

# Ruptured Mucinous Cystadenoma Pancreas: A Case Report and Review of Literature

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## ABSTRACT

Mucinous cystic neoplasm (MCN) of the pancreas is a rare neoplasm affecting the elderly women. They vary in their clinical presentation and biological behavior. Spontaneous rupture of MCN is very rare and only eight cases have been reported so far in the English literature. We report a case of a young woman presenting with abdominal pain following spontaneous contained rupture of MCN managed with surgical resection.

**Keywords:** Cystic neoplasm of pancreas, Distal pancreatectomy, Mucinous cystadenoma, Pancreas, Rupture.

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## INTRODUCTION

Mucinous cystic neoplasm (MCN) of the pancreas is a rare neoplasm affecting the elderly women.<sup>1</sup> They encompass a spectrum of biological behavior from benign cystadenoma to malignant cystadenocarcinomas. They are usually detected incidentally or during evaluation of the vague abdominal symptoms. Spontaneous rupture of MCN is very rare and only eight cases have been reported so far in the English literature. We report a case of a young woman presenting with abdominal pain following spontaneous contained rupture of MCN managed with surgical resection.

## CASE DESCRIPTION

A 39-year-old woman presented to the surgical outpatient department with complaints of diffuse abdominal pain for 2 months. There was no other contributory history. Clinical examination was unremarkable except for fullness in the left hypochondrium. Contrast-enhanced computed tomography showed a large peripherally enhancing collection measuring 9.2 × 3.1 cm posteroinferior to proximal body of the stomach. Another peripherally enhancing collection measuring 4.7 × 3.8 cm was noted posteroinferior to the antropyloric region of stomach communicating with the previous collection. The collections were in close relation to the body and the tail of pancreas. Wall of the collection measured 2 to 3 mm in thickness with mild enhancement suggesting mild peritonitis. Endoscopic ultrasound confirmed the collection with aspiration showing mucoid material with no atypical cells on cytology. Fluid analysis showed elevated amylase (1,52,400 IU/L) and lipase (2,10,400 IU/L) with normal carcinoembryonic antigen (CEA) (3.59 ng/mL) and CA19-9 (87.27 U/mL). Magnetic resonance cholangiopancreatography (MRCP) confirmed the suspicion of pancreatic duct communication with an associated proximal duct dilatation (Fig. 1).

In view of mucinous aspirate and pancreatic ductal communication, distal pancreatectomy was planned. Intraoperatively, serous fluid of 300 mL in the lesser sac was found tracking along the superior border of the pancreas from body to tail and abutting the transverse mesocolon (Fig. 2). Pancreatic duct disruption was noted at the level of body with mucinous material around the duct (Fig. 2). Distal pancreatectomy with splenectomy

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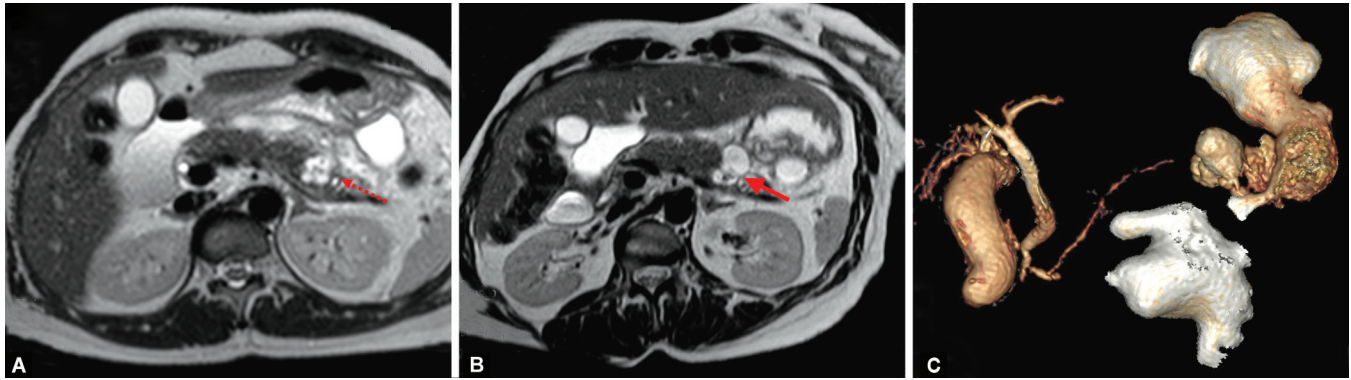
**Conflict of interest:** None

was done. The patient had an uneventful postoperative period. Histopathological examination was consistent with the ruptured benign epithelial cystic lesion with ovarian type stroma arising from distal pancreas suggestive of the mucinous cystadenoma (Fig. 3). Twelve regional lymph nodes with reactive changes were also noted. Patient is doing fine at 6 months of follow-up.

## DISCUSSION

MCN of the pancreas represent 1% of pancreatic neoplasms affecting predominately women during the fifth and sixth decades.<sup>2</sup> The tumor commonly affects the body and tail of the pancreas.<sup>3</sup> De novo mucinous cystadenocarcinoma and malignant transformation of cystadenoma have been described.<sup>2</sup> Given their insidious growth and predisposition for affecting the body and tail of the pancreas, patients are often asymptomatic or minimally affected.<sup>4</sup> Significant symptoms such as jaundice, back pain, and weight loss may indicate invasive cystadenocarcinoma.<sup>4</sup>

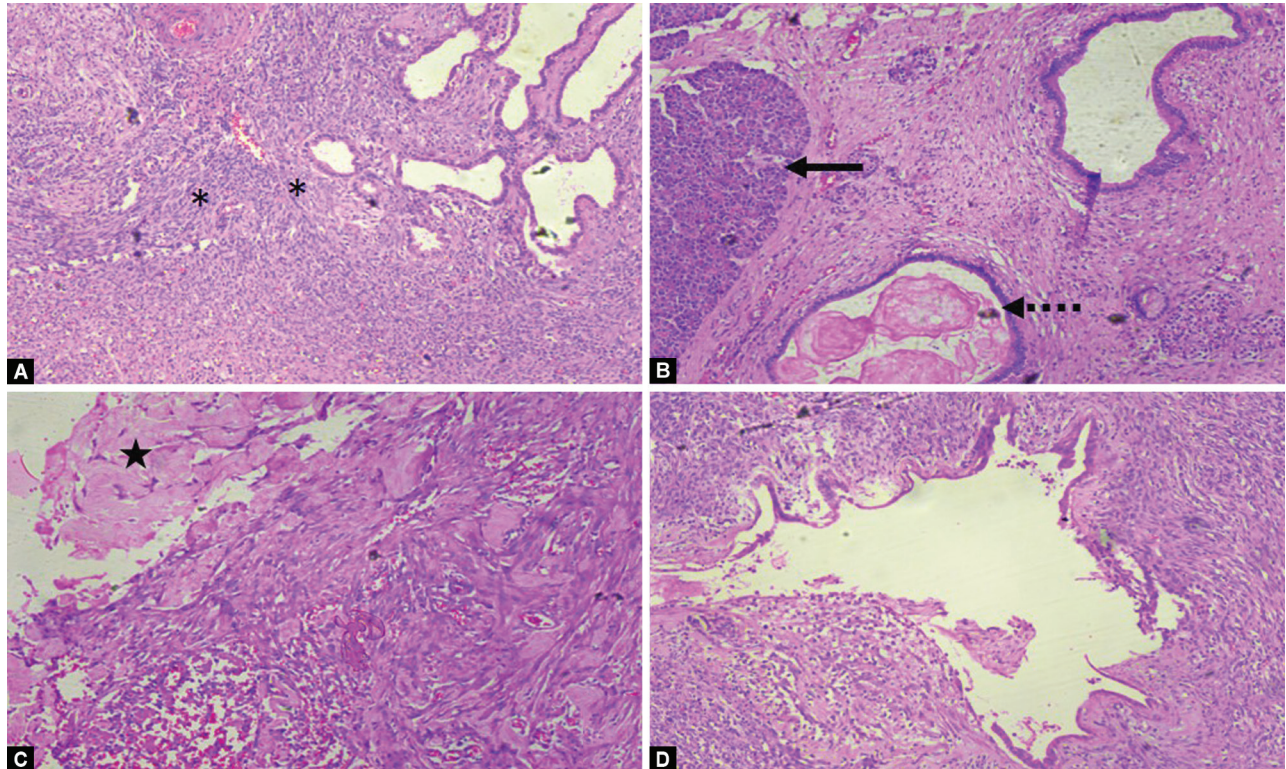
Apart from malignant transformation, MCNs have very few complications. Spontaneous rupture is one such complication reported. Eight cases of spontaneous rupture of MCN have been reported in the English literature. Table 1 depicts the summary of these cases. The initial case reports were in pregnant women, suggesting a possible stimulation of estrogen or progesterone receptors found on the neoplasms by hormonal changes during pregnancy leading to a rapid increase in size and rupture. However, similar ruptures have been found in nonpregnant women and men. It is also interesting that the histology of the ruptured tumors



**Figs. 1A to C:** Magnetic resonance imaging. (A) Axial T2-weighted image showing T2 hyperintense lesion in the pancreatic tail (\*) with proximal ductal dilatation (dotted red arrow). (B) Reformatted T2-weighted image showing the communication between the lesion and the collection (solid red arrow). (C) Reconstructed volume-rendered image showing the dilated and the relation of the collection



**Figs. 2A to C:** Intraoperative image. (A) Collection bulging through the transverse colon (\*). (B) After complete mobilization of the pancreatic tail and the spleen. (C) Opening of the fistula between the lesion and the pancreatic duct (solid black arrow)



**Figs. 3A to D:** Photomicrograph of H&E of resected specimen. (A) High-power field (40x) showing presence of ovarian stroma (\*). (B) High-power field (40x) showing the presence of adjacent pancreatic acinar tissue (black solid arrow) and dilated pancreatic duct with inspissated mucinous material (dotted black arrow). (C) High-power field (100x) showing corpora albicans. (D) High-power field (40x) showing dilated pancreatic duct

**Table 1:** Summary of case reports of ruptured mucinous cystic neoplasm

Author	Year	Age	Gender	Presentation	Imaging	Surgery	Final HPR	Follow-up
Smithers et al. <sup>5</sup>	1996	33	Female	Acute left iliac fossa pain radiating to left shoulder	—	Distal pancreatectomy + splenectomy after cystogastrostomy	Mucinous cystadenocarcinoma	—
Ozden et al. <sup>2</sup>	2007	32	Pregnant female	Acute upper abdominal pain	—	Cystectomy with spleen preservation	Well-differentiated mucinous cystadenocarcinoma	Gemcitabine for 6 months. DFS 12 months
Bergensfeldt et al. <sup>4</sup>	2007	42	Female	Vague abdominal pain with increasing abdominal distention	USG—massive ascites with large cystic lesion in body of pancreas	Distal pancreatectomy with splenectomy with colectomy	Borderline mucinous cystic neoplasm	DFS—19 months
Biswas et al. <sup>6</sup>	2007	59	Female	Acute abdominal pain with loss of consciousness	Hemoperitoneum and a large cystic lesion involving the body and tail of the pancreas	Distal pancreatectomy with splenectomy	Mucinous cystadenoma	—
Naganuma et al. <sup>7</sup>	2011	32	Pregnant female	Acute abdomen	Rapidly growing mucinous cystic neoplasm in pancreatic head	Emergency cesarean section with pancreatoduodenectomy	Mucinous cystadenocarcinoma	Four cycles of gemcitabine—local recurrence at 7 months surgically excised. Received additional 16 courses of GEM. DFS—3 years
Imoto et al. <sup>8</sup>	2013	69	Female	Abdominal pain	CT—multilocular cystic lesion of the pancreas with solid components with dilated PD. ERCP showed leakage of contrast medium from the main pancreatic duct into the peritoneal cavity	Distal pancreatectomy with splenectomy	Mucinous cystadenocarcinoma	DFS—2 months
Woo et al. <sup>9</sup>	2016	72	Male	Epigastric pain for 2 weeks followed by acute exacerbation	CT—irregular high attenuation within the cystic lesion with wall defect. Fluid noted in bilateral paracolic gutter and pelvic cavity	Splenic artery ligation and hematoma evacuation	Frozen biopsy of the nodules of peritoneum, mesentery of the small and large bowel—mucinous cystadenocarcinoma	Adjuvant chemotherapy with gemcitabine and erlotinib. Expired after 189 days of operation due to aspiration pneumonia DFS—3 years
Haddad et al. <sup>3</sup>	2018	30	—	Acute epigastric pain	MRI—cystic mass at the expense of the body of the pancreas. T2- and T1-weighted imaging showed hypersignal and hyposignal respectively with regular wall enhanced after gadolinium injection. The anterior component had lobulated contours and contained an enhanced septum. A communication between the posterior cystic wall and the retroperitoneum was observed	Distal pancreatectomy with splenectomy	Pancreatic mucinous cystadenoma with low-grade dysplasia	DFS—3 years
Our case	2018	39	Female	Vague abdominal pain	CT and MRCP—Lesser sac collection with pancreatic duct communication	Distal pancreatectomy with splenectomy	Mucinous cystadenoma	DFS – 3 months

Abbreviations: USG, ultrasonogram; CT, computed tomography; MRI, magnetic resonance imaging; ERCP, endoscopic retrograde cholangiopancreatogram

range from cystadenoma to borderline tumors to even mucinous cystadenocarcinoma. Thus, the exact cause for such a complication is thus difficult to explain. Though the rupture is usually intraperitoneal, even retroperitoneal rupture has been reported.<sup>3</sup>

Our index patient was unique in many ways. While she was younger than the usual age of presentation, the subacute nature of the presentation of the contained rupture was not seen in the other cases reported. The imaging and the fluid analysis such as high amylase and normal CEA were favoring pseudocyst. However, the lack of clinical symptom suggestive of acute or chronic pancreatitis and mucinous aspirate suggested otherwise. Communication with pancreatic duct is known to occur in 7% of mucinous cystadenoma explaining the high amylase value and imaging of ductal communication.<sup>1</sup> The presence of ovarian stroma differentiates it from the intraductal papillary mucinous neoplasm.<sup>10</sup>

Emergency surgical intervention was performed in all these cases except one that was incidentally detected during endoscopic retrograde cholangiopancreatogram.<sup>8</sup> Excision of the tumor with curative intent has been the principle line of management. In a case of bleeding ruptured MCN, only splenic artery ligation was done by Woo et al.<sup>9</sup> in view of biopsy-proven peritoneal disease.

The impact of spontaneous rupture of MCN on overall survival is difficult to ascertain, given the rarity of the complication. Though concerns have been raised about possible tumor spillage during the intervention performed for obtaining tissue or fluid, its impact on survival is not known. We believe that given the good survival shown by other reports, ruptured MCN, especially cystadenoma and borderline tumors, should not be equated to peritoneal involvement, and management should be radical excision with curative intent wherever feasible. In patients with ruptured malignant MCN, adjuvant therapy should be considered to improve overall survival.

## CONCLUSION

Spontaneous rupture of MCN is a very rare complication. Preoperative imaging plays a crucial role in identification and planning surgery. Radical excision with curative intent, especially nonmalignant MCN, should be attempted.

## CLINICAL SIGNIFICANCE

Spontaneous rupture of cystic neoplasm of pancreas is an uncommon complication. Though more commonly reported in

intraductal papillary mucinous neoplasm, clinicians should be aware that the MCNs can also rupture. Moreover, the outcome following rupture depends on the malignant potential of the MCN. Benign and borderline lesions require aggressive surgery to achieve complete cytoreduction and are associated with good overall survival. Malignant lesions have poor outcomes despite adjuvant chemotherapy.

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