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Spontaneous rupture of a pancreatic acinar cell carcinoma presenting as an acute abdomen

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ABSTRACT

our knowledge, there has been no report on spontaneous rupture of acinar cell carcinoma. *PRESENTATION OF CASE*: A 39-year-old Azari male presented with a history of sudden onset, acute epigastric pain of 12-h duration. Eight hours later the patient's general condition rapidly deteriorated, blood pressure was decreased to 90/70 mm/Hg and heart rate was increased to 120 beat/min. Emergent abdominal computed tomography scan showed a well-defined hypo-dense, necrotic mass, measured 12 cm × 12 cm that was originating from the uncinate process of pancreas with marked free peritoneal fluid and extensive haziness of retroperitoneal and mesenteric fat compatible with marked bleeding.

INTRODUCTION: Pancreatic acinar cell carcinoma is a rare malignant pancreatic neoplasm. To the best of

DISCUSSION: Pancreatic acinar cell carcinoma (ACC) usually presents with abdominal pain, nausea and vomiting. To best of our knowledge, no report has been made of spontaneous rupture of ACC. CONCLUSION: Pancreatic carcinoma may present as acute abdomen due to rupture of underlying neoplasm

Emergent abdominal operation was performed and histopathology revealed acinar cell carcinoma of the

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1. Introduction

Pancreatic acinar cell carcinoma (ACC) is a rare malignant tumor, accounting for approximately 1% of primary pancreatic neoplasms of exocrine origin. 1

Clinical presentation is different from adenocarcinoma. Jaundice is an infrequent finding (12%) and complications related to lipase release (panniculitis) are common.² Abdominal pain is reported to be present in 32% and nausea and vomiting in 23% of patients.³

To the best of our knowledge, this is the first described case of spontaneous rupture of ACC that presented as an acute abdomen.

2. Case presentation

A 39-year-old Azari male was referred to our tertiary referral hospital's emergency department with a history of sudden onset, acute epigastric pain of 12 h duration. The pain was progressive and increased in intensity over the past 12-h period. The pain was accompanied by nausea, vomiting. There was no past medical history of a specific disease or past trauma.

Physical examination revealed abdominal distention with tenderness and rebound tenderness in the epigastric area. The blood

* Corresponding author. Tel.: +98 4413455810; fax: +98 4413455810. E-mail addresses: afshin.mohdi@gmail.com, mohamadi_afshin@yahoo.com (A. Mohammadi). pressure was $110/70 \, \text{mm} \, \text{Hg}$, heart rate was 90 beat/min and body temperature was 38.3° .

Laboratory findings in our hospital were as follows: white blood cell count: 18,000 per ml, hemoglobin: 8.5 g/dl, hematocrit: 25, prothrombin time: 14s. Liver enzyme, alkaline phosphatase, serum amylase were within normal limit.

Eight hours after admission the patient's general condition deteriorated rapidly, blood pressure was decreased to 90 over 70 mm/Hg and heart rate was increased to 120 beat/min.

Emergent abdominal ultrasonography revealed a well-defined hyper-echoic mass ($10\,\mathrm{cm} \times 12\,\mathrm{cm}$) in the epigastrium compatible with head of pancreas and marked free peritoneal fluid. Emergent abdominal computed tomography scan showed a well-defined hypo-dense necrotic mass measuring $12\,\mathrm{cm} \times 12\,\mathrm{cm}$ originating from the uncinate process of pancreas. There was also evidence of neoplastic invasion to the second part of duodenum (thickening of bowel wall), marked free peritoneal fluid and extensive haziness of retroperitoneal and mesenteric fat compatible with marked bleeding (Fig. 1).

Emergency laparotomy was performed. In the operation room, the abdominal cavity was full of gross blood and clot and there was active bleeding from a ruptured mass in the pancreatic head. A large tumor was found originating from uncinate process of pancreas extending into the retroperitoneal space, transverse mesocolon and duodenum. Pancreaticoduodenectomy was not possible to perform and only cyto-reduction (debulking) surgery was performed. There was no signs of distant metastasis or regional lymphadenopathy

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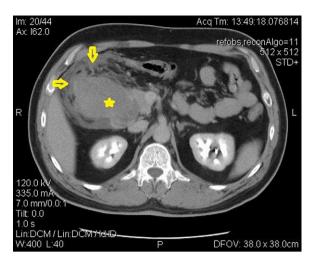


Fig. 1. Abdominal computed tomography scan revealed a hypodense mass originate from pancreatic uncinate process (stellate) accompanied with marked mesenteric haziness (arrow)

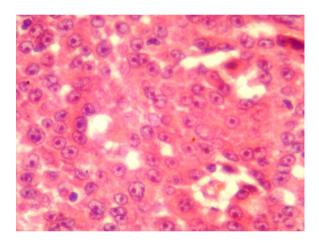


Fig. 2. Histopathology revealed. The neoplastic cells had relatively uniform nuclei, minimal pleomorphism, large central single nuclei and moderate amphophilic to eosinophilic granular apical cytoplasm reflecting the accumulation of zymogen granules.

during laparotomy but there was extensive local spreading and adhesion to omentum. Wound healing was well and he had no postoperative complication.

Histopathology revealed neoplastic cells that were arranged in solid sheets, nests and less organized acini. The neoplastic cells had relatively uniform nuclei, minimal pleomorphism, large central single nuclei and moderate amphophilic to eosinophilic granular apical cytoplasm reflecting the accumulation of zymogen granules (Fig. 2). These findings were consistent with those of ACC.

The patient was treated with chemotherapy for four weeks with a two-week interval as a single cycle. After chemotherapy,

CT scan demonstrated remarkable shrinkage of the residual tumor. The patient is still alive with no distant metastases after 6-month follow-up.

3. Discussion

Spontaneous abdominal hemorrhage is one of the rare causes of acute abdomen. It is defined as non-traumatic and non-iatrogenic intra-abdominal bleeding.⁴ Spontaneous abdominal hemorrhage can have visceral sources. The main visceral sources of spontaneous intra-abdominal bleeding that have been previously reported in the literatures are; underlying hypervascular tumor,⁴ spontaneous rupture of hepatic adenoma or hepatocellular carcinoma,⁵ renal cell carcinoma,⁶ renal angiomyolipoma,⁷ adrenal pheochromocytoma⁸ and gastrointestinal stromal cell tumor.⁹

Pancreatic neoplasms are an extremely rare source of spontaneous bleeding and only few cases have been reported. In review of literature we found only five cases of spontaneous rupture of cysadenoma of pancreas. ^{10–15} Naganuma et al. ¹⁰ reported ruptured mucinous cystic neoplasm with associated invasive carcinoma of pancreatic head in a pregnant woman. Heim et al. ¹¹ reported ruptured cystadenoma of the pancreas in a 85-year-old woman. Ozden et al. ¹² reported rupture of pancreatic mucinous cystadenocarcinoma in a pregnant woman. Kobayashi et al. ¹⁴ reported spontaneous rupture of pancreatic metastasis from renal cell carcinoma in a 53-year-old female patient, which presented with acute abdomen. Table 1 summarizes the previous cases of spontaneous rupture of pancreatic neoplasm.

Acinar cell carcinoma of the pancreas is a rare malignancy that originates from pancreatic exocrine gland.¹ Because of frequent metastasis, a high recurrence rate, and low resectability it has been considered as a cancer with a poor prognosis.

Most of ACC occurs in white elderly men and are asymptomatic. The most frequent presentations are weight loss (50%), abdominal pain (32%), nausea and vomiting (20%), elevated lipase levels (16%) and jaundice being an uncommon presentation. The prognosis of ACC is better than pancreatic ductal carcinomas but worse than pancreatic neuroendocrine tumor. 16

Review of the literature revealed only one somewhat similar case which was rupture of a splenic metastasis from an acinar cell carcinoma. 17

Our case was unusual both in the age and in clinical presentation of ACC. We are presenting a case of spontaneous rupture of ACC for the first time. Our patient like the previously reported cases, presented with an acute abdomen and hypotension as the first sign.

We diagnosed and localized intra-abdominal bleeding due to pancreatic mass preoperatively by computed tomography. Furlan et al.⁴ revealed that CT is essential for prompt diagnosis of intra-abdominal bleeding.

Our study suggests that pancreatic carcinoma may present as an acute abdomen due to rupture of pancreatic neoplasm and physician should bare in mind the pancreatic carcinoma as one of the rare tumors presenting with acute abdomen and abdominal bleeding.

 Table 1

 Table summarized the previous cases of spontaneous pancreatic rupture due to pancreatic neoplasm.

	Author	Age	Sex	Type of tumor
1	Naganuma et al. ¹⁰	32 years old	Pregnant women	Mucinous cystic neoplasm
2	Ozden et al. 12	28 years old	Pregnant women	Mucinous cystadenocarcinoma
3	Heim et al.11	85 years old	Women	Cystadenoma
4	Kobayashi et al. ¹⁴	53 years old	Women	Metastasis from renal cell carcinoma
5	Shimizu et al. ¹⁵	77 years old	Men	Intraductal papillary mucinous neoplasm
Present case	Mohammadi et al. (2012)	39 years old	Men	Acinar cell carcinoma

Conflict of interest

None.

Funding

None.

Consent

We have obtained written consent from the patient and that we can provide this should the editor ask to see.

Author contributions

Afshin Mohammadi: manuscript writing; Jalal Porghasem: perform of surgery; Arefeh Esmaili: histopathology evaluation; Mohammad Ghasemi-rad: editing.

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