

## Case 5.1

### A 71 year-old Thai male from Bangkok province

**Chief complaint** : Painful skin lesions on his legs for 2 days

**Present illness** : The patient's medical history included chronic pancreatitis with retained CBD stone, alcohol dependent, gouty arthritis. He was admitted to department of medicine with acute dyspnea, which later was diagnosed as acute pulmonary embolism. During the admission, he developed acute painful skin nodules on both legs

**Physical exam** : Multiple discrete painful erythematous indurated plaques and nodules on both legs with moderate swelling of right ankle

**Investigation** : serum amylase 1825 U/L, serum lipase > 20,000 U/L

**CT abdomen** : Mild atrophic change of pancreas with some calcification suggest chronic pancreatitis. A small CBD stone with tortuous dilated CBD

#### **Histopathology** (S09-7427) (Fig 5.1.2, 5.1.3)

- Dense mixed inflammatory cell infiltrate of mostly neutrophils and basophilic debris around necrotic fat lobules.
- Necrotic fat cells show anucleate, hyalinized cell rim (ghost cells), some with calcified saponification.
- Superficial and deep perivascular infiltrate of lymphocytes and neutrophils in the dermis.



Fig. 5.1.1

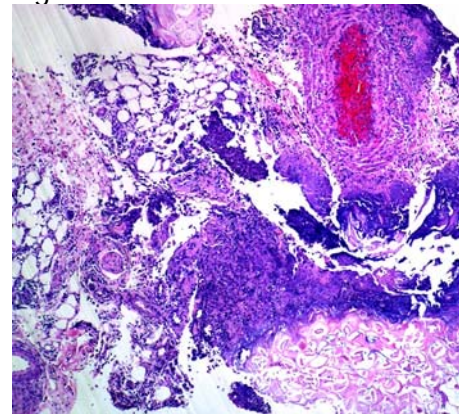


Fig. 5.1.2

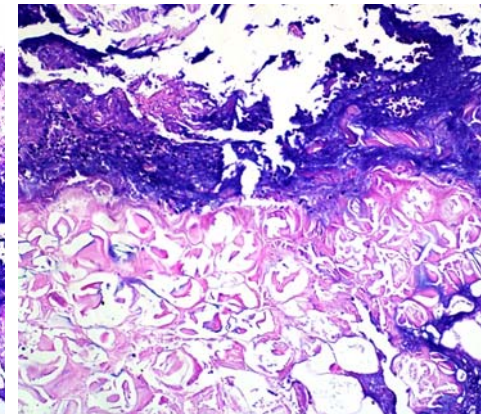


Fig. 5.1.3

**Diagnosis** : Pancreatic panniculitis associated with chronic pancreatitis with retained CBD stone

## Case 5.2

**A 45 year-old Thai male from Nontaburi province**

**Chief complaint** : Skin lesions at both thighs and legs

**Present illness** : He was admitted to department of medicine with acute abdominal pain under the impression of acute alcoholic pancreatitis with respiratory and renal failure. He was treated by respiratory support and hemodialysis. Then he developed few subcutaneous nodules on his both thighs and legs.

**Physical exam** : Few discrete ill-defined erythematous indurated plaques and nodules on both thighs and legs ( Lt > Rt )

**Investigation** : serum amylase 185 U/L, serum lipase 7808 U/L

**CT abdomen** : Suggestive of acute pancreatitis with necrosis, hemorrhage and large amount of free fluid collection



Fig. 5.2.1

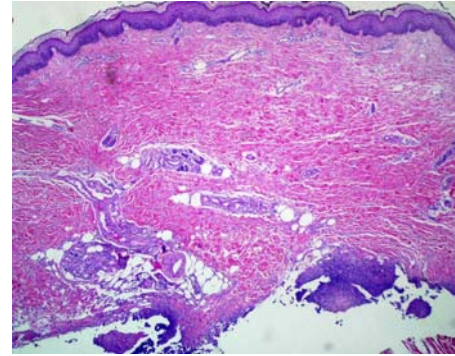


Fig. 5.2.2

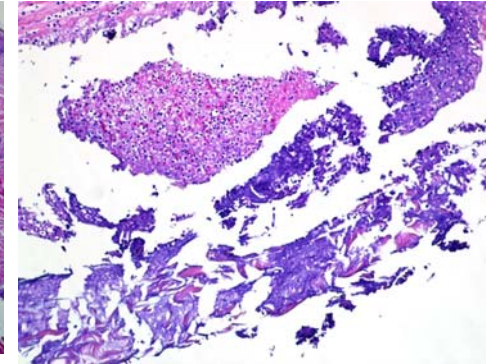


Fig. 5.2.3

**Histopathology** (S90-8134A) (Fig 5.2.2, 5.2.3)

- Dense inflammatory infiltrate of neutrophils and cell debris around necrotic fat cells and calcified saponification.

**Diagnosis** : Pancreatic panniculitis associated with acute alcoholic pancreatitis

### Case 5.3

#### A 47 year-old Thai male from Bangkok province

**Present illness :** A healthy man with a history of alcohol abuse presented with severe abdominal distension. The diagnosis was acute alcoholic hemorrhagic pancreatitis with pancreatic necrosis. He was consulted for maculopapular rash at trunk which drug eruption was suspected. On physical examination, there are diffuse erythematous maculopapular rashes at trunk and groin region. After reviewing the history of drug administration, the impression was meropenem induced exanthematous drug eruptions. In addition, non pitting edema was observed on his lateral side of both thighs. The surface revealed few ill-defined round firm erythematous nodules.

**Investigation :** serum amylase 497 U/L, serum lipase 15692 U/L

**CT abdomen :** Suggestive of acute pancreatitis with suspected focal necrotic area at pancreatic body-tail and fluid collections at right upper abdomen

**Histopathology** (S09-7084A) (Fig. 5.3.2, 5.3.3)

- Dense inflammatory cells infiltrate of neutrophils and basophilic debris necrotic fat cell and calcified saponification.
- Infiltrate of neutrophils with thrombi in the adjacent large vessels.



Fig. 5.3.1

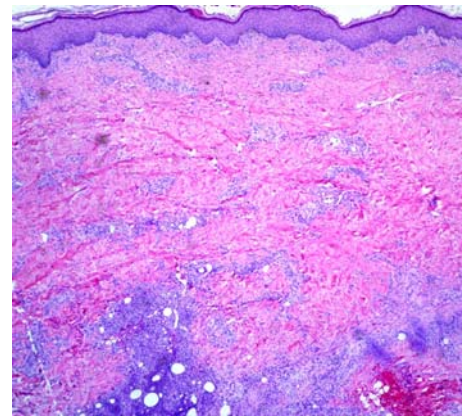


Fig. 5.3.2

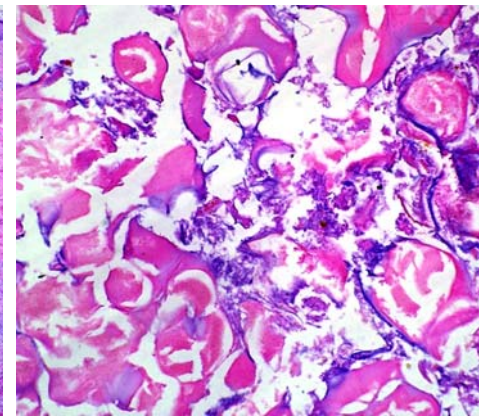


Fig. 5.3.3

**Diagnosis :** Pancreatic panniculitis associated with acute alcoholic pancreatitis with pancreatic necrosis

**Presenter :** Chatchadaporn Keerathikajornchai

**Consultant :** Ploysyne Busaracome

## Discussion

Panniculitis developing in patients with pancreatic diseases was first described by Chiari in 1883 but was not reported in the English literature until 1947 by Szymanski and Bluefarb. Although several review papers of pancreatic panniculitis have been published, most of them only include reports of one or two cases. We present here three cases of pancreatic panniculitis associated with different conditions of pancreatitis. This variant of panniculitis appears in approximately 2% to 3% of all patients with pancreatic disorders<sup>1</sup>, with a higher incidence among alcoholic men. The exact pathogenic mechanism of pancreatic panniculitis is still unclear, but release of pancreatic enzymes, such as lipase, phosphorilase, trypsin, and amylase, may be involved. Trypsin may increase the permeability of the microcirculation within lymphatic vessels, allowing the enzymes, such as lipase or amylase, to enter into the fat lobules and hydrolyze neutral fat to form glycerol and free fatty acids, which results in fat necrosis and inflammation<sup>2</sup>.

The most common pancreatic disorders associated with pancreatic panniculitis are acute or chronic pancreatitis<sup>3</sup> (especially alcohol related) and pancreatic carcinoma (usually acinar cell carcinoma, less frequently islet cell carcinoma<sup>4</sup>). Rarely are other pancreatic disorders associated with pancreatic panniculitis, including posttraumatic pancreatitis<sup>5</sup>, pancreatic pseudocysts, pancreas divisum<sup>6</sup> and vascular pancreatic fistulas<sup>1</sup>. Pancreatic panniculitis has been reported in association with primary human immunodeficiency virus (HIV) infection and a hemophagocytic syndrome<sup>7</sup>. Furthermore, allograft pancreatitis and rejection in pancreas-kidney transplant recipient presented with pancreatic panniculitis<sup>8</sup>. Recently it has been described in association with a liver carcinoma<sup>9</sup>, and a sign of adenocarcinoma of unknown origin<sup>10</sup>.

Clinically, pancreatic panniculitis presents with ill defined, tender, edematous, erythematous, or red-brown nodules that may spontaneously ulcerate and drain an oily brown, sterile, and viscous substance that results from liquefaction necrosis of adipocytes. These lesions are usually located on the distal parts of the lower extremities,

around the ankles and knees, although nodules can spread over the thighs, buttocks, arms, abdomen, chest, and scalp. In moderate cases associated with pancreatitis, the nodules usually do not break down and tend to resolve when the inflammatory pancreatic episode regresses, leaving an atrophic hyperpigmented scar<sup>11</sup>. In contrast, in patients with pancreatic panniculitis associated with pancreatic carcinoma, the nodules tend to be more persistent with frequent recurrences, ulceration, and involvement of cutaneous areas beyond the lower extremities.

Extrapancreatic manifestation may be present in patient with pancreatic disease. Monoarticular or oligoarticular arthritic symptoms have been reported in 56% of the patients<sup>11</sup>. It is usually symmetric, although it may be intermittent, migratory, or persistent, caused by focal necrosis of the periarticular fat. Other foci of metastatic fat necrosis include abdominal fat, pleural effusions, pulmonary infiltrates, mesenteric thrombosis and bone marrow fat necrosis. Schmid's triad is the presence of panniculitis, polyarthritis and eosinophilia in a patient with pancreatic tumor. It is associated with poor prognosis<sup>12</sup>.

The diagnosis of this condition frequently requires deep skin biopsy. The most pathognomonic feature is a predominantly neutrophilic lobular panniculitis without vasculitis with characteristic coagulative necrosis of the adipocytes and saponification of fat from pancreatic enzyme, which leads to ghost adipocytes. However, septal panniculitis has been reported. It is postulated that reaction is the earliest histopathologic change in pancreatic panniculitis because the first enzyme attack is directed against the endothelial lining of septal veins and venules<sup>13</sup>. Serum levels of amylase, lipase, or trypsin are usually elevated in pancreatic panniculitis, although not in all cases and degree of panniculitis does not correlate with enzyme levels.

Treatment of pancreatic panniculitis is primarily supportive and should be directed to the underlying pancreatic disease. Sometimes, complete resolution of the symptoms occurs when the pancreatic anomaly is surgically corrected. Administration of the somatostatin

analogue octreotide, a synthetic polypeptide that inhibits pancreatic enzyme production, resulted in a significant resolution of symptoms in a patient with pancreatic carcinoma<sup>14</sup>, but this effect could not be reproduced in other cases.

## References

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