



PPP Syndrome Caused by Pancreatic Pseudocyst with Fistula to Inferior Vena Cava

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Abstract

Introduction: In rare cases pancreatitis can manifest in extra-abdominal symptoms as polyarthritits and panniculitis leading to PPP syndrome.

Case Report: 34-year-old man was diagnosed with a pancreatic pseudocyst draining to Inferior Vena Cava. Because of the damage done by circulating pancreatic enzymes intraosseous fat necrosis, polyarthritits, panniculitis symptoms appeared. Surgical closing of the fistula was performed. As a result, pancreatitis and panniculitis were successfully treated while arthritis symptoms remained.

Discussion: Most often PPP syndrome appears due to the pancreatitis. The exact pathophysiological chain of the development of extra abdominal symptoms is not clear, however some authors suggest it might be the result of pancreatic lipase entering bloodstream. Pancreatic enzymes circulating in the blood tend to damage adipose tissue in the lower limbs causing the signs of panniculitis. The manifestation of arthritis differs depending on the clinical case as it can involve different joints, vary in number and symmetry. The best outcomes are usually achieved by performing the surgery, while conservative treatment lacks effectiveness.

Conclusion: Pancreatitis, pancreatic polyarthritits and panniculitis simultaneously are diagnosed in rare cases. However, it is important to include PPP syndrome into early diagnostics to improve the prognosis of the condition.

Introduction

PPP syndrome is a rare pancreatic disease complicated by panniculitis and polyarthritits [1-5]. In rare cases pancreatic cysts can form fistula to the adjacent blood vessels. Pancreatic juice draining to the blood circulation cause damage to the capillary and lymphatic vein walls [6]. Further damage to the surrounding tissue manifests in panniculitis and polyarthritits symptoms [2].

We present a case of PPP syndrome in a patient with acute pancreatitis alongside pancreatic pseudocyst fistula to Inferior Vena Cava. In this report, surgery is described as the method of choice to treat the condition.

Materials and Methods

To describe a rare case of PPP syndrome with fistula between pancreatic pseudocyst and Inferior Vena Cava by highlighting its radiological images to guide management and describing surgical technique.

Case Presentation

A 34-years-old man without previous history of chronic disease arrived at the regional hospital because of the intense pain in the epigastric area, nausea, vomiting, swollen, painful hand and leg joints with the restricted kinetic function. Arthritis symptoms were significantly pronounced in the joint of the left knee. Patient admits chronic use of alcohol for the past 1.5 years. The diagnosis of moderate severity acute pancreatitis was made based on clinical presentation, elevated serum amylase (1735 U/L) and ultrasound investigation. Crystalloid infusions, adequate pain management

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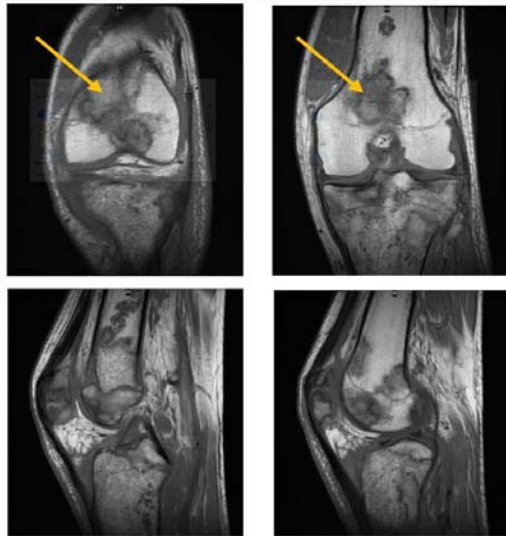
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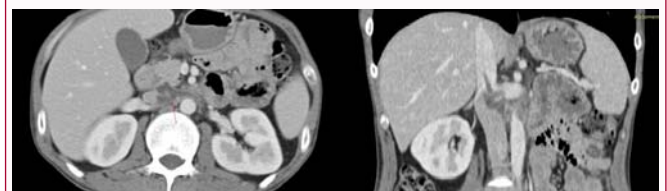
Picture 1: MRI images of the left knee. Multiple damage zones due to lipolysis in femur, patella and tibia.



Picture 4: Extent of the arthritis. The extent of swelling before the surgery in the cachexic patient compared with the hand of the nurse. The elbow of the same patient after 6 months – swelling resorbed.



Picture 2: The site of Paniculitis damage. Degraded adipose tissue localized in subcutaneous layer of skin.



Picture 5: Abdominal CT images of the pseudocyst with fistula to vena cava inferior.



Picture 3: Aspirate from the left knee joint. Brown, opaque liquid, containing tissue detritus and lipid degradation particles, aspirated from the site of shin affected by panniculitis.

(Ketoprofen and Metamizole), starvation diet was prescribed. CRP (185.7 mg/L) was significantly elevated during the time of admission. Therefore, Cefuroxime and Dexamethasone were administered. As the amylase concentration fluctuated wildly, patient's condition did

not improve, transfer to Klaipeda University Hospital (KUH) was arranged.

At the time of arrival, the patient was stable, but previously described symptoms, especially arthralgia, abdominal pain, were expressed. Urgent abdominal computer tomography revealed typical signs of pancreatitis without necrosis, soft tissue infiltration up to 12 mm around pancreas, 4 mm local thrombus in Inferior Vena Cava superiorly to the right renal vein branch. During the first days of conservative treatment, intensity of the abdominal pain decreased, but arthralgia was not manageable even with several analgesics, including Tramadol. As typical inflammatory signs were expressed in the left knee, synovial liquid was collected for the microbiological examination. However, growth of the pathological bacteria was not detected. Magnetic resonance images of the knee showed multiple acute infarcts of the bone marrow of femur, patella and fibula, signs of synovitis and bursitis (Picture 1). Simultaneous abdominal MRI was performed. The infiltration around the pancreas started to resorb. Therefore, a pseudocyst with a connection to the pancreatic duct was visualized.

Later patient's condition started to deteriorate as the pain of left knee, elbow and both ankle joints became more pronounced, abdominal pain returned, erythema and swelling in left shin appeared. Arthralgia could only be managed by prescribing Morphine. Puncture of the damaged tissue in the shin was done (Picture 2). Aspirated liquid is shown in the Picture 3. Cytologic examination of the liquid showed tissue detritus and lipid degradation particles. Consequently, blood lipase concentration was tested and it came

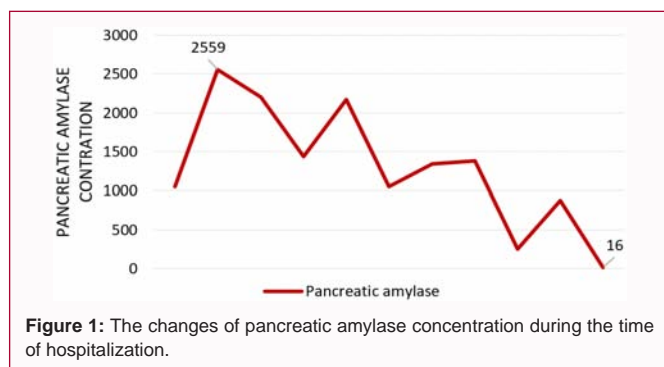


Figure 1: The changes of pancreatic amylase concentration during the time of hospitalization.

back 52 times higher than normal range (8404 U/L). At this point PPP syndrome was diagnosed because of the pancreatitis, multiple arthritis, panniculitis (Picture 4). Abdominal CT was repeated to determine the cause of the syndrome. Liquid collection medially to Inferior Vena Cava with the wide connection to its wall was observed. It was deduced that pancreatic pseudocyst had a fistula to Inferior Vena Cava and the thrombus observed in the primary CT was the opening of the canal (Picture 5).

A laparotomy was performed under general intubation anesthesia. During the surgical exploration the pancreas was stiff only in the head area, the body and tail were soft with normal pancreatic structure. Duodenum was mobilized by Kocher manoeuvre. Pancreatic pseudocyst of 3 cm × 4 cm × 3 cm in size and surrounding inflammation in the posterior aspect of pancreas head were observed. After the mobilization of Inferior Vena Cava, pancreatic pseudocyst was opened. Old blood clots were found inside of it. During the removal of the clots, an inferior vena cava fistula opened. Fistula was located at the conjunction of the left renal vein and inferior vena cava. The fistula was closed with monofilament suture. Connection of the pseudocyst and pancreas was visualized and sutured. Area of the pseudocyst was drained. Postoperative course was uneventful.

During the follow-up period of 3 years, panniculitis symptoms disappeared, but arthralgia in the left knee joint persists to this day. Kinetic function of the left knee is restricted because of the pain, but patient is capable of walking without the support. No analgesics are used for the pain management (Figure 1).

Discussion

In rare cases, pancreatic disease can be complicated by panniculitis and arthritis. The triad leads to the diagnosis of PPP syndrome. However, due to the limited number of reported cases the conception of PPP syndrome is not clear: The primary cause, extent of the damage differs depending on the case [1].

Literature review of 148 cases found that 49% and 46% of cases were caused by pancreatitis and pancreatic cancer respectively [7]. Although pathophysiology of PPP syndrome is not fully understood, this case supports the hypothesis that increased concentrations of circulating pancreatic enzymes cause the tissue damage [8]. In this case, pancreatic juice collected in the pseudocyst. We believe that when the pressure inside of the pseudocyst reached the pressure of Inferior Vena Cava, enzymes entered systemic circulation through the fistula between the two. Further, damage to the lymphatic vessels increased permeability of the wall. Pancreatic lipolytic enzymes hydrolyzed fats producing glycerol and free fatty acids within the tissue. Because of the lipolysis, inflammatory response developed and caused the symptoms [9].

Studies show that pancreatitis caused by alcohol abuse is prone to formation of pseudocyst as it accounts for more than ¾ of cases [10]. Although pseudocysts usually remain asymptomatic, it played significant role in PPP syndrome pathogenesis in this case. Clinically, only 54% of patients exhibit gastrointestinal symptoms, for example pronounced abdominal pain [1]. Pancreatic enzymes usually affect organs and systems with the highest quantity of adipose tissue: Mostly subcutaneous fat and bone marrow. Most often, pancreatic panniculitis is described as multiple erythematous nodules localizing mostly in lower limbs [11,12]. However, this patient developed panniculitis 1 month from the onset of arthralgia and the damage was localized in the left shin. Joint damage varies depending on the case. In the study of 59 case reports, more than 4 joints were damaged in ¾ of patients. Arthritis symptoms being expressed in the major limb joints, such as ankle, knee and elbow [1,13-15].

The treatment must concentrate on the pancreatic disease, as panniculitis and arthritis symptoms improve simultaneously [10]. Positive outcomes of the syndrome are usually described following partial or total pancreas resection as well as endoscopic stenting of disrupted pancreatic duct. The surgical strategy should be designed to prevent pancreatic enzymes from entering systemic circulation [2,16]. In this case successful treatment by suturing Vena Cava fistula and pancreatic duct was achieved. In the literature review, 78.6% of patients poorly responded to conservative treatment, for example NSAID, steroids or immunosuppressants [1]. Although some authors mention Octreotide as it could decrease the secretion of pancreatic enzymes, low numbers of PPP syndrome cases restrict the research on the drug [10].

Conclusion

We reported a rare case of pancreatitis with fistula between v. cava inferior and pancreatic pseudocyst. The condition being associated with subcutaneous and intraosseous fat necrosis. The possibility of PPP syndrome should be considered in situations of unexplained polyarthritis or panniculitis. If the syndrome is caused by pancreatitis, secondary radiologic imaging after the infiltration resorbed might be needed to confirm the diagnosis.

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