

You cannot miss it: Pancreatic mucoepidermoid carcinoma A case report and literature review

Running head: Mixed mucoepidermoid pancreatic carcinoma

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Abstract : Introduction. Mucoepidermoid carcinoma (MEC) of the pancreas is an exceptionally rare malignancy with fewer than twenty cases reported in the English-language literature. Its pathogenesis, optimal management, and prognosis remain poorly characterised. **Material and methods.** We report a case of mixed mucoepidermoid carcinoma of the pancreatic head diagnosed at our institution and provide a narrative review of all comparable cases identified in the literature through 2024. **Results.** A 69-year-old man presented with progressive cholestatic jaundice and weight loss. CT imaging revealed a 28 mm mass in the pancreatic head with portal vein invasion. CA19-9 was 500 U/mL. Biliary decompression was achieved by ERCP with stenting. Histopathological biopsy confirmed mixed mucoepidermoid pancreatic carcinoma. Given locally advanced unresectable disease, gemcitabine monotherapy (1000 mg/m², D1-D8-D15, q28d) was initiated. After three cycles, imaging showed tumour progression (40×34 mm), portal thrombosis, ascites, and CA19-9 rising to 28,940 U/mL. A switch to FOLFOX was planned but could not be administered due to rapid clinical deterioration. The patient died approximately five months after diagnosis. **Conclusions.** Mixed mucoepidermoid carcinoma of the pancreas is an aggressive and therapeutically challenging entity. Surgical resection remains the only treatment associated with prolonged survival. Gemcitabine monotherapy appears inadequate for locally advanced disease. Immunohistochemical and molecular characterisation is essential. This case is, to our knowledge, the first reported from Africa.

Keywords: mucoepidermoid carcinoma; pancreatic neoplasms; case report; gemcitabine; cholestatic jaundice; rare tumour

Introduction

Mucoepidermoid carcinoma (MEC) is a well-recognised salivary gland tumour characterised by a mixture of mucin-secreting, epidermoid, and intermediate cells. Its occurrence in the pancreas is exceptional; fewer than twenty cases have been documented in the English-language literature since Frantz's first description in 1959 [1, 2]. The term "mixed" MEC designates tumours in which the mucoepidermoid component coexists with an additional histological contingent, further complicating classification. The present case is, to our knowledge, the first reported from the African continent.

Case Report

A 69-year-old man with type 2 diabetes mellitus and hypertension presented with a one-month history of progressive cholestatic jaundice, significant weight loss, and general deterioration. Performance status was OMS 1. Examination revealed sub-icterus and mild epigastric tenderness without palpable mass.

Initial biochemistry showed markedly abnormal liver function: total bilirubin 291 µmol/L, direct bilirubin 155 µmol/L, alkaline phosphatase 1086 U/L, gamma-glutamyltransferase 3740 U/L, AST 388 U/L, ALT 108 U/L, haemoglobin 10.6 g/dL. CA19-9 was elevated at 500 U/mL.

Abdomino-pelvic CT (February 2021) demonstrated a 28 mm tissue mass in the pancreatic head with portal vein and retro-portal lamina invasion, regional lymphadenopathy, and bicanalicular dilatation. No distant metastatic lesion was identified. A multidisciplinary team (MDT) decision led to ERCP with biliary stenting for decompression.

Tissue biopsy was performed at an external laboratory. Histopathological analysis (March 2021) confirmed the diagnosis of pancreatic Mucoepidermoid Carcinoma (MEC) located in the pancreatic head. Immunohistochemical staining yielded the following results: CK wide (+), CK7 (+), CK19 (+), CK5/6 (+), P40 (partially +), CDX-2 (–), and CK20 (–) (Figure1). Metastatic salivary gland MEC was excluded by the absence of a salivary primary on clinical and radiological evaluation.

Given locally advanced unresectable disease, the MDT initiated palliative chemotherapy with gemcitabine monotherapy at 1000 mg/m² on days 1, 8, and 15 of a 28-day cycle (April 2021), after pre-treatment cardiac evaluation (transthoracic echocardiography: ejection fraction 70%). A totally implantable venous access port was placed.

Three cycles of gemcitabine were administered between April and June 2021. Tolerance was generally acceptable; one dose delay occurred at cycle 1 day 15 due to thrombocytopenia (platelets 99,000/mm³). Despite treatment, CA19-9 rose sharply to 51,650 U/mL after cycle 1.

Re-evaluation CT (June 2021) demonstrated clear tumour progression: mass increase from 31×30 mm to 40×34 mm, new partial portal vein thrombosis, and low-volume ascites. CA19-9 was 28,940 U/mL. The MDT decided to switch to FOLFOX (oxaliplatin, folinic acid, 5-fluorouracil). Unfortunately, the patient experienced rapid clinical deterioration and died before receiving any FOLFOX cycle. Overall survival from diagnosis was approximately five months.

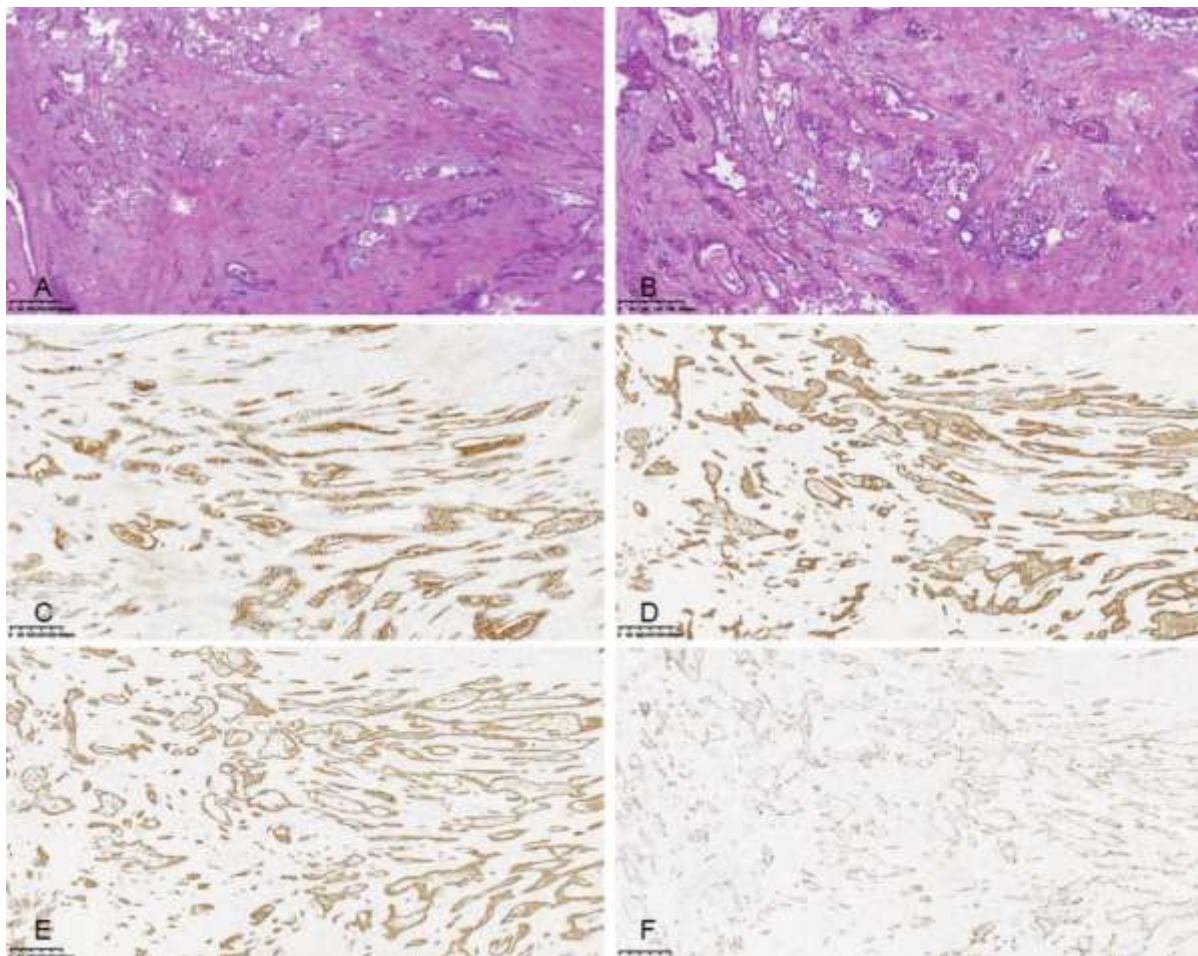


Figure 1. Pathological Examination and Immunohistochemical Findings. A (HE ×40) and B (HE ×100): The tumor shows sheet-like and nested growth within a hyalinized fibrous stroma. Focal keratinization is present, with occasional cells containing intracellular mucin. C-F: Immunohistochemical staining shows positivity for CK7 (C), CK19 (D), CK5/6 (E), and P40 (F) in the tumor cells.

Discussion

Pancreatic MEC is an exceedingly rare malignancy. The largest review to date, by Hu et al. [5], identified fewer than fifteen cases in the literature, noting a predilection for men aged 40–70 years, with jaundice and abdominal pain as the predominant symptoms — consistent with our patient's presentation. A summary of all comparable published cases, including the present one, is provided in Table 1.

Table 1. Reported cases of primary pancreatic mucoepidermoid carcinoma: comparison with the present case.

Author (Year)	Age/Sex	Site	Size (mm)	Vascular invasion	Diagnoses	Treatment	Survival	Reference
Onoda et al. (1995)	NR/M	Tail	Large, multi-organ	Yes (spleen, adrenal)	Surgery + histo	Surgery + adjuvant CT	4 months	[2]
Ma et al. (2012)	63/F	Body/Tail	45	No	Surgery + histo	Distal pancreatectomy	12 months	[3]
Pandey et al. (2016)	50/M	Head	NR	Yes (skin mets)	Surgery + skin biopsy	Pancreatectomy + CT	NR	[4]
Hu et al. (2018)	NR/M	Body	21×24	No	Surgery + histo	Pancreatectomy + cisplatin/gemcitabine ×8	23 months	[5]
Imazu et al. (2021)	75/M	NR	85×85	Yes (LN, liver, lung)	EUS-FNA + autopsy	Chemotherapy only	84 days	[6]
Chen et al. (2021)	56/M	Tail	46×37	Yes (liver)	Surgery + histo + seq.	Pancreatectomy + RFA	NR	[7]
Zhang et al. (2024)	65/F	Body	NR	Yes (liver)	Surgery + IHC + FISH	Pancreatectomy + adjuvant CT	Alive at submission	[8]
Wang et al. (2024)	51/F	Head	28×19	Yes (liver post-op)	PD + IHC	PD + paclitaxel/gemcitabine ×5	12 months post-op	[9]
Present case (2021)	69/M	Head	28→40×34	Yes (portal vein, thrombosis)	Biopsy only	Gemcitabine ×3 → FOLFOX planned (not given)	~5 months	—

CT, chemotherapy; PD, pancreaticoduodenectomy; EUS-FNA, endoscopic ultrasound-guided fine-needle aspiration; IHC, immunohistochemistry; FISH, fluorescence in situ hybridisation; LN, lymph node; RFA, radiofrequency ablation; NR, not reported.

An important and unresolved nosological issue concerns the molecular identity of pancreatic MEC. Saeki et al. demonstrated that these tumours do not harbour the CRTC1/3–MAML2 fusion gene characteristic of salivary gland MEC, and proposed that they be reclassified as pancreatic adenosquamous carcinoma with mucoepidermoid features [10]. This distinction carries prognostic significance: unlike salivary MEC, in which MAML2 fusion confers a favourable outcome, pancreatic MEC behaves as aggressively as adenosquamous carcinoma. Immunohistochemical characterisation — including p63, CK5/6, CK7, and mucin markers — is therefore mandatory for accurate diagnosis, as well as to exclude metastatic salivary MEC [8].

Among the published cases, surgical resection was the only treatment associated with meaningful survival. Hu et al. reported 23 months of survival after distal pancreatectomy and cisplatin/gemcitabine [5]; Wang et al. reported stable disease at 12 months after pancreaticoduodenectomy and paclitaxel/gemcitabine [9]. In contrast, non-resected cases, including ours, uniformly demonstrated survival of less than six months. The autopsy case of Imazu et al., in which a rapidly progressive mixed anaplastic and mucoepidermoid tumour caused tumour rupture and death within 84 days despite chemotherapy [6], illustrates the extreme biological aggressiveness of this spectrum.

In our patient, gemcitabine monotherapy resulted in clear progression after three cycles, reflected by both imaging and a dramatic CA19-9 rise. This is consistent with the known resistance of squamous-component pancreatic tumours to gemcitabine-based regimens. By analogy with pancreatic adenosquamous carcinoma — the closest histological entity — platinum-containing regimens

are expected to yield superior results [10]. The planned switch to FOLFOX was clinically sound; the patient's rapid deterioration, however, highlights the extremely narrow therapeutic window in this disease and the importance of early recognition of first-line failure.

This case carries several limitations: immunohistochemical data were unavailable from the external biopsy report, molecular profiling was not performed, and post-mortem examination was not carried out. Future cases should systematically include comprehensive IHC, MAML2 FISH analysis, and next-generation sequencing to advance understanding of this rare entity.

Conclusions

Mixed mucoepidermoid carcinoma of the pancreas is an exceptionally rare and aggressive malignancy. When diagnosed at a locally advanced, unresectable stage, prognosis is dismal. Gemcitabine monotherapy appears inadequate; platinum-based combinations should be preferred by analogy with adenosquamous carcinoma. Comprehensive histopathological and molecular workup is essential. Reporting such cases remains critical to advancing understanding of this rare entity.

Article Information and Declarations

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S.N ; M.B ;M.M : Writing – review and editing,

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