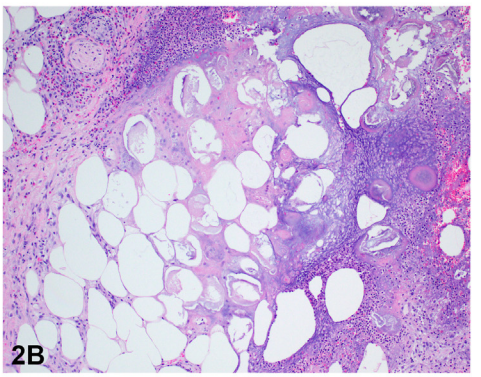

CASE

A 63-year-old woman presented with 1 week of worsening lower extremity lesions that started as multiple red pustules and evolved into subcutaneous nodules with overlying ulceration. On exam, there were many 0.5- to 2-cm dark pink and purple papulonodules with central hemorrhagic crust and erosions (Fig 1) and 2 tender,

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The patient gave consent for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available.

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several-centimeter ulcerations with violaceous, rolled borders and necrotic bases. The lesions were debrided on admission due to concern for infection. She had 2 weeks of diarrhea secondary to her chronic pancreatitis but denied acute pancreatitis symptoms including epigastric pain, fever, or nausea. Punch biopsy demonstrated lobular panniculitis, "ghost cells," calcification, and neutrophil-rich infiltrate (Fig 2).

Question 1: What is the most likely diagnosis?

A. Pyoderma gangrenosum

B. Pancreatic panniculitis

C. Alpha-1 antitrypsin deficiency panniculitis

D. Nodular vasculitis

E. Factitial vasculitis

Answer:

A. Pyoderma gangrenosum — Incorrect. Histopathology would demonstrate dense neutrophilic inflammation and the presence of ghost cells, and saponification would not be seen.[1]

B. Pancreatic panniculitis — Correct. Pancreatic panniculitis presents clinically with lesions to the lower extremities that are tender, erythematous, red-brown in color, nodular, ulcerated, and potentially with an oily drainage.[2] In 40% of cases, cutaneous findings precede the diagnosis of pancreatic disease by 1 to 7 months.[3,4] Along with these clinical features in the setting of pancreatic disease, an incisional biopsy can help confirm the diagnosis. A deep incisional biopsy is necessary to ensure adequate tissue sampling that includes subcutaneous fat. The main feature on histology is mostly lobular panniculitis without vasculitis. "Ghost cells" are also pathognomonic for panniculitis and result from coagulative necrosis of adipocytes due to pancreatic enzymes. Within "ghost cells," saponification can be seen as fine basophilic granular material due to dystrophic calcification.[2]

C. Alpha-1 antitrypsin deficiency panniculitis — Incorrect. Histopathology is absent of skip areas, making this diagnosis less likely.

D. Nodular vasculitis — Incorrect. Although this pathology also demonstrates erythematous, tender, ulcerated nodules of the lower extremities, its histopathology would reveal a mixed inflammatory infiltrate and would not demonstrate "ghost cells."[5]

E. Factitial vasculitis — Incorrect. Although some of our patient's clinical symptoms can be self-induced, the histopathology is more specific for pancreatic panniculitis.

Question 2: Pancreatic panniculitis is a cutaneous presentation that occurs in 2% to 3% of patients with pancreatic disease. Which of these etiologies is not associated with pancreatic panniculitis?

A. Malignancy

B. Infection

C. Trauma

D. Metabolic

E. Chemical

Answer:

A. Malignancy — Incorrect. Pancreatic panniculitis can be associated with pancreatic acinar carcinoma, can be associated with islet cell carcinoma, and even have nonpancreatic associations such as intrahepatic cholangiocarcinoma.[2,6,7] The use of imaging, tumor markers, and biopsies should be utilized to rule out malignancy as the etiology of pancreatic panniculitis.

B. Infection — Incorrect. Certain infections may result in acute pancreatitis which can progress to pancreatic panniculitis. For example, viral infections of mumps, coxsackievirus, hepatitis B, cytomegalovirus, varicella-zoster, herpes simplex, and HIV; bacterial infections of *Mycoplasma, Legionella, Leptospira*, and *Salmonella*; fungal infections of *Aspergillus*; or parasitic infections of *Toxoplasma, Cryptosporidium,* and *Ascaris* all can cause pancreatitis.[2,6-8] The clinical presentation of ulcerated nodules could mimic infection in the skin, and the presence of neutrophils on histopathology also can make this a concern, but the presence of ghost cells and calcification supports pancreatic panniculitis.

C. Trauma — Incorrect. Pancreatic panniculitis can arise from traumatic pancreatitis or status after endoscopic retrograde cholangiopancreatography (ERCP).[2,6,7]

D. Metabolic — Correct. Metabolic processes such as exocrine pancreatic insufficiency have not been associated with pancreatic panniculitis.[2,6,7]

E. Chemical — Incorrect. Pancreatic panniculitis can arise in pancreatitis secondary to sulindac intake.[2,6,7]

Question 3: What is the most effective treatment recommendation for pancreatic panniculitis?

A. Wide local excision

B. Lower pancreatic enzymes

C. Supportive

D. Immune support

E. Topical steroids

Answer:

A. Wide local excision — Incorrect. This has not been shown to treat pancreatic panniculitis since it does not address the root cause of the pathology. However, punch biopsies can be used to help in the diagnosis of pancreatic panniculitis as seen in our patient.

B. Lower pancreatic enzymes — Correct. A goal of pancreatic panniculitis treatment is to decrease pancreatic enzyme levels.[2,6] High pancreatic enzyme levels indicate continued fat saponification and progression of cutaneous lesions. Our patient had asymptomatic acute pancreatitis, so obtaining an ERCP was made to search for an underlying cause. ERCP identified a portion of the pancreatic duct that appeared extrinsically compressed to a tight stricture by a pancreatic cyst along with dilation of the main pancreatic duct within the body of the pancreas. There was a significant downtrend in lipase and amylase levels after ERCP. It is also important to note that elevated amylase, lipase, and aspartate aminotransferase/alanine aminotransferase are not diagnostic of pancreatic panniculitis as these abnormalities are not always observed in patients.

C. Supportive — Incorrect. Supportive measures, such as analgesics, can help the patient tolerate the tender, ulcerated lesions seen in pancreatic panniculitis. However, pancreatic panniculitis will not resolve without an intervention that will decrease pancreatic enzyme levels.

D. Immune support — Incorrect. Increased immunosuppressive medications in patients with pancreatic panniculitis status after transplant have been shown to rapidly normalize pancreatic enzyme levels.[9]

E. Topical steroids — Incorrect. May provide symptomatic relief, but they have shown to be ineffective in treatment of pancreatic panniculitis.[6]

Abbreviation used:

ERCP: endoscopic retrograde cholangiopancreatography

Conflicts of interest

None disclosed.

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