Beth Israel Lahey Health
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A Case of Polyarthritis Panniculitis and Pancreatitis

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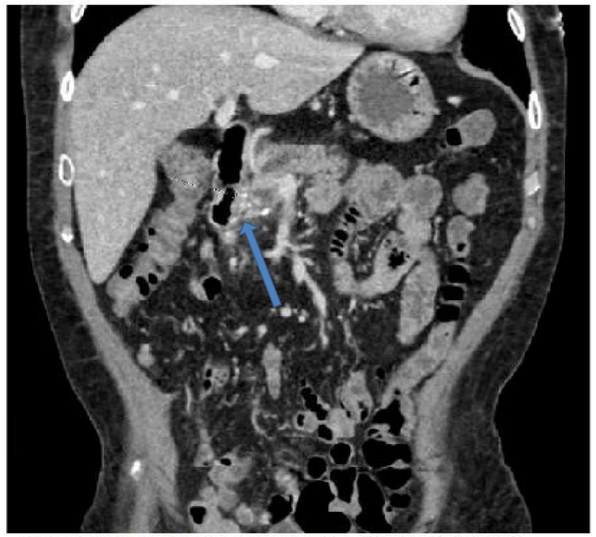
Introduction

Polyarthritis Panniculitis and Pancreatitis (PPP) is a rarely seen extra pancreatic morbidity hallmarked by the triad of joint pain (polyarthritis), tender skin lesions (panniculitis), and pancreatic inflammation (pancreatitis). The pathogenesis is mediated by pancreatic enzyme lipolysis of lipid rich skin and joint sites. Unfortunately, PPP is an elusive diagnosis given the minimal intrabdominal symptoms and unfortunately a delayed diagnosis may worsen prognosis by as much as 24%. As such, we aim to present a case of this rare diagnosis to familiarize clinicians with the diagnosis of PPP.

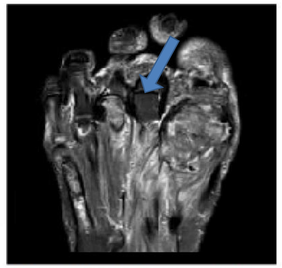
Case Background

67M with prior alcohol use disorder, recurrent pancreatitis, and complex pancreatic cyst (2.1 x 1.4 cm) status-post fine needle aspiration presented with fever, malaise, diffuse joint pain, and rash. Exam was notable for diffuse visible synovitis, tender joints, and subcutaneous nodules. Patient denied abdominal pain and was hemodynamically stable. Laboratory findings are shown in Table 1. Work-up including tickborne panel, Hepatitis B & C serology, Blood cultures, Urinalysis, Chlamydia, Gonorrhea, ASO, CCP, ACE, ANA, ANCA, IgG4, SSA, and SSB were all within normal limits. Right knee aspiration revealed straw colored fluid with 8552 WBC, 86 Neutrophils, no crystals, no growth on culture, and <3000 RBCs. CT of the Abdomen & Pelvis revealed complex cystic lesion of the pancreatic head measuring 2.2 cm, punctate foci of calcification, no pancreatic ductal dilatation, distal common bile duct within normal limits (Fig. A). MRI of the left foot revealed multifocal intramedullary osteonecrosis with bone marrow edema, multifocal synovitis, and prominent intermetatarsal bursitis (Fig B,C). Skin biopsy of the right thumb revealed lobar panniculitis with necrosis of adipocytes and residual “Ghost cells”. Patient was managed with IV methylprednisolone 80 mg, and prednisone taper. On Follow-up one month later, patient’s symptoms completely resolved. Follow-up MRI of the abdomen showed stable 2 cm pancreatic cyst which not consistent with underlying malignancy.

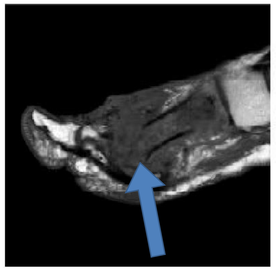
Imaging and Figures



a.) Peripancreatic inflammatory change and cystic lesion within the pancreatic uncinate process on CT Abd.



b.) Necrosis of 1st, 3rd, & 5th digits on STIR MRI



c.) L Foot Bone Marrow Edema of 1st digit on MRI

Figure 1

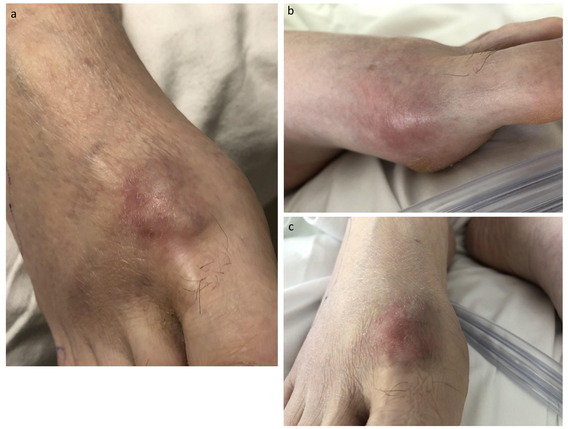


Image 1 a-c: Subcutaneous skin nodules of right (a,c) and left (b) 1st metatarsal

Table 1

CBC	Value
Hemoglobin	12.2 g/dL
Hematocrit	37 %
White Blood Count	12.4 K/uL
Platelets	304 K/uL
Chemistry	**Value**
Sodium	139 mmol/L
Potassium	3.8 mmol/L
Chloride	104 mmol/L
Bicarbonate	26 mmol/L
Blood Urea Nitrogen	18 mg/dL
Creatinine	0.87 mg/dL
Glucose	94 mg/dL
Total Bilirubin	0.68 IU/L
Alkaline Phosphatase	69 IU/L
AST	27 IU/L
ALT	34 IU/L
Lipase	20,521 U/L
C Reactive Protein	14 MG/L
Uric Acid	3.6 mg/dL

Discussion

PPP is a rare, but potentially devastating phenomenon which is most commonly observed in males with prior alcohol use. The phenotypic manifestations of the disease (skin nodules and synovitis) are thought to be mediated by lipolysis via systemically released pancreatic enzymes at lipid rich regions. The diagnosis often proves elusive given the lack of abdominal symptoms, but early treatment may be crucial in reducing mortality. While therapy is largely supportive; definitive resolution is achieved by treatment of the underlying pancreatic process.

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