Pancreatitis, Panniculitis, Polyarthritis Syndrome (PPP syndrome): A Case Report

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Abstract*—Pancreatitis presenting without abdominal pain is very unusual. A 35-year-old man with acute pancreatitis presented with prolonged fever, polyarthritis and multiple subcutaneous swellings, but without any abdominal pain. His serum lipase and amylase were very high. Biopsy from the subcutaneous swellings revealed fat necrosis and CT scan abdomen showed features of acute pancreatitis. He was managed conservatively. The available literature on this syndrome was reviewed.*

Keywords**: **Pancreatitis, Panniculitis, PPP syndrome.

I. INTRODUCTION

The pancreatic disease, panniculitis, polyarthritis (PPP) syndrome was first described by Berner in 1908[1]. Although the pancreatic pathology with high blood levels of pancreatic enzymes is considered causative, abdominal symptoms are often mild or absent. The delay in diagnosis and specific treatment of the underlying pancreatitis worsens the prognosis of this condition, which has a high mortality rate[2]. In nearly 45% of the patients, the arthritis follows a chronic course with a poor response to nonsteroidal anti-inflammatory drugs and corticosteroids, and the rapid development of radiographic joint damage.

As this is rare cases so the case was thoroughly evaluated and a case report was prepared to publish.

II. METHODOLOGY

A rare case of pancreatic disease, panniculitis, polyarthritis (PPP) syndrome was presented in Medicine department of SMS Medical College, Jaipur (Rajasthan) India. As it is a rare case so evaluated thoroughly to prepare a detailed case report to published.

III. CASE REPORT

A 35 yr. Old chronic alcoholic male was admitted with chief complaints of fever, multiple joint pain and nodular lesion beneath skin from 15 days. There was no complaint of any migratory joint pain, palpitation, palmarerythma, abdominal pain, loss of appetite or decreased urine output. He had taken multiple courses of oral antibiotics over the past 15 days but his symptoms continued to worsen.

Figure 1
Reduced joint space and erosion on X-ray B/L knee joints

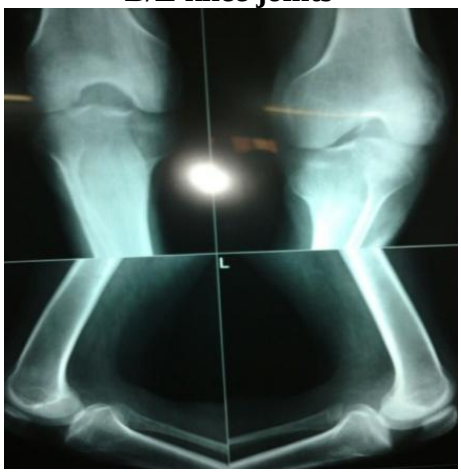


Figure 2
Reduced joint space and erosion on x-ray small joints of hands

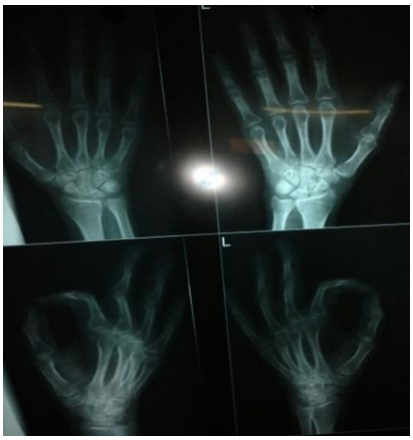
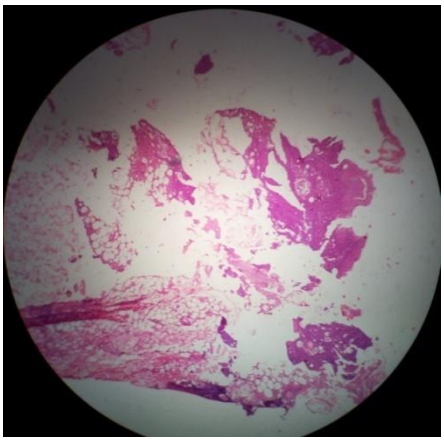


Figure 3
Subcutaneous Tissue shows Fat Necrosis



On Physical examination: Patient was febrile (104.2 µF) with multiple, discreet, tender, palpable nodular skin lesions of varied size from 2cm x 1cm to 1.5 cm x 2.5 cm over abdomen and calf region. Joint tenderness, local swelling and increased local temperature were present over both knee joints and over small joints of hands. Rest of the physical and systemic examination were within normal limits. His preliminary workup showed normal Renal function tests (RFTs) and Liver Function tests (LFTs), Serum albumin and normal urine examination findings. Total leucocyte count was markedly high (48500/µL) with 90% neutrophils and 05% lymphocytes. Peripheral Blood Film showed marked leukocytosis and shift to left. ASLO, RF and anti CCP antibodies were negative and HIV test was nonreactive. Chest X-ray was normal. X-ray both knee joints and small joints of hands revealed reduced joint space with periarticular erosions suggestive of polyarthritis (Figure 1 &2). Serum amylase (7900 U/L) and Serum lipase (26000 U/L) were markedly elevated. CECT abdomen was suggestive of acute pancreatitis (modified CTSI 4/10).

Next an excisional biopsy was taken from the subcutaneous nodule in view of the possibility of vasculitis syndrome. It revealed mild lymphocytic infiltrate in dermis. Subcutaneous tissue showed fat necrosis as evidenced by necrotic degenerating fat cells surrounded by abundant foreign body giant cell reaction along with few foamy histiocytes, no evidence of atypia or malignancy. Overall histopathology was suggestive of panniculitis with no evidence of vasculitis. No malignant cells or granuloma were

seen (Figure 3). With these pathognomonic findings the diagnosis of acute pancreatitis complicating into polyarthritis and panniculitis was made.

Patient had significant discomfort from his symptoms. As no specific treatment is available, the patient was kept nil per oral, managed conservatively with intra venous fluids and antipyretics. Patient was stable and recovered gradually and discharged 20 days later on oral antipyretics.

IV. DISCUSSION

The syndrome of pancreatitis, panniculitis, and polyarthritis has been rarely reported. The pathogenesis of this syndrome is unclear; however it is thought to be caused by release of pancreatic enzymes in the systemic circulation, which may lead to lipolysis and secondary inflammation in peripheral tissues, like subcutaneous tissue and joints.[10]

Pancreatic panniculitis can occur in 2-3% of patients with pancreatitis, first described in 1883 by Chiari.[3,4] Panniculitis is characterized by erythematous nodular lesions over lower limbs and abdomen. Pathognomonic histo-pathological findings are anucleate adipocytes with a thickened shadowy wall and granular basophilic cytoplasm ("ghost-like cell" adipocytes)[7,11,18]. Pain abdomen is usually absent and therefore pancreatitis is diagnosed late[2]. Serum lipase levels are exceptionally high and can rise even upto 80-100 times the upper normal limit[10,15]. Joint involvement is nonspecific and may present as monoarthritis as well as polyarthritis which can be asymmetrical or symmetrical.[1,19]

PPP syndrome can occur at any age, although the typical patient is a middle-aged male with history of heavy alcohol abuse[2,7,11]. Pancreatic pathology is considered as the main etiologic factor. It can precede, coincide or succeed the pancreatic disease. Pancreatic pathology can be acute or chronic pancreatitis, pancreatic carcinoma, neuroendocrine carcinoma, insulinoma, ischemic pancreatic disease, abdominal trauma or pancreatic duct stenosis.[5-7,11]

Overall mortality is about 24% when associated with acute or chronic pancreatitis and can be as high as 75% in cases of pancreatic carcinoma[2]. As there is no specific treatment available patients are managed conservatively, long term NSAIDs are often required.[2,5,6,10,11] Total pancreactectomy is practiced when there is evidence of pancreatic carcinoma and has shown to decrease mortality rate significantly.

V. CONCLUSION

'PPP' syndrome is a very rare clinical entity. Only about 66 cases have been reported till date. It is often misdiagnosed. Pancreatitis in these patients is usually silent with no clinical symptoms pertaining to the pathology and hence patient is diagnosed only late in the course. Histopathological demonstration of panniculitis is a must for diagnosis. Management is nonspecific and no specific treatment is available presently .Patients are treated for pancreatitis and often require long term NSAIDs for prolonged fever and Total pancreatectomy is indicated in cases of pancreatic carcinoma.

CONFLICT OF INTEREST

None declared till now.

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