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Fistulising Pancreatic Mucinous Cystic Neoplasm to the Portal Vein

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ABSTRACT

Mucinous Cystic Neoplasms (MCNs) of the pancreas are rare, typically affecting women and carrying potential for malignant transformation. While complications like fistulas between MCNs and adjacent structures are rarely reported, this case presents the first documented fistula between a pancreatic MCN and the portal venous system. A 56-year-old woman with acute abdominal pain was found to have a 10.4 cm cystic mass in the pancreatic tail, consistent with an MCN. The lesion exhibited high-risk features. During her admission, a progressive venous filling defect, presumed to be a thrombus, developed despite anticoagulation therapy. Percutaneous thrombectomy revealed a communication between the splenic vein and the cystic mass, with mucinous content aspirated, confirming a fistula with direct extension of mucin into the vein. Distal pancreatectomy confirmed the diagnosis of an MCN with high-grade dysplasia and ovarian-like stroma. This case highlights the rare occurrence of a fistula between a pancreatic MCN and the portal venous system, underscoring the importance of early recognition for proper treatment and surgical planning.

Mucinous cystic neoplasms (MCNs) of the pancreas are rare cystic tumors that typically affect women in their 4-5th decade [1]. Risk factors include female gender. 95% are located in the pancreatic body or tail with a cyst in cyst appearance and whilst rarely contain calcification, when present appear as curvilinear density in the cyst wall on CT. The main pancreatic duct is often non dilated or deviated due to mass effect [1]. The diagnosis of MCNs is largely based on imaging findings, with Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) playing key roles in identifying their characteristic features. Endoscopic Ultrasound (EUS) and fluid sample analysis can be used as an adjunct [2].

The natural history of these lesions generally demonstrates a low aggressive behaviour, particularly when under 4cm in size [3]; however, they have the potential for malignant transformation, particularly when they exhibit certain high-risk features such as cyst size > 3cm, mural nodularity, dilated main pancreatic duct >6mm or peripheral egg shell calcification [3]. Historically, management is resection for the majority of patients with an MCN. However, European Guidelines [2] suggest a conservative approach with surveillance for asymptomatic MCN under 4cm without an enhancing nodule.

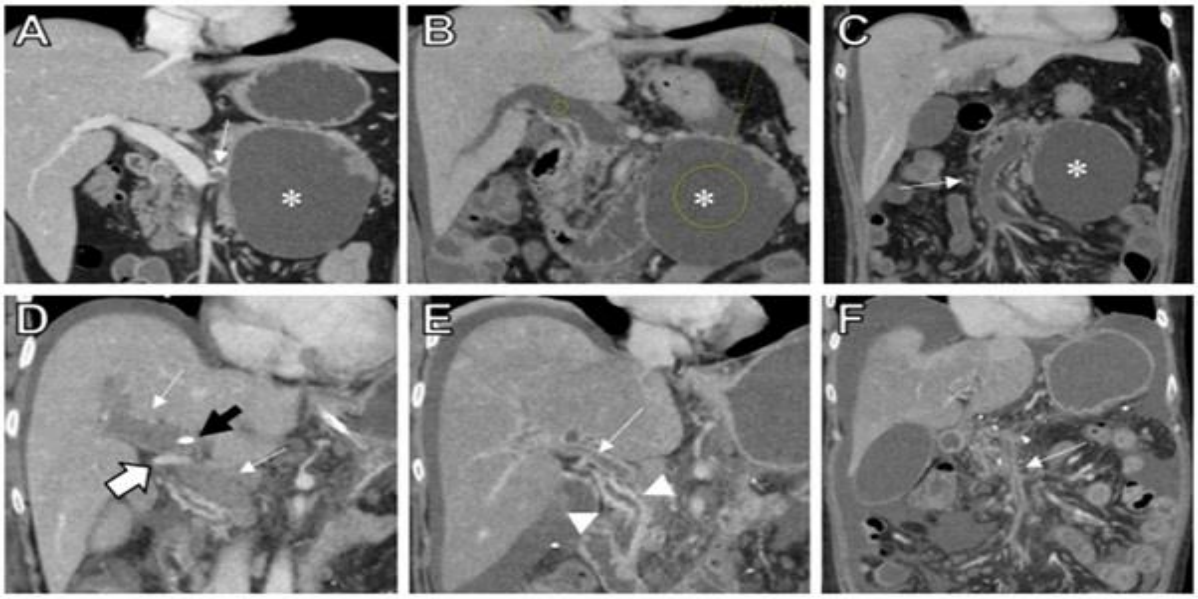
A fistula is an abnormal communication between two epithelialized surfaces. Fistulas associated with pancreatic cystic neoplasms, specifically IPMN have been described in case reports with an incidence of 1.9% [4], although more commonly cause pancreaticobiliary or pancreaticoduodenal fistulas [4]. However, IPMN with pancreaticovenous fistulas and extension of mucin into the vein have been described in case reports [5,6]. Although MCNs are well-documented in the literature, complications associated with these lesions, particularly those involving unusual fistulous connections, are rarely reported.

This case presents an exceptional occurrence of a fistula between a pancreatic MCN and the portal venous system, a phenomenon previously described in other types of pancreatic mucinous tumors, such as intraductal papillary mucinous neoplasms (IPMN). The significance of this case lies in its potential to inform clinical and radiologic practice, highlighting the need for awareness of such rare complications that may impact both the diagnostic and therapeutic approach. The early recognition of these features could improve patient outcomes by guiding more appropriate treatment strategies, including surgical intervention. Here, we discuss the clinical presentation, imaging findings, and management of this rare complication in a 56-year-old woman with a pancreatic MCN and portal venous fistula.

Case Presentation

A 56-year-old woman presented with acute abdominal pain to the Emergency Department. An abdominopelvic Computed Tomography (CT) demonstrated a large 10.4 cm cystic mass in the pancreatic tail with internal mural nodularity consistent with an imaging diagnosis of Mucinous Cystic Neoplasm (MCN). There were high-risk and worrisome imaging features according to the Kyoto guidelines [1], characterized by enhancing nodular thickening of the cyst wall over 0.5 cm and an overall size above 3 cm with enhancing mural nodules. Notably there was no associated dilatation of the main pancreatic duct (Figure 1). Her pain significantly increased during her hospital admission, and a follow-up CT demonstrated a new filling defect in the splenic vein, presumed to represent thrombus. Serial follow-up CTs showed progression of the filling defect, which gradually expanded through the superior mesenteric, splenic, and portal veins despite therapeutic anticoagulation (Figure 1). The portal vein filling defect had an HU of 4.6, similar to that of the primary tumour, which had a HU of 3.1.

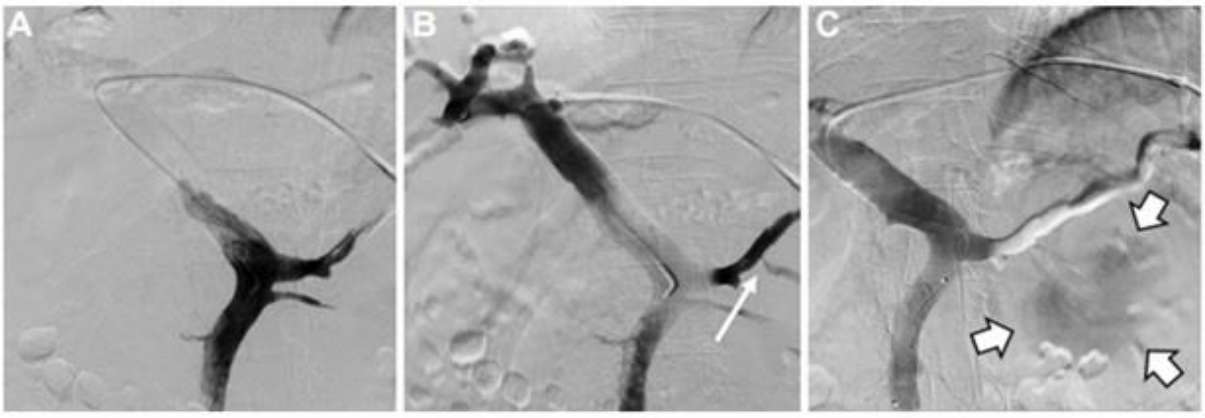
Figure 1. Contrast enhanced CT demonstrating the mucinous mass and its fistulization to the portal vein.



Note. Figure 1. Contrast enhanced CT abdomen and pelvis in portal-venous phase, coronal reformat. (A) Admission. Large pancreatic mucinous cystic neoplasm (*) with a solid irregular wall. Small thrombus within the proximal splenic vein (white arrow). (B, C) Images obtained 5 days after admission. B. Progression of thrombosis into the PV despite therapeutic anticoagulation. ROIs placed on the main PV and pancreatic tumor showed a density of 4.6 and 3.1 HU, respectively. The low density of the 'thrombus' within the PV should raise the concern for fistulation of the mucinous content from the pancreatic neoplasm. C. Occlusion extending into the SMV (white arrow). (D) Immediate status post distal pancreatectomy. Persistent portal vein occlusion (white arrows). Note the compensatory hepatic artery hypertrophy (thick white arrow). 5 Fr catheter that was kept within the PV (black arrow). (E, F) 1 month postop. E. Chronic portal vein occlusion (white arrow). There is secondary engorgement of the peribiliary plexus (arrow heads). F. Chronic SMV thrombosis (white arrow).

The patient was referred to interventional radiology and a mechanical percutaneous thrombectomy was performed. A vascular sheath was placed in the portal vein for aspiration of the thrombus. A dark thick gelatinous mucus drained from the port site as the wire was removed from the dilator, highly suspicious for mucin. The correct position of the catheter was then reconfirmed with a portal venogram. Fluoroscopy demonstrated an overt communication between the splenic vein and the large cystic mass consistent with a pancreatico-venous fistula. The injected contrast slowly filled the cavity within the pancreatic mass (Figure 2). The content aspirated from the vascular sheath in the portal vein was composed of mucin and hemorrhagic thrombus.

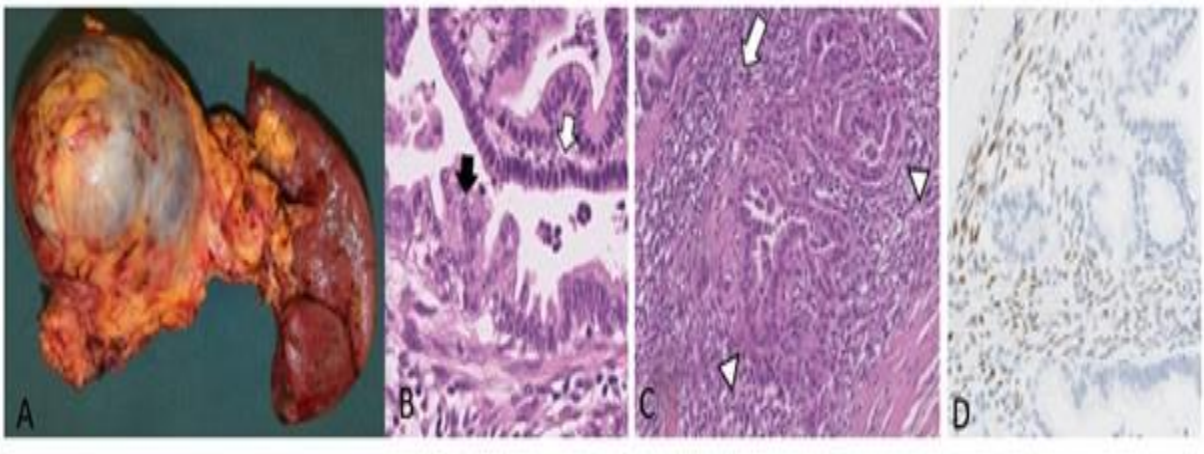
Figure 2. Angiographic images demonstrating a communication between the mucinous mass and PV.



Note. Figure 2. Conventional portogram through a 5 Fr catheter in the porto-mesenteric confluence, DSA images. (A) The main PV, proximal splenic vein, SMV and its branches are opacified. (B) Contrast leak from the mid splenic vein (arrow). (C) Extravasation of contrast into the pancreatic cystic tumor. Given the mucinous content aspirated through the sheath placed in the left portal vein consistent with fistulation of the pancreatic mucinous neoplasm into the splenic vein with extension into the portal system.

Following this discovery, the patient had a distal pancreatectomy (Figure 3). Pathology confirmed a multiloculated cystic neoplasm lined predominantly by mucinous epithelium with multifocal high-grade dysplasia with ovarian-like stroma in the wall of the pancreatic cystic neoplasm (Figure 3) consistent with the pre-operative diagnosis of MCN. There was no communication with the pancreatic duct system.

Figure 3. Histological images post resection.



Note. Figure 3. (A) Surgical specimen of distal pancreatectomy and splenectomy. (B,C) Microphotographs from the cyst show tall columnar mucin-secreting epithelium (white arrow in A) with multifocal high grade dysplasia (black arrow in B) and the subepithelium shows ovarian-type stroma in the wall (white arrow in C). A focus of atypical glands with high grade features (arrow heads in C), surrounded by reactive ovarian like-stroma is identified; these atypical glands are still confined to the wall of the cyst and not entirely fulfill the criteria for invasion. (Hematoxylin-eosin stain). (D) Immunohistochemistry shows the stroma is positive for estrogen receptor, seen as nuclear (brown) positivity. This is consistent with the diagnosis of mucinous cystic neoplasm (MCN).

This is the first case described in the literature of a fistula between a pancreatic MCN and the portal venous system causing mucin to extend into the vein, a rare event that has been documented in other types of pancreatic mucinous tumors, such as IPMN. Awareness of this disease could improve prospective imaging diagnosis that would guide medical treatment and surgical planning.

Discussion

The differential imaging diagnosis for his mass includes a mucinous cystic neoplasm (MCN), side branch IPMN, macrocystic serous cystadenoma (SCA), and walled-off pancreatic necrosis. The lack of prior history of pancreatitis and unremarkable laboratory makes pancreatic walled-off necrosis less likely. An IPMN could demonstrate similar imaging characteristics, but given the large size of this lesion, there would typically be evidence of communication with the main pancreatic duct. A macrocystic serous cystadenoma (SCA) is rare, with a higher prevalence in older patients and is typically asymptomatic. On imaging, it is commonly located in the pancreatic head, demonstrates a lobulated contour, and has an absence of wall enhancement. None of these features were present in this case.

A unilocular cystic lesion with fluid density on CT and a thick nodular wall without clear communication with the main pancreatic duct are all features favoring a Mucinous Cystic Neoplasm (MCN) of the pancreas. The female gender of the patient also supports this diagnosis. The progressive expanding material within the portal venous system, its cystic nature on imaging and the lack of response to anticoagulation should raise the concern for fistulation and

extension of mucin into the adjacent vascular structures of the adjacent pancreatic mucinous tumor.

Bland thrombus attenuation on CT has been previously studied. In a comparison study [7] to distinguish the difference between tumor and bland thrombus on the portal vein, a value of greater than 54 HU in a thrombus ROI had a 92.1% sensitivity and 85.2% specificity for the diagnosis of tumour thrombus, suggesting while this is much higher than the values measured in our case (filling defect had HU of 5 in the portal and mesenteric vein), this was proven to represent tumour extension, exemplifying that mucin can be a pitfall in the diagnosis of tumour thrombus.

This is a rare event that has been documented in other types of pancreatic mucinous tumors, specifically IPMN [3-6]. Awareness of this condition could improve prospective imaging diagnosis, and therefore guide medical treatment and surgical planning.

Conclusion

This case represents the first documented instance of a fistula between a pancreatic Mucinous Cystic Neoplasm (MCN) and the portal venous system. While MCNs are well recognized in clinical practice, this unusual complication, previously described in other pancreatic mucinous tumors like Intraductal Papillary Mucinous Neoplasms (IPMN), underscores the need for heightened awareness in imaging diagnosis. The clinical presentation, progression of venous thrombus despite anticoagulation, and fluoroscopic findings all pointed toward an underlying communication between the MCN and the venous system. Early identification of such an anomaly is essential for guiding appropriate medical management, including surgical intervention. The case highlights the critical role of detailed imaging evaluation and interdisciplinary collaboration in the diagnosis and treatment planning for complex pancreatic lesions.

Declarations

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Ethics Approval

Not applicable.

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