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## Case Report

# Spontaneous pancreatic pseudocyst – superior mesenteric vein fistula: A rare complication of chronic pancreatitis

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## ABSTRACT

Pseudocyst formation is common in chronic pancreatitis. A rare subset of these patients may develop fistulization between the pseudocyst and the portal vein system. We report a case of spontaneous pancreatic pseudocyst – superior mesenteric vein fistula in a 61-year-old male with a history of chronic recurrent calcifying pancreatitis. The fistulous connection was correctly identified on both computed tomography and magnetic resonance cholangiopancreatography (MRCP), and the patient was treated successfully with a conservative approach. Our case report aims to educate on this rare and potentially fatal vascular complication of chronic pancreatitis and to discuss the role of modern noninvasive imaging techniques, such as T2-weighted MRI/MRCP, in establishing this diagnosis and making a decision regarding its management.

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## Case report

A 61-year-old male with past medical history of chronic calcifying pancreatitis due to previous alcohol abuse requiring multiple hospitalizations presented to the emergency department with sudden onset severe abdominal pain associated with nausea and vomiting. Exocrine and endocrine insufficiency was present and treated with enzyme supplementation and insulin. On admission the patient was afebrile and hemodynamically stable. Initial laboratory workup was significant for elevated lipase level of 277 and severe

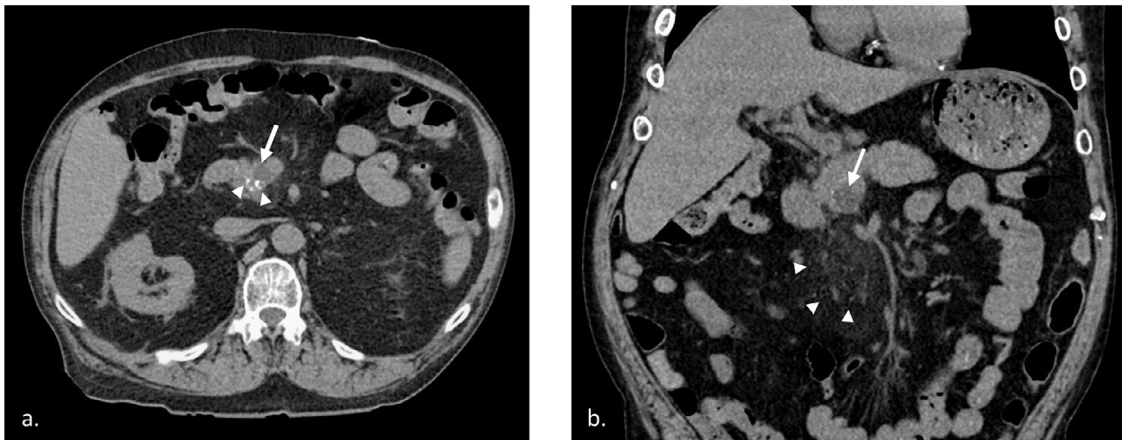
hypomagnesemia. Of note, the patient had multiple prior computed tomography (CT) examinations showing chronic calcifying pancreatitis, with the most recent study 2 months prior to hospitalization demonstrating a 1.3 cm pancreatic head pseudocyst (Fig. 1). A contrast-enhanced CT of the abdomen and pelvis was obtained, demonstrating pancreatic calcifications and a 2.5 cm hypoattenuating lesion in the pancreatic head with a slightly thickened wall, compatible with a pseudocyst. Short segment thrombosis of the SMV was identified near its confluence with the portal vein. An apparent communication between the lesion and adjacent SMV was suspected, as there was a central hypoattenuation extending from the lesion into the vessel (Fig. 2). This unusual appearance of SMV prompted further investigation for evidence of portal vein-pseudocyst fistula, given the proximity

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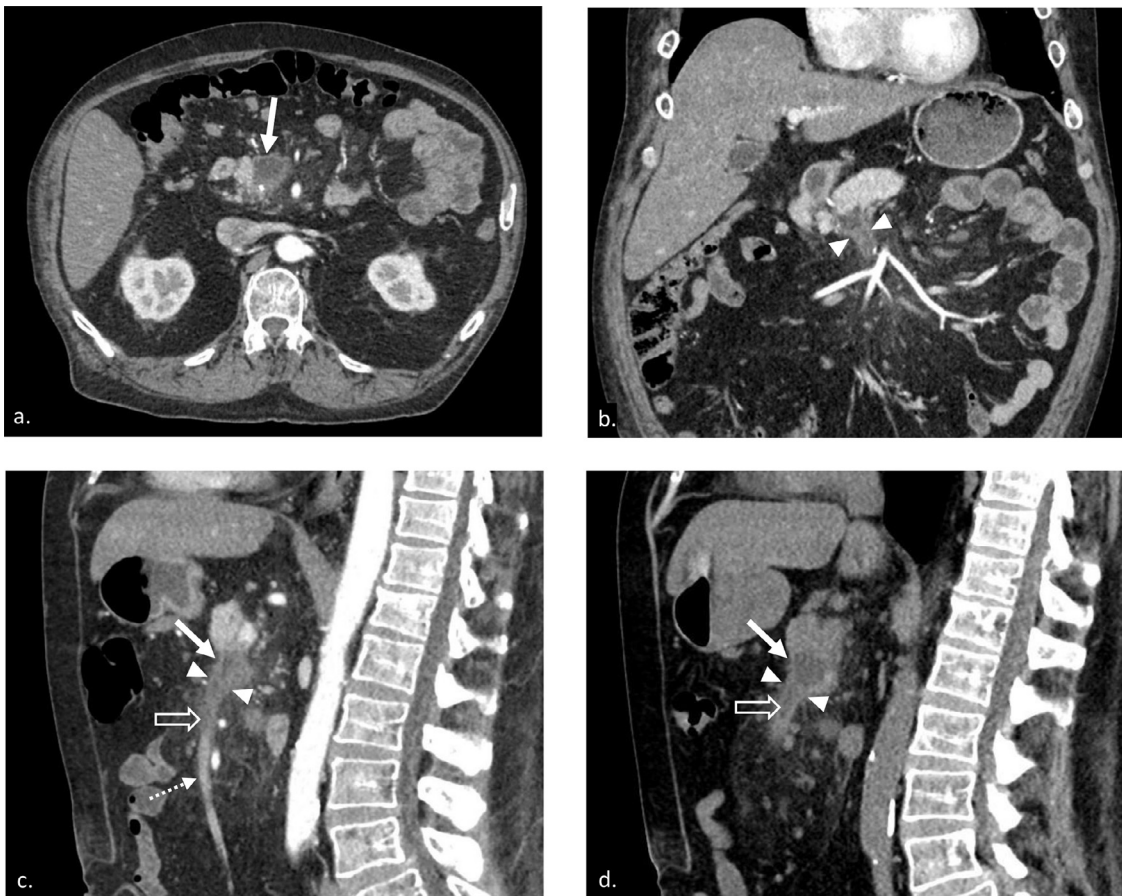
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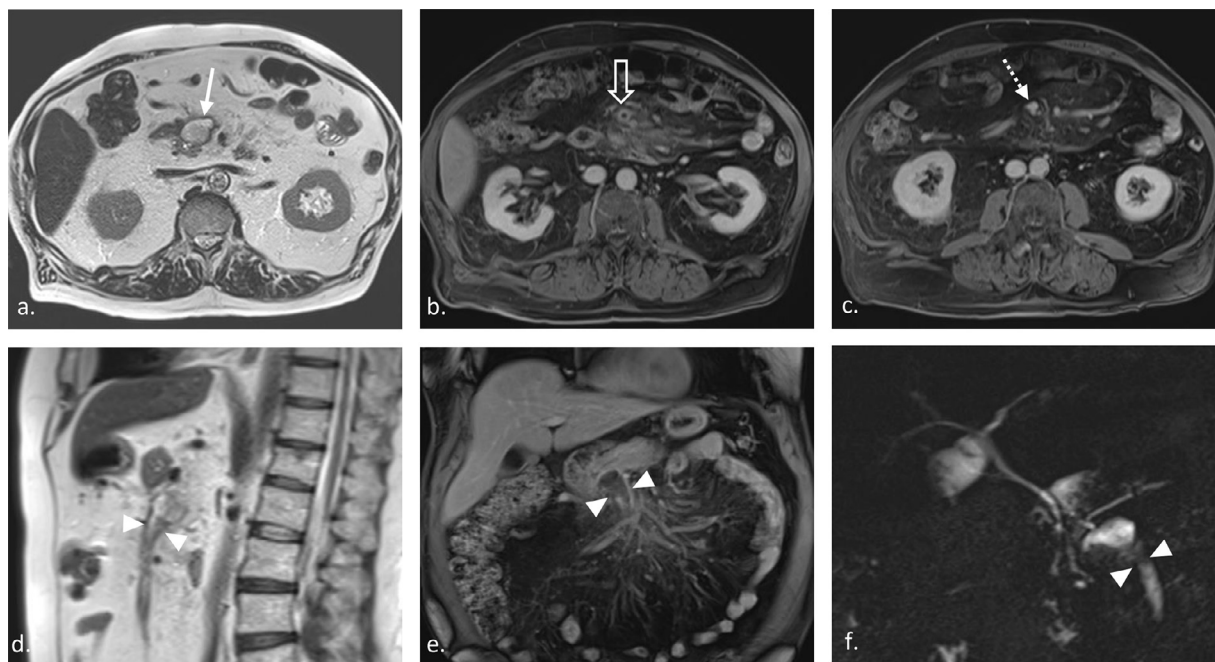
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**Fig. 1** – (a) Axial noncontrast CT image shows a small pseudocyst in the pancreatic head and multiple pancreatic parenchymal calcifications (arrowheads). (b) Coronal reconstructed CT image demonstrating pancreatic pseudocyst. Note increased attenuation of the mesentery (arrowheads), which in this case was chronic, reflecting sclerosing mesenteritis.



**Fig. 2** – Axial (a), coronal (b), sagittal (c) contrast-enhanced and sagittal noncontrast (d) CT images demonstrating pancreatic pseudocyst (arrow) with probable connection (arrowheads) with adjacent SMV. Note fluid attenuation of the proximal SMV (hollow arrow) and normal opacification with contrast of the distal SMV (dotted arrow).



**Fig. 3 – Axial T2-weighted (a) and postcontrast T1-weighted (b, c) images demonstrate pancreatic pseudocyst (arrow), low intensity thrombus in the proximal SMV (hollow arrow) and normal flow in the distal SMV (dotted arrow). Sagittal T2-weighted (d), coronal pos-contrast T1-weighted (e) and 3D MRCP reconstruction (f) images demonstrate the fistula between the pancreatic pseudocyst and proximal SMV (arrowheads).**

of the lesion to the SMV. A contrast-enhanced abdominal MRI/magnetic resonance cholangiopancreatography (MRCP) further demonstrated a 2.8 × 2.6 cm pancreatic head pseudocyst containing internal debris, showing a wide connection with adjacent SMV and abnormal fluid intensity signal in the long proximal segment of the vein, compatible with fistulization and thrombosis (Fig. 3). A decision was made by gastroenterology and surgical team to treat the patient conservatively, given a relatively small pseudocyst size, only mildly elevated lipase levels, and overall stable clinical condition. The patient underwent supportive medical management for symptom control and anticoagulation for SMV thrombosis. The patient's symptoms improved significantly, and he was doing well on 1-month follow-up.

## Discussion

Pancreatic pseudocyst formation is a well-known complication of chronic pancreatitis, reported in up to 40% of cases [1,2]. Vascular complications of chronic pancreatitis are also relatively common, occurring in up to 12% of patients [3]. While portal vein thrombosis and arterial pseudoaneurysms are well described in the literature, pancreatic pseudocyst - portal vein fistula (PPVF) is an extremely rare complication of chronic pancreatitis, with only a few cases reported till date [4]. The majority of these cases are alcohol-related and more often seen in men [4,5]. The mechanism of fistulization remains poorly understood. Some authors suggest that primary pathogenesis is related to portal vein thrombosis

resulting from mass effect and vessel compression by the pseudocyst, and serving as a nidus for fistula formation [6]. Others believe that high concentrations of pancreatic enzymes within the pseudocyst erode into adjacent structures and directly cause intravascular thrombosis [7]. It is important to note, however, that not all PPVFs are associated with pancreatic pseudocysts [6]. In some cases, a direct connection occurs between the portal system and main pancreatic duct. Several symptoms are observed in the patients with PPVFs, with severe abdominal pain and high serum amylase/lipase being the most common [5]. Serious complications include septic shock, systemic lipolysis, hemorrhage, and death. Systemic lipolysis (Weber-Christian disease) is a dreaded complication of PPVF due to release of pancreatic contents into the systemic circulation through the fistula, associated with a high mortality rate [3–5]. Clinically, patients present with polyarthritis, polyserositis, necrotic bone lesions, and purpuric nodules on the extremities. Biopsy of these nodules demonstrates subcutaneous lipolytic necrosis. Chronic complications reported in the literature include development of portal hypertension and portal biliopathy secondary to cavernous transformation of the portal vein [8].

Definitive diagnosis of PPVF in the setting of chronic pancreatitis can be challenging. Endoscopic retrograde cholangiopancreatography was historically considered the most useful method by several authors before the availability of modern noninvasive imaging techniques such as high-resolution CT and heavily T2-weighted MRI/MRCP imaging [5,9]. Recent retrospective studies demonstrated that all cases that underwent initial MRCP imaging were correctly diagnosed with PPVFs, while the cases initially evaluated with CT

were diagnosed only with portal vein thrombosis, and the correct diagnosis was eventually made with MRCP [3]. The classic imaging finding of PPVF on CT is a fluid-attenuated portal vein with possible periportal inflammatory changes, collateral vessels, and associated pancreatic pseudocysts [3]. These findings are particularly suspicious in patients with history of chronic pancreatitis. On MRI/ MRCP, a corresponding hyperintense central fluid signal is often seen within the portal vein on T2-weighted sequences. Direct visualization of a T2-hyperintense fistulous tract may be seen and can be highlighted by the use of 3-dimensional rendering techniques [3]. Not all fistulous communications, however, may be seen on MRI. Endoscopic ultrasound may be useful both for diagnosis and intervention, demonstrating complex fluid within the portal vein with absent flow on color Doppler. An important imaging pitfall is differentiating PPVFs from bland portal vein thrombosis due to hypercoagulable state or extrinsic compression, and pancreatic malignancy with tumor thrombus invasion of the portal venous system [4]. Unlike pancreatic neoplasms, pseudocysts demonstrate central fluid signal with only peripheral wall enhancement after contrast administration. Treatment strategies for PPVFs highly differ in literature, ranging from medical supportive treatment to surgical interventions, with varying outcomes. Most reported cases who survived required aggressive surgical treatment including portal venoplasty (for primary closure of the fistula) or pancreatectomy and pancreaticojejunostomy (for blocking the flow of pancreatic secretions) as a definitive treatment [10]. In 1 reported case, a pancreatic stent alone was successfully used to treat the fistula [11].

In summary, pancreatic pseudocyst-portal fistula is a rare, potentially life-threatening complication of chronic pancreatitis. The diagnosis in our case was established by contrast-enhanced CT and MRCP, and the patient responded well to medical management and anticoagulation. Findings of a fluid attenuation within the portal vein system on contrast enhanced CT in a patient with chronic pancreatitis and pseudocysts should raise suspicion for pseudocyst-portal fistulization. Subsequent imaging should be performed to definitively identify the fistula for possible surgical planning. MRCP is a reasonable diagnostic confirmatory study, that can be followed by endoscopic retrograde cholangiopancreatography if findings remain equivocal. Treatment decisions are made on a case-by-case basis, with conservative or

minimally invasive approach favored in the acute stage, as surgery carries a high risk of mortality and morbidity [10].

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