

## Case Report

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# Pancreatic mucinous cystadenocarcinoma: Case report of a rare malignancy

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## Abstract

Pancreatic mucinous cystadenocarcinoma (MCAC) is extremely rare predominantly occurs in women between 5th and 6th decades; the majority arises from pre-existing mucinous cystadenoma. Commonly diagnosed incidentally during investigation for often unrelated and nonspecific abdominal complaints using state-of-the-art abdominal imaging. Here we report the case of a 69-year-old woman initially brought to the emergency department with nausea and vomiting. The clinical diagnosis of epigastric mass was made, the blood tests were normal. Abdominal and pelvic CT showed irregular defined multiloculated large cystic lesion in the body and tail of the pancreas with secondary locations. MRI showed fluid signal of the cyst with fleshy portion on intermediate signal, hyperintense signal diffusion (DWI) with restriction of ADC and heterogeneous gadolinium enhancement. Histopathological examination revealed a pancreatic mucinous cystadenocarcinoma. Therefore, the patient referred for neoadjuvant chemotherapy, given the extensive nature of the tumor.

**Keywords:** Mucinous; Cystadenocarcinoma; Pancreas; Imaging.

## Introduction

Mucinous Cystic Neoplasm (MCN) degenerative is an uncommon cystic tumor of the pancreas, accounting for about 10% of pancreatic cystic lesions. It occurs almost exclusively in women (female to male ratio of 20:1), with a mean age of diagnosis at 40-60 years, usually located in the pancreatic body and tail (93%-95%) [1,2]. Small MCNs are usually asymptomatic and found incidentally on imaging during clinical evaluation for

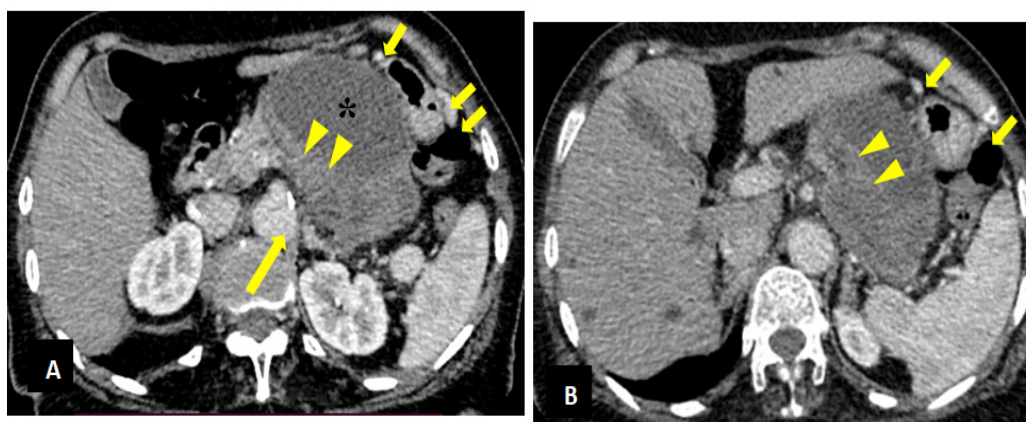
other conditions. Larger MCNs may produce symptoms including abdominal pain, nausea and vomiting, anorexia, and weight loss [3,4]. Imaging studies often show a large cyst, within which thin-walled, daughter cysts can be seen. Calcification may be present in the wall in 20% of cases. The tumor is not connected to the duct system. Features suggestive of malignancy include large size, irregular thickening of the cyst wall, mural nodules, and/or papillary excrescences projecting into the cyst lumen

[4-6]. Correct and early characterization of a premalignant or malignant mucinous cystic neoplasm and subsequent adequate surgical resection, if possible, offers a comparably favorable prognosis. However, once invasive or metastasized, the outcome of a cystic pancreatic carcinoma is merely as poor as for ductal adenocarcinomas of the pancreas [7,8].

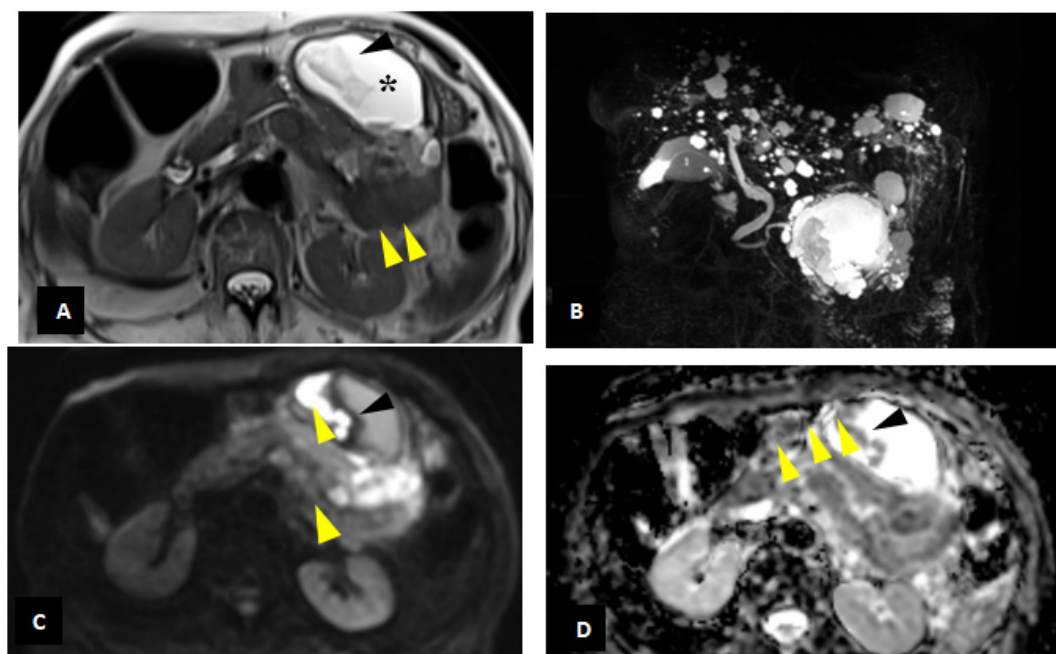
### Case presentation

The patient in the present study was 69 year-old woman, presented with nausea and vomiting. Upon examination, an epigastric mass was found, blood tests were normal. Abdominal ultrasound showed a large epigastric mass. Computed tomography scan of the abdomen showed an irregular defined multiloculated large cystic lesion (9.56 × 6.48 cm) in the body and tail of the pancreas have a limit with the stomach, left liver,

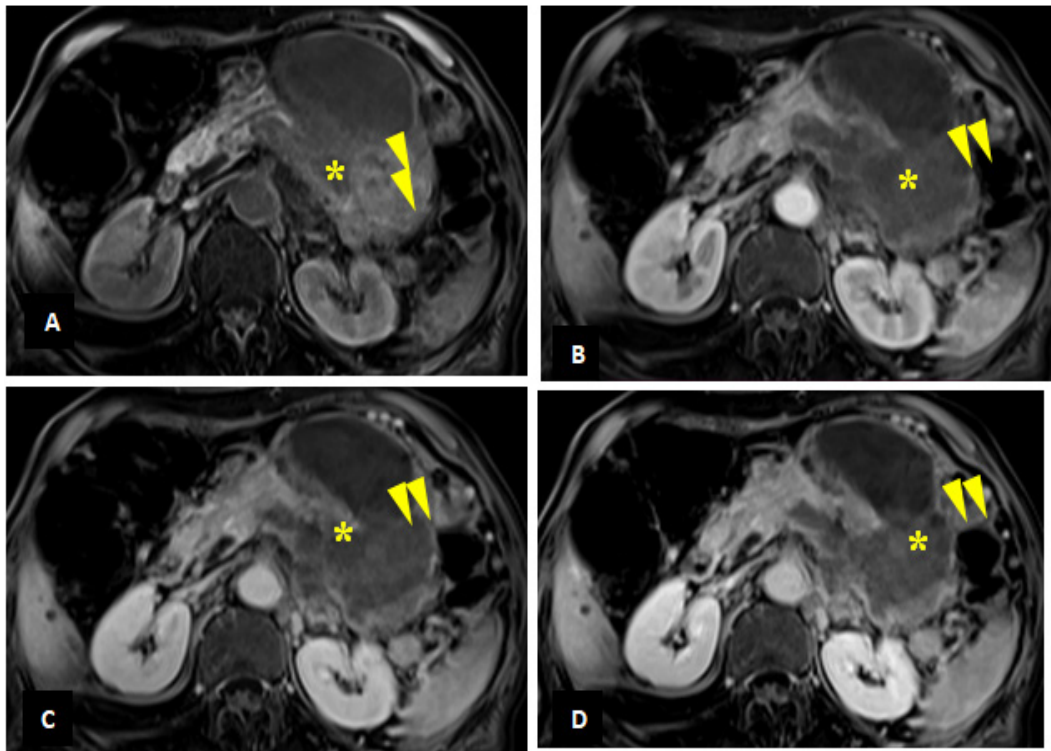
porto-mesenteric axis with splenic vein thrombosis and portal hypertension (Figure 1). MRI showed liquid signal of the cyst on hyperintense signal T2 with solid portion on intermediate signal, hyperintense signal diffusion weighted images (DWI) with restriction of ADC of solid portion. Three-dimensional MR cholangiography with maximum intensity projection reconstruction shows no communication with the pancreatic duct, associated with biliary hamartomas (complexes de Von Meyenburg) (Figure 2). Contrast-enhanced T1-weighted MR images show enhancement of the intervening septa and solid portion on arterial, venous and delayed phase, with non-enhancing cystic spaces (Figure 3). Preoperative diagnosis was a cystic neoplasm of the pancreas associated with invasive carcinoma. Histopathological examination revealed mucinous cystadenocarcinoma of the pancreas. Therefore, the patient referred for neoadjuvant chemotherapy, given the extensive nature of the tumor.



**Figure 1:** Contrast-enhanced CT shows a multiloculated large cystic lesion in the body and tail of the pancreas (A,B). The mass has areas of cystic degeneration (\*) but has solid portion (arrowheads). Splenic vein thrombosis (long arrow) with gastroepiploic collateral channel along the greater curvature of stomach (short arrow).



**Figure 2:** MRI shows liquid signal of the cyst (\*) on hyperintense signal T2 (A) with solid portion (arrowheads) on intermediate signal, hyperintense signal diffusion weighted images (DWI) (C) with restriction of ADC (D) of solid portion. Three-dimensional MR cholangiography with maximum intensity projection reconstruction (B) shows no communication with the pancreatic duct, associated with biliary hamartomas (complexes de Von Meyenburg).



**Figure 3:** Montrast-enhanced T1-weighted MR images show enhancement of the intervening septa (arrowheads) and solid portion (\*) on arterial (B), venous (C) and delayed (D) phase, with non-enhancing cystic spaces.

### Discussion

Pancreatic mucinous cystic neoplasms are rare tumors, accounting for only 2-5% of primary exocrine pancreatic tumors [9]. Mucinous cystic neoplasm is the currently recognized terminology for this tumor, which has been referred to previously as mucinous cystadenoma and mucinous cystadenocarcinoma (non-invasive or invasive) [4]. MCNs are generally considered to progress from low-grade dysplasia through high-grade dysplasia to invasive carcinoma and are one of the three main precursors of pancreatic invasive carcinoma, the other two being PanIN and intraductal papillary neoplasm [4]. They should always be sampled thoroughly to establish the diagnosis and the grade of dysplasia. Diagnosis is based on the presence of ovarian type fibrous stroma and no communication with the pancreatic ductal system. These tumors occur predominantly in women between 5th and 6th decades. Malignant lesions usually present about 10 years later than cystadenoma. Pancreatic mucinous cystadenocarcinoma is extremely rare and the majority arises from preexisting mucinous cystadenoma [10]. Small MCNs are usually asymptomatic and found incidentally on imaging during clinical evaluation for other conditions. Larger MCNs may produce symptoms including abdominal pain, nausea and vomiting, anorexia, and weight loss. Patients may present with a palpable abdominal mass [3,4,11,12]. Upper gastrointestinal bleeds due to involvement of the porto-mesenteric axis with portal hypertension, direct invasion of the gastric wall with ulceration of the gastric mucosa [4]. Once the diagnosis of cystic neoplasm of the pancreas is suspected, further evaluation should be made with abdominal ultrasonography, computed tomography scan and MRI. In either way, a pathological histochemical examination is required to achieve a definitive diagnosis. Presence of ovarian stroma on histology is essential for diagnosis of MCAC. This also suggests a possible tumorigenic pathway for both ovarian and pancreatic MCAC. Correct and early characterization of a premalignant or malignant mucinous cystic neoplasm and subsequent adequate surgical resection,

if possible, offers a comparably favorable prognosis. However, once invasive or metastasized, the outcome of a cystic pancreatic carcinoma is merely as poor as for ductal adenocarcinomas of the pancreas [7,8]. The 5-year survival rate for resected MCN with associated invasive carcinoma is up to 60%, which is much better than for conventional, non-MCN-related pancreatic ductal adenocarcinoma. This probably reflects the earlier stage at diagnosis [4,13].

### Conclusion

Pancreatic MCNs are mucin-producing cystic neoplasms occurring almost exclusively in women, predominantly located in the body and tail of pancreas, and do not communicate with the duct system. Prognosis is excellent for non-invasive MCNs. Given their malignant potential, all MCPNs should be excised in patients fit to undergo surgery.

### Declarations

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