Case 26
A 80-year-old Thai male from Nonthaburi
Chief complaint: A 2-week history of painful lumps on both legs and feet

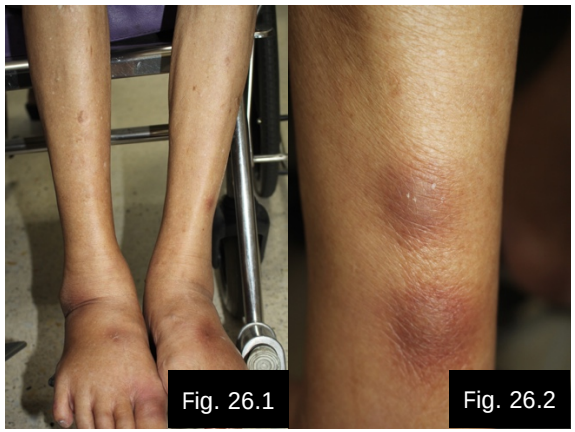
Fig. 26.1

Fig. 26.2

Present illness:
The patient presented with a 2-week history of painful lumps on both legs and feet. He firstly noticed a loss of appetite 1 year prior to OPD visit. 9 months later, he had developed progressive non-productive cough without neither fever nor night sweats. He refused to have any abdominal symptoms or arthralgia. Prior treatment with oral antibiotics was unsuccessful.

Past history:
- He has underlying of type 2 diabetes mellitus and hypertension.
- He has been diagnosed with aortic aneurysm for 3 years.
- Pancreatic mass was being detected accidentally on imaging at the same time. Expectant management was considered to be the choice of treatment.
- He was an ex-smoker.
- He has quit drinking alcohol for 20 years.

Family history: There was no family history of cancer or similar cutaneous lesion.

Physical examination:
- He looked mildly pale
- Other systemic examination revealed no abnormality, no lymphadenopathy, and no hepatosplenomegaly.

Dermatological examination: (Fig 26.1, 26.2)
Multiple discrete ill-defined, erythematous to brownish, edematous, and tender subcutaneous nodules on both legs and feet

Histopathology: (S19-010034, Left leg) (Fig. 26.3, 26.4)
- Lobular panniculitis with numerous neutrophils and basophilic degeneration
- Numerous ghost cells and saponification are noted

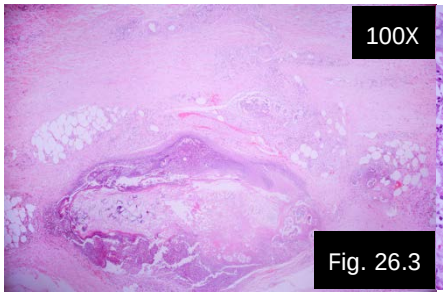

Fig. 26.3

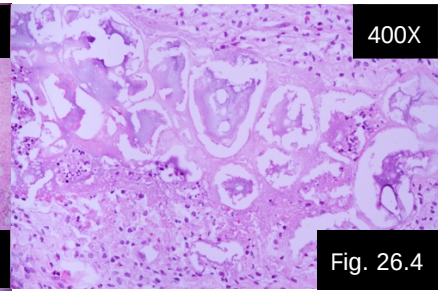

Fig. 26.4

Laboratory investigations:

- CBC: Hb 10.5 g/dL, Hct 31.7%, MCV 84.5 fL, Plt 501,000 /mm^3, WBC 15,750 /mm^3 (N 86%, L 8%, M 6%)
- Lipase: >20,000 U/L (23-300 U/L)
- Amylase: 103 U/L (30-110)
- AST/ALT: 39/11 U/L
- ALP/GGT: 138/117 U/L
- BUN/Cr: 18/0.83 mg/dl

- CXR: Focal increased patchy opacity in the right middle lung and solitary pulmonary nodule in the left upper lung
- CT chest and abdomen: Few large conglomerate masses with internal necrosis along the pancreas, suggestive of pancreatic cancer with finding of pulmonary and hepatic nodules, as well as few osteolytic lesions at right iliac bone, suspicious of metastasis

Diagnosis: Pancreatic panniculitis associated with pancreatic cancer with lung, liver, and bone metastasis
Treatment: Supportive treatment due to advance stage of cancer
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Consultant: Phatcharawat Chirasuthat, MD

Discussion:

Pancreatic panniculitis is a rare form of panniculitis, first described by Chiari in 1883, occurring in 2-3% of patients with pancreatic disorders which usually described in association with benign diseases, such as acute pancreatitis, mainly secondary to alcoholism, cholelithiasis, chronic pancreatitis, and less commonly with pancreatic carcinoma, mostly in acinar cell carcinoma.[1,2] The incidence rate is higher among males with alcoholism.[3] Interestingly, even though it was named as pancreatic panniculitis, it can be associated with non-pancreatic conditions , such as neuroendocrine carcinoma of adrenal gland, sulindac therapy, or intrahepatic cholangiocarcinoma.[4,6] Other recently reports of benign conditions associated with pancreatic panniculitis were pancreatic-portal fistula, systemic lupus erythematosus[7] or even in pancreas-kidney transplantation in association with acute pancreas allograft rejection patient.[8,9]

The exact pathogenesis of pancreatic panniculitis is still unclear, but release of pancreatic enzymes such as trypsin, amylase, lipase and phospholipase may lead to fat necrosis concomitant with inflammatory reaction.[1,4] However, there are reports of pancreatic panniculitis in the setting of normal serum levels of pancreatic enzyme.[2] These findings lead to the hypothesis that pancreatic enzymes may not be the only etiological factor, an immunological process, such as alpha–antitrypsin deficiency, can also initiate subcutaneous fat necrosis.[2]

The cutaneous lesions of pancreatic panniculitis are usually presented with ill-defined erythematous to red-brown edematous and tender nodules that may spontaneously ulcerate and drain an oily brown, sterile, viscous substance resulting from liquefaction necrosis of adipocytes. These lesions typically located on the lower extremities, especially around the ankles and pretibial areas.[4] Although there are no specific clinical findings, it seems that pancreatic panniculitis associated with pancreatic carcinoma seems to be more persistence with frequent recurrences, ulceration, fistulization, and involvement of cutaneous areas beyond lower extremities.[3] Extracutaneous involvements can cause acute arthritis, necrosis of abdominal or bone marrow fat, polyserositis, mesenteric thrombosis, leukemoid reactions, eosinophilia and fever.[1] Monoarticular or oligoarticular arthritic symptoms have been reported in 50–80% of cases, mainly affects the ankles, knees and metacarpophalangeal joints, and association with osteonecrosis.[1] The association of pancreatitis, panniculitis and polyarthritis (PPP syndrome) is a very rare extra-pancreatic complication and associated with a poor prognosis, other than the Schmid's triad. A skin biopsy is a mandatory for diagnosis of this disease. The presence of neutrophilic lobular panniculitis with the typical ghost cells and focal calcification in the saponified fat lobules are characteristic.[1,2,4] Laboratory investigations usually reveal elevated serum amylase, lipase, or trypsin, but not in all cases and degree of panniculitis does not correlate with enzyme levels. Leukocytosis and an increase of acute phase reactants usually occur.[1] Eosinophilia and leukemoid reactions are also commonhematological abnormalities, particularly in patients with pancreatic carcinoma.[1]

Treatment for pancreatic panniculitis is based on treatment of the underlying pancreatic disease. Prognosis of pancreatic panniculitis depends on the prognosis of the underlying disease and the extent of fat necrosis.[1] Zhang et al. reported a case of pancreatic panniculitis

associated with pancreatic mucinous adenocarcinoma with increased of serum amylase, lipase, and carbohydrate antigen 19–9. After the administration of octreotide acetate, a synthetic polypeptide that inhibits pancreatic enzyme production, and Whipple procedure, the serous amylase and lipase levels returned to normal, and the pancreatic panniculitis had almost resolved by 4 weeks later.[2] According to our case with advance stage of pancreatic cancer, the management was supportive care. Finally, the patient died due to the advance stage of disease.

References:

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