Single Case

Acinar Cell Cystadenocarcinoma of the Pancreas

Keita Aoto^a Tatsuo Shimura^a Yasuhide Kofunato^a Ryo Okada^a
Rei Yashima^a Yuichiro Kiko^b Seiichi Takenoshita^a

^aDepartment of Organ Regulatory Surgery, Fukushima Medical University, Fukushima City, Japan; ^bDepartment of Pathology, Fukushima Medical University, Fukushima City, Japan

Keywords

Acinar cell cystadenocarcinoma · Pancreatic cancer · Alpha 1-antichymotrypsin · Alpha 1-trypsin

Abstract

Acinar cell cystadenocarcinoma is a rare malignant epithelial neoplasm of the pancreas with a diffusely cystic, gross architecture in which the cysts are lined with neoplastic epithelial cells that demonstrate evidence of pancreatic exocrine enzyme production. This is the 10th case that has been reported in the literature. A 77-year-old male complaining of left hypochondrial pain was referred to our hospital for treatment of a pancreatic tumor. A huge, honeycomb-structured tumor was detected in the pancreatic tail. Distal pancreatectomy with total resection of the residual stomach and partial resection of the transverse colon were performed. Microscopically, there were variably sized cystic lesions in the tumor. Immunohistochemical examinations revealed that tumor cells were positive for alpha 1-antichymotrypsin and alpha 1-trypsin, showing that tumor cells had features of pancreatic acinar cells. Thus, the tumor was diagnosed as acinar cell cystadenocarcinoma. Herein, we report a rare case with acinar cell cystadenocarcinoma, which is the 10th case reported in the literature based on a PubMed search. We managed to resect the tumor completely by distal pancreatectomy with

total resection of the residual stomach and partial resection of the transverse colon. The patient is still alive 26 months after surgery without any recurrence after 1 year of adjuvant chemotherapy with S-1.

© 2016 The Author(s)
Published by S. Karger AG, Basel

Introduction

Acinar cells make up the bulk of the pancreas, whereas pancreatic neoplasms exhibiting predominantly acinar differentiation are relatively uncommon. Almost all acinar neoplasms of the pancreas are malignant. The most common type (more than 90%) of carcinoma of the pancreas is infiltrating ductal adenocarcinoma. Acinar cell carcinoma accounts for 1–2% of pancreatic neoplasms in adults and 15% of pediatric neoplasms, and most patients are adults [1]. Acinar cell cystadenocarcinoma is a rare malignant epithelial neoplasm of the pancreas with a diffusely cystic, gross architecture in which the cysts are lined with neoplastic epithelial cells that demonstrate evidence of pancreatic exocrine enzyme production [1], and only 9 cases have been reported so far [2–10]. The cystic structure, which is separated by a thin fibrous septum, looks sponge-like. The clinicopathological features of pancreatic acinar cell cystadenocarcinoma remain to be elucidated, and therapeutic strategies for the disease have not yet been developed. Herein, we report a case with acinar cystadenocarcinoma of the pancreas complaining of abdominal pain.

Case Presentation

A 77-year-old male complaining of left hypochondrial pain was referred to our hospital for treatment of a pancreatic tumor. His medical history demonstrated distal gastrectomy with Billroth type II reconstruction for a gastric carcinoma 30 years ago (poorly differentiated adenocarcinoma, submucosal infiltration, n0, and stage I). Carbohydrate antigen 19-9 was elevated to 582.0 U/ml, but there were no other abnormalities in blood chemical analyses. Abdominal computed tomography revealed a huge tumor (123 × 104 × 86 mm in size), compressing the surrounding organs, in the pancreatic tail (Fig. 1a). A magnetic resonance imaging enhanced with gadolinium revealed a well-margined, honeycomb-structured tumor with a high-intensity area in the tumor on T2-weighted image, which was estimated to be a fluid collection (Fig. 1b). Abnormal accumulations with a maximum standardized uptake value of 7.1 were detected in ¹⁸F-fluorodeoxyglucose-positron emission computed tomography (Fig. 1c).

Distal pancreatectomy with total resection of the residual stomach and partial resection of the transverse colon were performed and reconstructed in a Roux-en-Y fashion. The resected specimen is shown in Figure 2. A cut surface of the tumor looked like a sponge. Pathological findings showed variably sized cystic lesions containing eosinophilic substances, lined with flat or cubic eosinophilic cells with granules in the cytosol (Fig. 3a, b). Immunohistochemical analyses revealed that tumor cells were positive for alpha 1-antichymotrypsin (Fig. 3c) and alpha 1-trypsin, showing that tumor cells had features of pancreatic acinar cells. The tumor invaded into the spleen without lymph node metastasis. The tumor was diag-

nosed as acinar cell cystadenocarcinoma, with pT3, pN0, cM0, and final stage III using the TNM classification of the Union for International Cancer Control, 7th edition.

The patient was discharged on the 9th day after surgery with an uneventful course. We prescribed S-1 (80 mg/body) as adjuvant chemotherapy for 1 year. He is still alive at 26 months after surgery without recurrent disease.

Discussion

To the best of our knowledge, this is the 10th case of acinar cell cystadenocarcinoma reported in the literature (7 male cases and 3 female cases) as shown in [Table 1](#). The average age of the patients was 53.2 years (range: 4–77), and the mean tumor size was 17 cm (3–39) at the time of diagnosis. Six cases were complaining of abdominal pain. The involved portion of the pancreas was the head in 3 cases, the body in 1 case, the tail in 2 cases, and both the body and the tail in 3 cases. Five cases developed hepatic metastasis, whereas 2 cases suffered from peritoneal dissemination after surgery. Three cases died 2, 13, and 37 months after surgery, respectively. Six patients were alive at the time of the report, with a mean survival time of 14.3 months. Acinar cell cystadenoma/adenocarcinoma should be distinguished from serous cystadenoma and neuroendocrine tumor. It is requisite for a diagnosis of acinar cell cystadenocarcinoma that tumor cells contain pancreatic exocrine enzymes without markers showing a differentiation into neurons in immunohistochemistry. Microscopic features of acinar cell cystadenoma/adenocarcinoma comprise acinar-like formation and eosinophilic cytosol without abundant glycogen. Other cystic tumors of the pancreas are usually excluded by absence of mucinous epithelium. Our case was confirmed to be acinar cell cystadenocarcinoma by immunohistochemical analyses for alpha 1-antichymotrypsin and alpha 1-trypsin which are pancreatic exocrine enzymes. The number of cases with acinar cell cystadenocarcinoma reported so far is so small that a standard therapeutic strategy has not yet been established. However, surgical removal is the only way to cure the disease.

Conclusions

We reported a rare case with acinar cell cystadenocarcinoma, which is the 10th case reported in the literature based on a PubMed search. We managed to resect the tumor completely by distal pancreatectomy with total resection of the residual stomach and partial resection of the transverse colon. The patient is still alive at 26 months after surgery without any recurrence after 1 year of adjuvant chemotherapy with S-1.

Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent form is available for review from the Editor-in-Chief of this journal.

Disclosure Statement

The authors declare that they have no competing interests.

Author Contributions

K.A., T.S., and S.T. conceptualized and designed the study. R.O., R.Y., and Y.K. participated in data acquisition. K.A., T.S., and S.T. performed the analysis and interpretation of the data. T.S. and S.T. helped draft the manuscript. All authors read and approved the final manuscript.

References

- 1 Hruban RH, Pitman MB, Klimstra DS: Tumors of the Pancreas. Atlas of Tumor Pathology, 4th series, fascicle 6. Washington, DC, Armed Forces Institute of Pathology, 2007, pp 191–210.
- 2 Cantrell BB, Cubilla AL, Erlandson RA, Fortner J, Fitzgerald PJ: Acinar cell cystadenocarcinoma of human pancreas. *Cancer* 1981;47:410–416.
- 3 Stamm B, Burger H, Hollinger A: Acinar cell cystadenocarcinoma of the pancreas. *Cancer* 1987;60:2542–2547.
- 4 Hoorens A, Lemoine NR, McLellan E, Morohoshi T, Kamisawa T, Heitz PU, Stamm B, Rüschoff J, Wiedenmann B, Koppel G: Pancreatic acinar cell carcinoma. An analysis of cell lineage markers, p53 expression, and Ki-ras mutation. *Am J Pathol* 1993;143:685–698.
- 5 Ishizaki A, Koito K, Namieno T, Nagakawa T, Murashima Y, Suga T: Acinar cell carcinoma of the pancreas: a rare case of an alpha-fetoprotein-producing cystic tumor. *Eur J Radiol* 1995;21:58–60.
- 6 Joubert M, Fiche M, Hamy A, Heymann MF, Sagan C, Valette PJ, Le Bodic MF: Extension of an acinar cell pancreatic carcinoma with cystic changes invading the Wirsung canal. *Gastroenterol Clin Biol* 1998;22:465–468.
- 7 Colombo P, Arizzi C, Roncalli M: Acinar cell cystadenocarcinoma of the pancreas: report of rare case and review of the literature. *Hum Pathol* 2004;35:1568–1571.
- 8 Beltraminelli HS, Buechner SA, Hausermann P: Pancreatic panniculitis in a patient with an acinar cell cystadenocarcinoma of the pancreas. *Dermatology* 2004;208:265–267.
- 9 Huang Y, Cao YF, Lin JL, Gao F, Li F: Acinar cell cystadenocarcinoma of the pancreas in a 4-year-old child. *Pancreas* 2006;33:311–312.
- 10 Perrone VG, Mariniello DM, De Lio N, Caniglia F, Cappelli C, Campani D, Funel N, Amorese G, Boggi U: The odd case of a small and mucinous-like acinar cell cystoadenocarcinoma of the pancreas. *Pancreatol* 2012;12:421–422.
- 11 Klimstra DS, Hruban RH, Koppel G, Morohoshi T, Ohike N: Acinar cell neoplasms of the pancreas; in Bosman FT, Carneiro F, Hruban RH, Theise ND (eds): WHO Classification of Tumours of the Digestive System. Lyon, IARC Press, 2010, pp 314–318.

Fig. 1. **a** Computed tomography with enhancement. A huge, mixed-density tumor in the tail of the pancreas compressing the surrounding organs (arrows). The tumor invaded into the spleen. **b** T2-weighted magnetic resonance imaging. A solid and cystic component was shown on magnetic resonance imaging with a honeycomb structure (arrows). **c** ^{18}F -fluorodeoxyglucose-positron emission tomography. Abnormal accumulation of the radioisotope was detected in the tumor with the maximum standardized uptake value of 7.1 (arrows).

Fig. 2. Resected specimen. **a** The tumor invaded into the spleen. **b** A cut surface of the tumor resembling a sponge.

Fig. 3. Microscopic findings. **a** Variably sized cystic lesions in the tumor. **b** The tumor cells formed an acinar-like structure, containing eosinophilic substances at the center of the tubular formation. **c** Alpha-1 antichymotrypsin was positive in the tumor cells.

Table 1. Summary of the reported cases

Case	Age, years	Sex	Size, cm	Location	Symptoms	Operation	Metastasis	Outcome	Authors [Ref.]
1	64	male	39	body and tail	epigastric pain, abdominal mass	DP	liver, omentum	dead (13 months)	Cantrell et al. [2]
2	42	male	25	body and tail	weight loss, abdominal mass	DP, PH	liver	alive (18 months)	Stamm et al. [3]
3	64	male	17	body	NA	none	none	NA	Hoorens et al. [4]
4	57	male	3	tail	epigastric pain, vomiting	DP, PH	liver	dead (37 months)	Ishizaki et al. [5]
5	32	female	13	head	abdominal pain	none	liver	alive (13 months)	Joubert et al. [6]
6	69	male	25	body and tail	abdominal pain	EL	peritoneum	alive (6 months)	Colombo et al. [7]
7	60	male	NA	NA	painful erythematous nodules	none	liver	dead (2 months)	Beltraminelli et al. [8]
8	4	female	18	head	none	PPPD	none	alive (12 months)	Huang et al. [9]
9	63	female	3	head	none	PD	none	alive (12 months)	Perrone et al. [10]
10	77	male	12	tail	abdominal pain, abdominal mass	DP	none	alive (20 months)	our case

DP, distal pancreatectomy; PH, partial hepatectomy; NA, not assessed; EL, exploratory laparotomy; PPPD, pylorus-preserving pancreaticoduodenectomy; PD, pancreaticoduodenectomy.