

You cannot miss it: Pancreatic mucoepidermoid carcinoma

A case report and literature review

Hai-Jie Hu, MD, Rong-Xing Zhou, MD, Fei Liu, MD, Jun-Ke Wang, MD, Fu-Yu Li, MD, PhD*

Abstract

Rationale: Pancreatic mucoepidermoid carcinoma (MEC) is a rare disease with no more than 10 cases reported in literature. The prognosis is poor and few patients can survival more than 1 year.

Patient concerns: We presented a case of patient manifested as left upper abdominal pain. Computed tomography demonstrated a low-density shadow measuring 2.1 × 2.4 cm situated at the transition area of neck and body of the pancreas with obscure boundary and irregular enhancement. The preoperative symptoms and imaging features were unspecific.

Diagnoses: Pancreatic MEC.

Interventions: Curative surgery of distal pancreatectomy was conducted. Postoperatively, the patient subsequently underwent 8 cycles of chemotherapy using cisplatin (25 mg/m²) on day 1 to day 3; and gemcitabine (1000 mg/m²) on day 1 and day 8, repeated every 21 days.

Outcomes: The patient was monitored on a regular basis at our outpatient department and survived 23 months after surgery.

Lessons: Preoperative diagnosis of pancreatic MEC is difficult. Patients with pancreatic MEC may have a survival benefit from the multimodal treatment of curative surgery combined with chemotherapy.

Abbreviation: MEC = mucoepidermoid carcinoma.

Keywords: mucoepidermoid carcinoma, multimodal treatment, pancreas, poor prognosis

1. Introduction

Salivary gland carcinoma is a rare malignant tumor, accounting for approximately 5% to 6% of all head and neck cancers.^[1] The salivary gland carcinoma mainly consists of myoepithelial

Editor: N/A.

Hai-Jie Hu and Rong-Xing Zhou contributed equally to this manuscript; Hai-Jie Hu and Rong-Xing Zhou collected the clinical data and drafted the manuscript; Fei Liu and Jun-Ke Wang collected the clinical data; Fu-Yu Li revised the manuscript. All authors read and approved the final manuscript.

Patient's informed consent could not be obtained because the patient died due to progression of the disease before we wrote the case report.

This article is a case report and does not contain any studies with human participants or animals performed by any of the authors.

The financial support from the National Nature Science of China (30801111) and the Science and Technology Support Project of Sichuan Province (No 2014SZ0002-10).

The authors have no conflicts of interest to disclose.

Department of Biliary Surgery, West China Hospital of Sichuan University, Chengdu, Sichuan, China.

* Correspondence: Fu-Yu Li, Department of Biliary Surgery, West China Hospital of Sichuan University, Chengdu, Sichuan 610041, China (e-mail: lfy_74@hotmail.com).

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Medicine (2018) 97:11(e9990)

Received: 22 December 2017 / Received in final form: 30 January 2018 /

Accepted: 31 January 2018

<http://dx.doi.org/10.1097/MD.0000000000009990>

neoplasm, mucoepidermoid carcinoma (MEC), salivary duct carcinoma, and polymorphous low-grade adenocarcinoma, among which, MEC is the most common histological subtype, representing 30% to 40% of the salivary gland malignancies.^[2,3] MEC is generally characterized as an indolent tumor; however, it always has a dismal prognosis.^[1] MEC is characterized by 3 kinds of cells: epidermoid cells, mucin-producing cells, and intermediate cells just between basal cells and epidermoid cells.^[4,5] The etiology and pathogenesis of MEC is unclear. Pancreatic MEC is an even uncommon entity with no more than 10 cases reported in the literature. Due to the heterogeneous clinical features and nonspecific radiological characteristics, pancreatic MEC cannot be easily differentiated from other pancreatic malignancy merely by the preoperative examinations. Operative resection seems to be the most effective option for this malignancy if the mass is resectable. However, the prognosis of pancreatic MEC is extremely poor; few patients can survive more than 12 months even after curative surgery.^[6] Herein, we presented a usual case of pancreatic MEC and conducted a brief literature review of reported series of patients with pancreatic MEC so as to better understand the clinical feature and treatment option of this disease.

2. Case presentation

A 48-year-old man suffered from a 2-month history of left upper abdominal pain. He denied any history of gastrointestinal illness or immunological disease previously. Physical examination was unremarkable. When presented in our department, the patient had a carbohydrate antigen 19-9 level of 300.50 U/mL (normal: <37 U/mL) and a ferritin level of 794.10 ng/mL (normal: 24–336



Figure 1. Computed tomography demonstrated a low-density shadow measuring 2.1×2.4 cm situated at the transition area of neck and body of the pancreas with obscure boundary and irregular enhancement.

ng/mL). The routine blood, liver function, and the inflammatory markers were all within the normal range. Abdominal ultrasonography revealed an equal echo mass ($4.0 \times 2.3 \times 2.6$ cm) in the conjunction of body and tail of the pancreas. Computed tomography demonstrated a low-density shadow measuring 2.1×2.4 cm situated at the transition area of neck and body of the pancreas with obscure boundary and irregular enhancement (Fig. 1). Thus, primary pancreatic carcinoma was suspected and curative surgery was planned after multidisciplinary team discussion. The patient underwent distal pancreatectomy. The tumor appeared as gray–yellow, firm, and medium texture. Microscopically, the tumor consisted of epidermoid cells, intermediate undifferentiated cells, and mucous cells as confirmed by hematoxylin–eosin staining (Fig. 2A). Immunohistochemical staining indicated the tumor was positivity for PCK, CK 7, CK 19, P 63 (Fig. 2B), CK 5/6 and showed negative staining for CgA, Syn, and CK 20. Moreover, it was also positivity for AB and PAS (Fig. 2C). These pathological findings were consistent with the diagnosis of pancreatic MEC. The postoperative course was uneventful and the patient was discharged 10 days after the surgery. Postoperatively, the patient subsequently underwent 8 cycles of chemotherapy using cisplatin (25 mg/m^2) on day 1 to day 3; and gemcitabine (1000 mg/m^2) on day 1 and day 8, repeated every 21 days. The patient was monitored on a regular basis at our outpatient department and survived 23 months after surgery.

3. Discussion

MEC is a malignant neoplasm, which is most frequently seen in the salivary glands or bronchi but extremely rare in the pancreas.^[6] Since first reported by Franz in 1959, about 10 cases have been reported in literature.^[4,6–8] We summarized all published series about pancreatic MEC which are presented in Table 1. Taking together, a majority of patients tend to be male (70%), with ages between 40 and 70 years. Meanwhile, there is a likelihood that the tumor is always located at the body/tail. In addition, although labeled as an indolent tumor, it always invades the surrounding tissues and lymph node, which was identified by our current manifestation. Curative surgery with R0

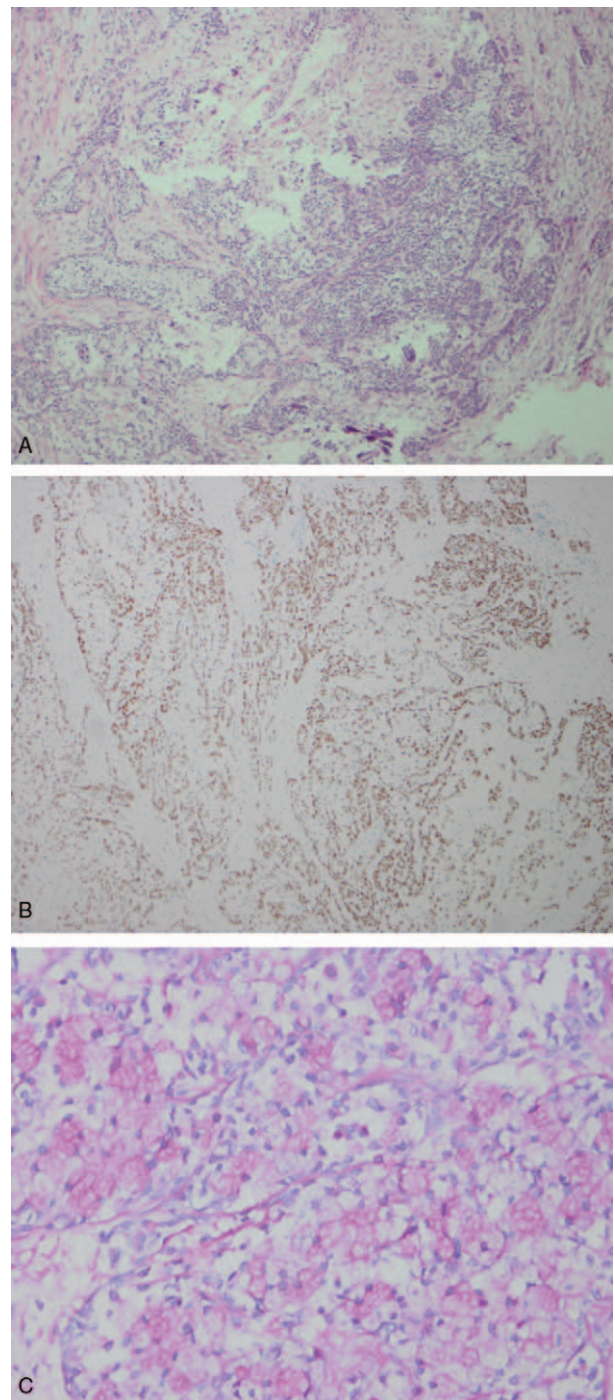


Figure 2. Hematoxylin–eosin staining revealed (A) the tumors consisted of epidermoid cells, intermediate undifferentiated cells, and mucous cells; immunohistochemical staining indicated (B) the tumor was positivity for P 63; special staining identified (C) the tumor was positivity for PAS.

resection margin seems to be the basis to guarantee an improved survival outcome, as the failure to undergo radical operation always resulted in an inferior survival outcome as described in our current literature review. Postoperative multimodal treatment may be a prognostic factor to improve survival outcome, which was identified by Pandey et al^[8] and re-confirmed by us in the present series. In that study, the patient received 6 months chemotherapy by using adjuvant gemcitabine and cisplatin first; then further FOLFIRINOX was also employed, and after that

Table 1**Current literature review about published series of pancreatic mucoepidermoid carcinoma.**

Case	Year	Age/Sex	Location	Tumor Size, cm	Surgical Procedure	Disease Progression	Survival, mo
Ohtsuki	1987	58/M	Tail	10	None	Lung, gallbladder, spleen, adrenals, and LN metastasis	2
Ohshio	1987	69/F	Head	T2	PD	Mesocolon and superior mesenteric vein invasion	3
Kishimoto	1991	48/F	Body	3.5	TP	Solitary liver metastasis	11
Hayashi	1992	58/M	Head	3.5	PD	LN metastasis	6
Kimura	1993	57/M	Body	8	Intraoperative radiation	Stomach, colon, liver, and lung metastasis	2
Onoda	1995	64/M	Tail	8	DP	Spleen, left kidney, and adrenal, colon, and liver metastasis peritoneal dissemination	11
Li	2002	65/M	Body	10	UN	None	UN
Ma	2012	63/F	Body, tail	4.5	DP	None	12
Pandey	2016	50/M	Head	17	TP	Liver, LN	45
Current	2018	48/M	Body, tail	4	DP	LN	23

DP = distal pancreatectomy, LN = lymph node, PD = pancreateoduodenectomy, TP = total pancreateoduodenectomy, UN = unknown.

several chemotherapy or biotherapy were also used so as to prevent disease progression. In our current study, the patient also accepted cisplatin-based therapy after major surgery. From this point of view, the multimodal treatment of curative surgery combined with chemotherapy could help improve survival outcome of these patients.

As an uncommon phenotype in pancreas, the preoperative diagnosis is always puzzle and difficult. Pancreatic MEC manifests nonspecific symptoms that all other tumors share. Computed tomography can accurately estimate the corresponded location of the mass and the relationship with the surrounding organs and vascular systems; however, it also has low specificity in distinguishing MEC from the other types of pancreatic malignancies just before pathological examination. However, there is still something in common. Given the notion that MEC is likely to happen in male patients with ages between 40 and 70 years; tumors located at the body/tail of the pancreas should be mentioned and the diagnosis of MEC should at least be kept in the differential diagnosis. Another aim of the current study was to enhance the awareness in the range of hepatobiliary surgeons that MEC can still happen in the pancreas and caution should also be paid so that omission diagnostic rate could be reduced. However, as to the relatively lower incidence of MEC in pancreas, further analysis is still required so as to establish better regimens and subsequently guide the preoperative diagnosis and treatment option of pancreatic MEC.

In conclusion, we reported a rare case of pancreatic MEC and a literature review of current studies; preoperative diagnosis seems to be difficult in consideration of the rarity of disease and unspecific manifestations. Patients may have a survival benefit from the multimodal treatment of curative surgery combined with chemotherapy.

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