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## Pancreatitis, panniculitis and polyarthritis syndrome caused by pancreatic pseudocyst with vena caval fistula

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**Introduction**: PPP syndrome is a triad of panniculitis, polyarthritis and pancreatitis. This syndrome is extremely rare extrapancreatic complication of pancreatic disease. We report a case of a patient with PPP syndrome due to formation of the fistula between inferior vena cava and pancreatic pseudocyst.

**Methods**: A 64-year-old man came to the hospital with bilateral leg pain that began a week ago. He had no specific disease other than hypertension. The vital signs were normal. The blood test revealed that WBC 28690 /uL, amylase 9055 U/L, lipase 2089 U/L, CRP 12.94 mg/dL. Recent acute pancreatitis was found on pancreas CT. NSAIDs were administered, but there was no improvement. After the steroid administration, the pain was slightly improved. Skin lesions were diagnosed as panniculitis. Bone scan and knee MRI revealed multiple osteoarthritis and bone marrow infarction.

**Results**: Surgical treatment was considered and total pancreatectomy with splenectomy was performed. A pseudocyst was present in the posterior side of the pancreas head, forming a fistula with suprarenal IVC. After surgery, amylase and lipase decreased. But the patient died of an uncontrolled infection on the 13th postoperative day.

**Conclusions**: PPP syndrome should be suspected if it accompanies with skin lesions and joint lesions. Delays in diagnosis can have catastrophic consequences. It is sure way to treat underlying pancreatic disease. The fistula into the systemic circulation is usually reported with mesenteric vein or portal vein, but the fistula with IVC like this case is reported for the first time in the literature.

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