Case Report

Spontaneous Rupture of Pancreatic Acinar Cell Carcinoma: Report of a Case

Sadaaki Yamazoe, Ryosuke Amano, Kenjiro Kimura, Go Ohira, Kohei Nishio, Kotaro Miura, Masatsune Shibutani, Katsunobu Sakurai, Hisashi Nagahara, Takahiro Toyokawa, Naoshi Kubo, Hiroaki Tanaka, Kazuya Muguruma, Hiroshi Ohtani, Masakazu Yashiro, Kiyoshi Maeda, Masasichi Ohira, Kosei Hirakawa

Department of Surgical Oncology (First Department of Surgery), Osaka City University Graduate School of Medicine, Osaka, Japan

Introduction: Pancreatic acinar cell carcinoma (ACC) is a relatively rare neoplasm. Furthermore, tumor rupture is extremely rare. Only 1 case of ruptured pancreatic ACC has been reported, and the long-term outcome of the case is unknown. Here, we present a case of spontaneously ruptured ACC with long-term survival after successful resection.

Case Presentation: A 67-year-old man was brought to our hospital by ambulance, presenting with progressive left abdominal pain. Laboratory data showed an increased inflammatory response, and contrast-enhanced computed tomography showed a mass in the pancreas tail with nonuniform enhancement in the early phase. Fluid collection was detected around the spleen to the left kidney. Spontaneous rupture of a pancreatic tumor was strongly suspected. After improvement of his general condition, the patient underwent resection of the pancreas and adjacent organs. The resected tumor was surrounded by organs and adipose tissue, so obvious exposure was not observed in the surgical margins. Pathologically, neither exposure of tumor cells at the surgical margins nor lymph node involvement was detected. The patient has survived 80 months since initial diagnosis without any evidence of recurrence.

Conclusion: Although ruptured pancreatic ACC has the potential for dissemination, surgical resection including adjacent organs remains an option for curative treatment.

Key words: Pancreas – Acinar cell carcinoma – Rupture – Surgical treatment – Long-term survival

Corresponding author: Sadaaki Yamazoe, Department of Surgical Oncology, Osaka City University Graduate School of Medicine, 1-4-3 Asahi-machi, Abeno-ku, Osaka 545-8585, Japan.
Tel.: +81 6 6645 3838; Fax: +81 6 6646 6450; E-mail: zoesada@yahoo.co.jp
Pancreatic acinar cell carcinoma (ACC) is a relatively rare malignant neoplasm, accounting for about 1% of all pancreatic exocrine tumors. Because of this rarity, therapeutic strategies have yet to be established. Surgical resection has been selected for patients showing localized, resectable disease, because of the metastatic potential of this entity and its resistance to anticancer drugs.

Only 1 case of ruptured pancreatic ACC has been reported; however, long-term outcome of the case is unknown. Therefore, selection of the most appropriate first treatment for ruptured pancreatic ACC that may have disseminated is difficult. In this case report, we describe a case of a long-term survivor who underwent radical resection with adjacent organs for ruptured pancreatic ACC. To the best of our knowledge, this is the first case of long-term survival after surgical treatment for ACC that ruptured spontaneously.

Case Report

A 67-year-old man had experienced dull, persistent left abdominal and shoulder pain during a period of a few days. He was brought to our hospital by ambulance after the pain had gradually progressed and increased in intensity.

Physical examination revealed marked abdominal tenderness and rebound tenderness in the epigastric region. The pain showed radiation to the flank from the left back. The patient’s blood pressure was 118/82 mmHg, heart rate was 109 beats per minute, and body temperature was 37.0°C. He had no recent history of abdominal trauma.

Contrast-enhanced computed tomography showed a large (67×51 mm), well-marginated, smooth-edged, oval-shaped mass in the pancreas tail. Hypervascular enhancement of the mass was nonuniform in the early phase and strongly prolonged in the late phase. Recognizing fluid collection around the spleen to the kidney from the left side of the mass, we suspected bleeding into the retroperitoneal space due to rupture of a tumor originating in the pancreas tail (Fig. 1). Laboratory data showed a white blood cell count of 10,600/μL, and a C-reactive protein level of 27.2 mg/dL. Total bilirubin level was mildly elevated, and carbohydrate antigen 19-9 and carcinoembryonic antigen levels were within normal ranges.

After admission, pain was well controlled, vital signs were stable, and anemia did not progress, so we decided to continue careful observation.

Magnetic resonance imaging showed a low-intensity tumor on both T1- and T2-weighted imaging, with small cystic change inside. On the outside of the tumor, a region with low and high intensity mixed in both T1- and T2-weighted images was observed, which was considered to be a mixture of hematoma and tumor tissue (Fig. 2).

Subsequent 18F-fluorodeoxyglucose positron emission tomography demonstrated intense 18F-fluorodeoxyglucose uptake (maximum standardized uptake value, 5.7) in the tumor, but no accumulation in other organs that would suggest metastasis.

Endoscopic ultrasonography revealed a hypoechoic, heterogeneous mass measuring 56 mm and with distinct borders. Blood flow was confirmed in the tumor by Doppler examination. The differential diagnosis of endoscopic ultrasonography included neuroendocrine tumor, acinar cell tumor, solid pseudopapillary neoplasm, atypical pancreatic adenocarcinoma, lymphoma, and metastatic tumor. Endoscopic ultrasonography–guided fine needle aspiration using a transgastric approach was performed for diagnosis histologically and for prediction of dissemination. The fine-needle aspiration specimen revealed tumor cells with eosinophilic cytoplasm proliferate to form fused ducts. On immunohistochemical examination, tumor cells were stained for α-antitrypsin, lipase, and p53. We therefore diagnosed ACC with spontaneous rupture.

These findings suggested a diagnosis of ACC that would be amenable to resection with adjacent organs, if seeding into the abdominal cavity had not occurred.

Laparotomy was performed 47 days after the initial visit. The patient underwent distal pancreatectomy with splenectomy and partial resection of the transverse colon and diaphragm on suspicion of direct invasion to these organs, but no dissemination or malignant cells were detected from intraoperative cytology.

Gross examination of the resected tumor, which measured 65 × 40 mm with expansive growth, revealed a white to yellowish mass with internal focal bleeding and a well-marginated appearance. The tumor was surrounded by organs and adipose tissue, with no obvious exposure in the surgical margins (Fig. 3).

Microscopically, the tumor comprised cells similar to eosinophilic acinar cells, displaying an acinar structure (Fig. 4). Infiltration into surrounding tissue was only observed in the spleen, but fibrous
adhesion to other organs was apparent. Neither exposure of tumor cells at the surgical margins nor lymph node involvement was detected. Immuno-histochemistry showed that tumor cells were positive for lipase and negative for synaptophysin and chromogranin A. These results confirmed the diagnosis of ACC, and R0 resection was considered to have been obtained by surgery with combined resection of the adjacent organs.

Postoperatively, minor leakage at the anastomosis of the transverse colon due to spreading inflammation from the pancreatic fistula was seen, and the patient was discharged from the hospital 50 days after surgery. We have continued to observe the patient without adjuvant chemotherapy, which he had declined. As of the time of writing, he has survived 80 months since initial diagnosis without any evidence of recurrent disease.

Discussion

Pancreatic ACC is a relatively rare pancreatic tumor that arises from the acinar cells of the pancreas. This entity has been reported to comprise less than 1% of
all pancreatic cancers. A report of 672 cases of ACC from the United States made comparisons to pancreatic ductal adenocarcinoma, finding that ACC tended to occur at a younger age, in males, and in the body/tail of the pancreas rather than in the pancreatic head. ACC is more indolent than invasive ductal cancer, with improved long-term survival among both resected and unresected patients. In a report of 115 cases from the Pancreatic Cancer Registry by the Japan Pancreas Society, the 5-year survival rate for patients with resected ACC was 43.9%, representing an improvement compared with cases with unresectable disease, and prolonged survival is expected with resection.

Rupture of pancreatic tumors seldom occurs. Spontaneous rupture in the absence of trauma has only been reported in 19 cases in PubMed (Table 1). The most frequently reported tumor is solid-pseudo-papillary neoplasm, followed by mucinous cystic neoplasm. Spontaneous rupture of ACC has been reported in only 1 case, presenting as acute abdomen, although the long-term prognosis was not described.

Fig. 2  Magnetic resonance imaging showed a low-intensity tumor on both T1- and T2-weighted imaging, with small cystic change inside. On the outside of the tumor, a region with low and high intensity mixed in both T1- and T2-weighted images was observed.

Figs. 3  Cut surface of the resected tumor, which measured $65 \times 40$ mm with expansive growth, reveals a white to yellowish mass with internal foci of bleeding and a well-marginated appearance.
Few reports have discussed the mechanisms underlying the spontaneous rupture of pancreatic tumors. Naganuma et al.⁹ reported a case in which mucinous cystic neoplasm of the pancreas grew rapidly during pregnancy and ruptured in the late stage of gestation. He suggested that pregnancy in that case may have increased tumor activity in a hormonally dependent manner, to elicit tumor rupture.⁹ Kobayashi et al.¹⁰ reported a rupture of pancreatic metastasis from renal cell carcinoma, potentially caused by intratumoral hemorrhage. They suggested that intratumoral hemorrhage might have occurred because of high blood pressure. Hypertension may elicit rupture of intratumoral vessels by overflow. In the present case, the tumor was hypervascular and showed expans-

Table 1  Previously reported cases: 19 cases were identified from PubMed

| Case | Author         | Type of tumor                        | Citation                          |
|------|----------------|-------------------------------------|-----------------------------------|
| 1    | Takamatsu S    | SPN                                 | Case Rep Med. 2013                |
| 2    | Honda S et al  | Pancreatoblastoma                   | J Pediatr Surg. 2013              |
| 3    | Imoto A et al  | Mucinous cystadenocarcinoma         | Endoscopy. 2013                   |
| 4    | Huang SC et al | SPN                                 | Obstet Gynecol. 2013              |
| 5    | Mohammadi A et al | ACC                             | Int J Surg Case Rep. 2012       |
| 6    | Kawaguchi K et al | Lymphangioma                  | Case Rep Gastroenterol. 2011     |
| 7    | Naganuma S et al | Mucinous cystadenocarcinoma       | Pathol Int. 2011                  |
| 8    | Bergenfeldt M et al | MCN                        | Acta Oncol. 2008                   |
| 9    | Lee SE et al   | IPMC                                | World J Gastroenterol. 2007       |
| 10   | Ozden S et al  | Mucinous cystadenocarcinoma         | Pancreas. 2007                    |
| 11   | Murai H et al  | Anaplastic mucinous cystadenocarcinoma | Nihon Ronen Igakkai Zasshi. 2006 |
| 12   | Omori H et al  | SPN                                 | Hepatogastroenterology. 2005      |
| 13   | Kobayashi A et al | Metastasis from renal cell carcinoma | Jpn J Clin Oncol. 2004          |
| 14   | Ideguchi K et al | Islet cell carcinoma                | J Hepatobiliary Pancreat Surg. 2001 |
| 15   | Panieri E et al | SPN                                 | J Am Coll Surg. 1998             |
| 16   | Heim D et al   | Cystadenoma                         | Chirurg. 1994                     |
| 17   | Jeng LB et al  | SPN                                 | Arch Surg. 1993                   |
| 18   | Errougani A et al | Mucinous cystadenoma              | J Chir (Paris). 1992              |
| 19   | Kawamura T et al | Cystadenocarcinoma                 | Nihon Rinsho. 1980                |

IPMC, intraductal papillary mucinous carcinoma; MCN, mucinous cystic neoplasm; SPN, solid-pseudopapillary neoplasm.

Fig. 4  Resected specimen reveals tumor cells proliferating to form like fusion glands. Immunohistochemical staining shows tumor cells stain positively for α-antitrypsin, lipase, and p53.
sive growth, and the patient had been under treatment for hypertension, suggesting that high blood pressure may have increased the internal pressure of the growing tumor by overflowing from the supplying vessels, potentially contributing to spontaneous rupture.

We did not perform the cytology from fluid collection; however, it is important to confirm the existence of tumor cells in the fluid collection for predicting the peritoneal dissemination. Although we were able to achieve R0 resection, strict follow-up is needed in cases of ruptured tumor because of the possibility of recurrence, particularly peritoneal dissemination. In this case, the patient declined to undergo adjuvant chemotherapy, and treatment regimens have yet to be established for this pathology.

We have reported an extremely rare case of spontaneously ruptured ACC with long-term survival after successful resection by distal pancreatectomy combined with resection of adjacent organs. Although ruptured pancreatic ACC has the potential for dissemination, a surgical approach with en bloc resection is important and may represent an effective treatment option.

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