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Early Stage Anaplastic Sarcomatoid Carcinoma of The Pancreas, A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 64
Final Diagnosis: Anaplastic sarcomatoid carcinoma of the pancreas
Symptoms: Abdominal pain
Medication: —
Clinical Procedure: Pancreatoduodenectomy
Specialty: Oncology

Objective: Rare disease

Background: Anaplastic sarcomatoid carcinoma of the pancreas (ASCP) is a rare variant of pancreatic malignancies. It is a high-grade epithelial carcinoma predominated with spindle cells.

Case Report: We report a case of a 65-year-old patient who presented with early stage, ampullary mass indicating malignancy, and who subsequently underwent pancreatoduodenectomy. Histopathology and immunohistochemistry were confirmatory for anaplastic, grade IV sarcomatoid adenocarcinoma arising from the head of the pancreas.

Conclusions: To our knowledge, this is a rare presentation with few cases reported in the literature.

MeSH Keywords: Adenocarcinoma • Ampulla of Vater • Carcinoma • Immunohistochemistry

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Background

Worldwide, pancreatic cancer is the eighth leading cause of cancer-related deaths in men and the ninth in women [1]. Ductal adenocarcinoma represents the most common type of pancreatic exocrine neoplasms [2]. Most of pancreatic ductal malignancies are moderately to poorly differentiated adenocarcinomas [2]. Histologic grading, which is based upon the degree of differentiation and the prevalence of mitotic cells, typically uses 3 grade levels (grade I, well differentiated; grade II, moderately differentiated; grade III, poorly differentiated), although highly anaplastic tumors are sometimes designated grade IV [3]. We report a case of anaplastic sarcomatoid carcinoma of the pancreas (ASCP), which is an aggressive and extremely rare type of pancreatic cancer.

In this report, we describe the immunohistochemical characteristics of sarcomatoid carcinoma involving the head of the pancreas in a 64-year-old male patient. This case represents the eleventh case of ASCP reported in literature.

Case Report

A 64-year-old male presented with upper abdominal pain and weight loss for almost 3 months. The patient was referred to our Hepatobiliary Team where he was further evaluated. Physical examination was remarkable for epigastric tenderness that prompted further investigations. Laboratory investigation results were as follows: hemoglobin was at 8.9 g/dL (normal range, 13.5–17.5 g/dL), WBC was 8.25×10^9/L (normal range, 4–11×10^9, platelet count was

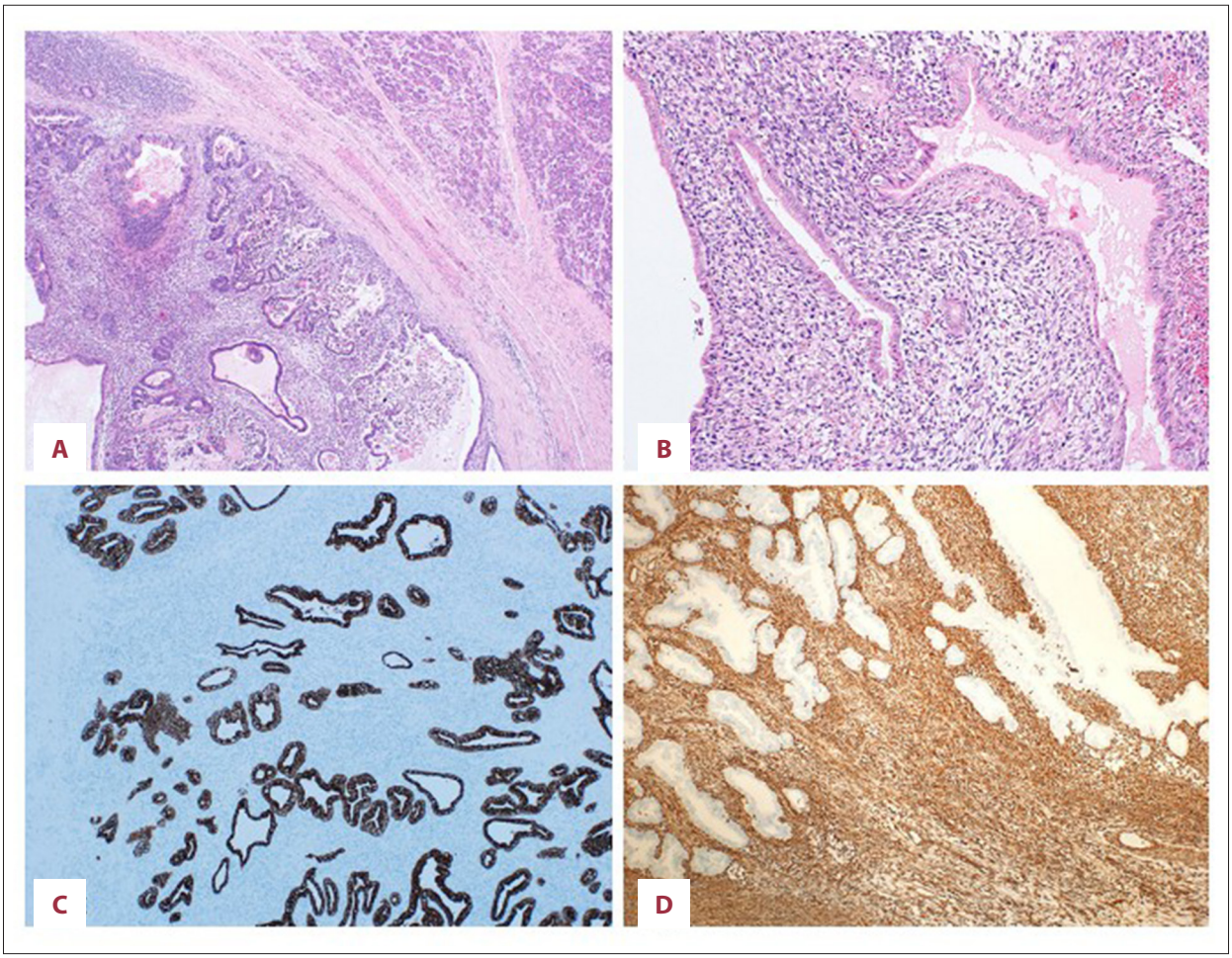

Figure 1. (**A, B**) Histological examination of the pancreatic neoplasm reveals infiltration by malignant cells displaying biphasic (epithelial and sarcomatoid) morphology. The epithelial component is in the form of ductal adenocarcinoma consists of malignant glands. The sarcomatoid component surrounds the malignant glands and is composed of sheets of malignant spindle-shaped cells. A rim of unremarkable pancreatic tissue is seen at the top right part of (**A**) Hematoxylin and eosin, 40×, (**B**) Hematoxylin and eosin 100×. (**C, D**) Immunohistochemical stains show that the neoplastic epithelial cells forming glands are strongly and diffusely positive for CAM5.2 and negative for vimentin while the neoplastic cells showing sarcomatoid differentiation lost CAM 5.2 positivity and are positive for vimentin (mesenchymal marker).

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374×10^9/L (normal range, 150–450×10^9/L). Carbohydrate antigen 19–9 (CA 19-9) was at 67.82 IU/L (normal range, <37 IU/L), amylase was 195 unit/L (normal range, 25–115 unit/L), lipase was 5566 unit/L (normal range, 147–310 unit/L). Liver enzymes concentrations were in the normal range.

A computed tomography (CT) scan of the abdomen revealed a well-defined mass involving the head of the pancreas, protruding through the distal pancreatic duct and highly suspicious for malignancy. Gallstones were also noted. Biopsy results were remarkable for adenocarcinoma. Positron emission tomography-CT (PET-CT) showed no evidence of distant metastasis.

The patient underwent a pancreatoduodenectomy with cholecystectomy without postoperative complications.

The surgical specimen consisted of duodenum, head of pancreas, common bile duct, cystic duct, and gall bladder. Opening of the duodenum revealed a gray-pink polypoid tumor protruding from the ampulla of Vater, measured 2.4×2×1.9 cm. The mass infiltrated into the pancreatic head parenchyma. Omental biopsies were negative for metastasis.

Microscopically, the tumor was consistent with anaplastic, grade IV, ductal adenocarcinoma with sarcomatoid features (Figure 1). The tumor was confined to the head of the pancreas with no invasion into ampulla of Vater or duodenal wall. All surgical margins were clear. Lymphovascular invasion and perineural invasion were both not identified. The tumor was pathologically staged at pT2N0M0. Twelve lymph nodes were examined and none of them were involved with disease.

The patient completed a 6-month course of adjuvant gemcitabine and was then followed on surveillance. At 19 months post-surgery, he was alive and continued to do well.

Discussion

ASCP is an extremely rare type of pancreatic carcinoma characterized by extremely rapid progression and poor outcome as compared to typical pancreatic ductal carcinomas. The World Health Organization classification of exocrine pancreatic tumors assigns spindle cell carcinoma, sarcomatoid carcinoma, and carcinosarcoma under the category of undifferentiated (anaplastic) carcinoma [3,4].

Sarcomatoid carcinoma comprises a mixture of carcinomatous and sarcomatous elements. ASCP usually demonstrates cellular patterns similar to those present in tumors of mesenchymal origin [3,4].

In addition to this patient, 10 cases of pancreatic sarcomatoid carcinoma with confirmed epithelial derivation of the spindle component and/or absence of specific mesenchymal differentiation have been reported [5–14]. Of the previously reported patients (Table 1) our case is the first T2N0M0 ASCP case reported.

Table 1. Summary of reported cases of localized and metastatic sarcomatoid carcinoma of the pancreas.

Cases	Age at diagnosis (years)	Gender	Tumor size (cm)	Involved part of the pancreas	Extent	Sarcomatoid component	Treatment	Follow-up, months/outcome
Case 1	74	Male	4.5×4×3	Head	Duodenal invasion, with blood vessel and perineural	Cytokeratin (CK) AE1, variable CK AE3, epithelial membrane antigen (EMA), apoprotein in MUC-1 (MUC1-ARA) (+), S100, smooth muscle actin (SMA) (+), desmin, vimentin, (–)	Pancreatoduodenectomy	3 months/ succumbed to diffuse peritoneal carcinomatosis [5]
Case 2	74	Male	4×3	Head	Peripancreatic adipose tissue and the duodenal wall.	Vimentin (+), CK (+)	Pancreatoduodenectomy	4 months/alive [6]
Case 3	67	Female	2.5×2.5×2	Head	Peripancreatic lymphadenopathy	Spindle cells (SC), separate focus of OGC; CK8/18 and vimentin (D+)	Pancreatoduodenectomy	Information not available [7]

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Table 1 continued. Summary of reported cases of localized and metastatic sarcomatoid carcinoma of the pancreas.

Cases	Age at diagnosis (years)	Gender	Tumor size (cm)	Involved part of the pancreas	Extent	Sarcomatoid component	Treatment	Follow-up, months/outcome
Case 4	72	Female	N/A	N/A	N/A	CK and vimentin (+)	Conservative	9 months/ succumbed to sarcomatoid carcinoma metastatic to the liver [8]
Case 5	82	Female	18×11×10	Head	Penetrated the transverse mesocolon, resulting in massive hemorrhagic clots in the abdominal cavity	SC, foci of OGC around hemorrhage; (SC): vimentin, CD10 (+), CK, AE1/AE3 (+)	Radical pancreatodu-odenectomy with partial resection of the transverse colon	Deceased due to DIC on postoperative day 13 [9]
Case 6	48	Male	3.5×2.5×1.5	Body	Liver metastasis	SC, scattered OGC; vimentin (+), pan-CK, CK, 7, CK8/18, EMA, CEA, CD34, CD56, CD68, CD117, desmin, SMA, myogenin, S100	Pancreatectomy with splenectomy and colonic segmental resection	4 months/ deceased secondary to hepatic and peritoneal metastases [10]
Case 7	85	Male	3.3×3×2.6	Body	Adherent to the serosa of the stomach	Diffuse pan-CK, CK5.2, p53 (+)	Distal (near-total) pancreatectomy, splenectomy, and partial gastrectomy	26 months/ alive and well [11]
Case 8	48	Male	10×8×5	Tail	Nil	CK 18 and vimentin	Left pancreatectomy, adjuvant gemcitabine 1 cycle	3 months/ succumbed [12]
Case 9	55	Male	14	Body and tail	N/A	CK, CK7, and vimentin	Distal pancreatectomy, splenectomy, and colonic segmental resection	Information not available [13]
Case 10	41	Male	2.2×2.1	Head and uncinate	Liver metastasis	CK and vimentin	Gemcitabine	1 month/on chemotherapy when reported [14]
Case 11	64	Male	2.4×2×1.9	Head	N/A	CAM 5.2 and vimentin	Pancreatodu-odenectomy/ cholecy-stectomy followed by adjuvant gemcitabine	19 months/ alive and well Our reported case

CK – cytokeratin; IHC – immunohistochemistry; SC – spindle cell; EMA – epithelial membrane antigen; SMA – smooth muscle actin; MUC1-ARA – apoprotein MUC1; ER – estrogen receptor; PR – progesterone receptor; OGC – osteoclastic giant cells; DIC – disseminated intravascular coagulopathy; N/A – not available; NSE – neuron-specific enolase; CEA – carcinoembryonic antigen.

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Of the patients with adequate follow up, 6 out of 7 patients succumbed to their condition within 9 months of surgery. The longest survival reported was 26 months, documented in an 85-year-old male who underwent distal (near-total) pancreatectomy, splenectomy and partial gastrectomy. Three patients had liver metastasis on presentation [14].

Notably, the patient of our present case study was alive and well 19 months after surgery and thus, to the best of our knowledge, is the second longest-living individual with ASCP reported in the English literature.

Conclusions

Anaplastic sarcomatoid ductal adenocarcinoma of the pancreas is an extremely rare variant of pancreatic cancer. The current case documents the eleventh case of ASCP, and the first case staged at T2N0M0. The patient described in this case study was alive and well at 19 months after surgery, and to our knowledge, this is the second longest-living individual with history of ASCP diagnosis.

Conflicts of interest

None.

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