

Pancreatitis, Panniculitis, Polyarthrititis (PPP) Syndrome Caused Post-Pancreatitis Mesenteric Vein Necrosis

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DESCRIPTION

Pancreatic disease can rarely express itself by extra-abdominal pathological processes. The symptomatic triad of pancreatic disease, panniculitis and polyarthrititis, also known as PPP syndrome, is characterized by severe chronic episodes and high mortality rate. The pathognomonic hallmark of PPP syndrome is fat necrosis in affected tissues. It is essential to identify its indicators due to complicated diagnosis procedure as a result of absent or mild abdominal symptoms.

Ascites, excessive oedematous changes in multiple organs were identified, whereas no pancreatic parenchymal changes were observed. Finally, this condition was named as PPP syndrome. Knowledge of association between panniculitis and polyarthrititis with acute pancreatic disease may lead to a prompt diagnosis and management. Histopathological features of skin lesions can be a valuable clue for focusing attention to a pancreatic disease

Pancreatitis, panniculitis and polyarthrititis syndrome (PPP-syndrome) may be a very rare symptom complex of additional pancreatic manifestations of pancreatitis with lobular panniculitis and (poly-) arthrititis with intra osseous fat necrosis. Although the pancreatic pathology with exceptional high blood levels of pancreatic enzymes (in particular lipase) is widely considered causative, abdominal symptoms are often mild or absent. This results in misdiagnosis, delay of appropriate therapy and worsening of prognosis [1].

Here we report a case of PPP-syndrome with extensive intraosseous fat necrosis in multiple bones and polyarthrititis. Outstanding in this case was the extremely high blood level of up to 600-fold elevated lipase caused by a pancreatic pseudocyst with contact and signs of pancreatitis in clinical symptoms, computed tomography and ultrasound it had been not related with the opposite symptoms. By this treatment, the elevated lipase levels and CRP-values were normalized, but joint pain was only partially reduced. After 2 weeks, the patient was readmitted with heavy pain in his left knee with distinct swelling and reddening.

In minor extent, he suffered also from pain within the left shoulder, wrists, cubital and both ankle joints. He added that 9 months ago, he suffered from similar symptoms with multiple joint pain. The patient had a history of acute, necrotizing head-pancreatitis caused by heavy alcoholic abuse 3 years ago, but now he clearly negated any abdominal symptoms and was plausibly abstinent of alcohol.

A cystic lesion of the uncinate process of the pancreas with constant size was described in imaging for 3 years. A key learning point is that the high value of vena mesenterica thrombosis as an indirect radiological sign of tiny fistulas. A PPP-syndrome has got to be taken into consideration if the origin of any osteonecrosis, polyarthrititis or panniculitis remains unclear, considering the high morbidity and mortality. Absent abdominal symptoms can delay the diagnosis of the underlying pancreatic disease.

Measuring of lipase, skin or bone biopsies, which show pathognomonic features, and if appropriate surgical exploration are the foremost important diagnostic procedures. Counting on the kind of the underlying pancreatic disease correct treatment can be successful and cause complete remission.

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Received: January 04, 2020; **Accepted:** January 18, 2020; **Published:** January 25, 2020

Citation: Musteikaite D (2021) Pancreatitis, Panniculitis, Polyarthrititis (PPP) Syndrome Caused Post-Pancreatitis Mesenteric Vein Necrosis. *Rheumatol Curr Res.* S7:003

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