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Pancreatitis, Panniculitis, and Polyarthritis Syndrome with a Fatal Course

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Dear Editor:

Pancreatitis, panniculitis, and polyarthritis (PPP) syndrome is a rare disease characterized by pancreatic disease, panniculitis presenting as tender erythematous nodules especially on the lower extremities, and polyarthritis showing swelling and inflammation without evidence of infection[1]. Prompt diagnosis is crucial because of its poor prognosis and high mortality with delayed treatment[2].

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An 64-year-old male visited the emergency department with pain on both legs for five days. He had history of frequent alcohol consumption and hypertension. The laboratory examination revealed leukocytosis (28,690/μl) with neutrophil dominance (88.7%), elevated amylase (9,055 U/L; 28~100 U/L), lipase (2,089 U/L; 13~60 U/L), C-reactive protein (12.94 mg/dl; 0~0.5 mg/dl), creatinine (2.71 mg/dl; 0.7~1.2 mg/dl), and blood urea nitrogen (41.0 mg/dl; 6~20 mg/dl). The peripheral blood cell morphologic analysis revealed normocytic normochromic anemia and neutrophilia with left-shifted maturation. The blood test revealed no evidence of viral infections, autoimmune diseases, or hematologic malignancies. There was no fever or abdominal pain, but abdominal computed tomography showed acute pancreatitis. He was admitted to the department of internal medicine and was consulted to the dermatology department for tender erythematous subcutaneous nodules with periarticular swelling on the knees and ankles (Fig. 1). A skin biopsy of the left ankle showed lobular

panniculitis with neutrophils and fat necrosis. The fat necrosis comprised pathognomonic anucleate ghostlike adipocytes. Basophilic granules comprised calcification in the cytoplasm of the necrotic fat cells (Fig. 2). The biopsy result was consistent with pancreatic panniculitis. The joint pain in both ankles, knees, and elbows persisted despite the administration of nonsteroidal anti-inflammatory drugs, antibiotics, and corticosteroids. Joint fluid aspiration showed suppurative inflammation without identified organisms. Based on the radiologic, clinical and pathologic findings, a diagnosis of the PPP syndrome was made. The patient underwent total pancreatectomy for the uncontrolled pancreatic enzyme level. The specimens showed chronic pancreatitis without malignancy. Despite the appropriate operation, the patient died 78 days after admission from acute kidney failure with pleural effusion.

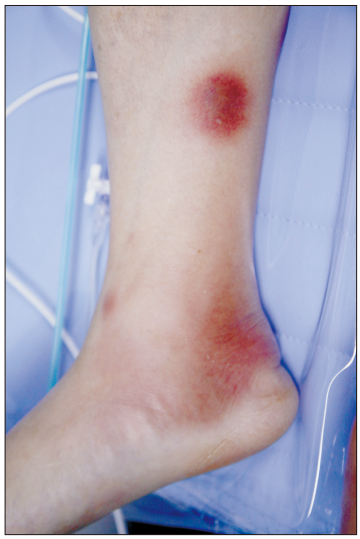

Fig. 1. Clinical photograph showing ill-defined erythematous edematous tender subcutaneous nodules with swelling around the ankle. We received the patient's consent form about publishing all photographic materials.

PPP is a rare syndrome of elevated serum pancreatic enzymes, lobular panniculitis presenting as painful erythematous nodules, and swollen peripheral joints without infection[1,2]. Only one case of PPP syndrome is reported in Korean dermatologic literature[3]. The PPP syndrome typically occurs in middle-aged males with a history of alcohol abuse[4], and two-thirds of patients show absent or mild abdominal symptom[1]. The exact pathophysiology of PPP syndrome is unclear. It is hypothesized that release of pancreatic enzymes, especially lipase, due to underlying pancreatic disease results in fat necrosis and secondary inflammation in multiple sites such as visceral organs, joints, and the skin[5]. In our case, skin and joint lesions presented without abdominal symptom. Differential diagnosis for tender subcutaneous nodules includes erythema nodosum, erythema induratum, and infectious panniculitis. However, male predominance, elevated pancreatic enzyme levels, and the histopathologic findings of enzymatic fat necrosis consisting of ghostlike adipocytes and calcification favored the diagnosis of PPP syndrome. The delay in diagnosis and treatment of the underlying pancreatitis results in poorer prognosis, with the mortality rate as high as 24%[1]. We knew the underlying pancreatitis thanks to early and thorough blood test in the emergency department, but we might have missed or delayed the diagnosis if we had waited for skin biopsy results without checking pancreatic enzymes. Thus, physicians should consider PPP syndrome and check for history of alcohol abuse and the serum pancreatic enzyme level when a patient presents painful erythematous subcutaneous nodules and arthritis, even when there is no abdominal pain. Herein, we report a fatal but instructive case of the PPP syndrome.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

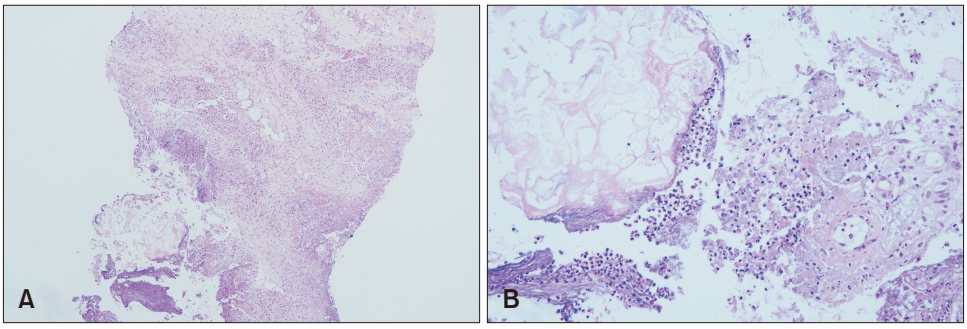

Fig. 2. (A) Lobular panniculitis with neutrophils and fat necrosis (H&E, ×40). (B) Enzymatic fat necrosis consisting of distinctive ghostlike adipocytes with thick, faintly stained cell peripheries and loss of basophilic nuclear staining. Inflammatory infiltrate surrounding the foci of fat necrosis. Basophilic granules comprising calcification in the cytoplasm of the necrotic fat cells (H&E, ×200).

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