Pancreatitis, Panniculitis, and Polyarthralgia (PPP) Syndrome: A Rare Complication of Pancreatic Pathology

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❖ Introduction:

- Pancreatitis, panniculitis, and polyarthritis (PPP) is a rare triad presenting with a unique diagnostic and therapeutic challenge.
- Patients with previous pancreatic pathologies can develop fistulas leading to the drainage of lipase from the pancreatic outflow tract into the vasculature, causing various clinical manifestations.
- Here, we present a patient with a previous history of pancreatitis and now diagnosed with PPP syndrome.

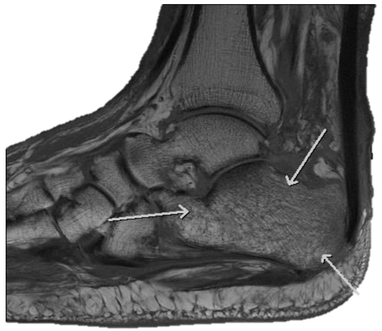

Figure 2: Reactive bone changes, caused by bone autodigestion by pancreatic enzymes

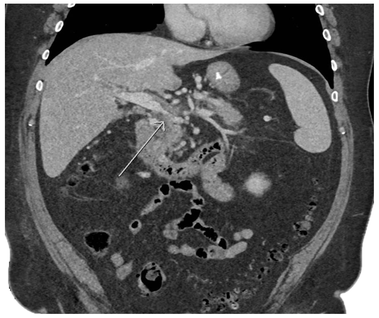

Figure 3: Thrombosis of superior mesenteric vein

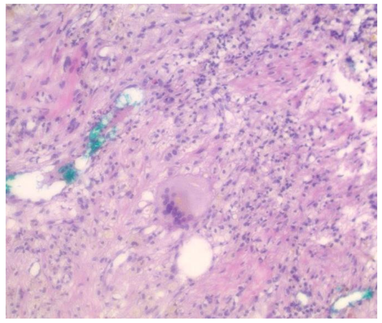

Figure 4: "Ghost cells" consisting of anucleate necrotic adipocytes with a basophilic material

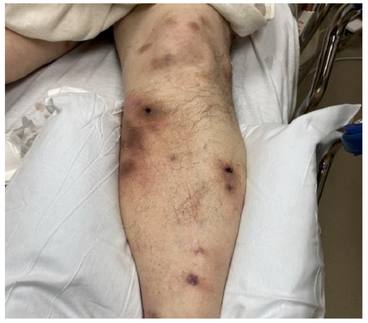

Figure 1: Pancreatic panniculitis of lower extremities

❖ Case Description:

- A 64-year-old gentleman presented with polyarthralgia in his lower and upper extremities for several months. Six months ago, he started having nodular lesions on his bilateral lower extremities, initially misdiagnosed as Erythema Nodosum, which later started draining. At that time, he had fevers and was hospitalized.
- Notably, the patient had a history of heavy alcohol use but did not have any typical symptoms associated with pancreatitis.
- On examination, the lesions were ulcerative with necrotic floor and serosanguinous discharge (Fig. 1), with the most prominent lesion in the right calcaneus.
- Lipase: 2328 U/L, ESR: 75 mm/hour, CRP: 5.7 mg/L, mild transaminitis.
- MRI of bilateral lower extremities revealed reactive bone changes in the left calcaneus (Fig. 2), diagnosed as osteomyelitis.
- CT Abdomen with contrast (Fig. 3) showed acute on chronic pancreatitis, complete thrombotic occlusion of the superior mesenteric vein, and partial occlusion of the portal vein.
- Biopsy of the lesions (Fig. 4) showed inflammation and ghost cells suggestive of pancreatic panniculitis.
- He received IV Amoxicillin and IV Ertapenem for six weeks. Due to non-response, his IV Ertapenem was continued for another six weeks. He was followed by Podiatry and received regular wound care.

❖ Discussion:

- The hypothesized pathogenesis for PPP syndrome is the formation of tiny fistulas between the pancreatic outflow and superior mesenteric vein leading to the intravascular leakage of lipase. These tiny fistulae may be difficult to visualize on imaging, but thrombosis of the mesenteric vein on imaging can indicate their presence.
- Panniculitis is believed to be caused by auto-digestion of adipocytes in subcutaneous tissue by lipase. On biopsy of skin lesions, the presence of ghost cells is pathognomonic for pancreatic panniculitis. The skin lesions can be misidentified as Erythema Nodosum; unlike EN, these lesions ulcerate and drain a serosanguinous discharge.
- Polyarthralgias are due to the deposition of free fatty acids in joints released subsequent to fat autodigestion. Although the lipase levels in the blood can be significantly elevated, patients usually present with no or mild abdominal symptoms.
- The reactive bone changes are caused due to bone necrosis and can be falsely diagnosed as osteomyelitis on imaging. This can delay diagnosis and treatment causing patient morbidity.
- Some previous cases have been successfully treated with surgical resection of the fistulae and steroids to control the inflammation
- Patients who present with history of alcohol use and necrotizing skin lesions need to be evaluated for possible PPP syndrome by abdominal CT and biopsy of skin lesions. The reactive bone changes can masquerade as osteomyelitis and the patient may be treated with antibiotics without any presence of infection.