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Long-term survival after resection of sarcomatoid carcinoma of the pancreas: an updated experience

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ABSTRACT

Background: Sarcomatoid carcinoma of the pancreas (SCP) is a rare histologic subtype of undifferentiated pancreatic carcinoma. Historically, this has been associated with a worse overall prognosis than adenocarcinoma. However, the clinical course and surgical outcomes of SCP remain poorly characterized owing to its rarity.

Methods: A single-institution, prospectively maintained database was queried for patients who underwent pancreatic resection with a final diagnosis of SCP. We describe their histology, clinicopathologic features, and perioperative outcomes. Survival data are highlighted, and common traits of long-term survivors are examined.

Results: Over a 25-year period, 7009 patents underwent pancreatic resection at our institution. Eight (0.11%) were diagnosed with SCP on final histopathology. R0 resection was achieved in six patients (75%). Four patients had early recurrence leading to death (<3 months). Two (25%) experienced long-term survival (>5 years), with the longest surviving nearly 16 years despite the presence of lymph node metastasis. There were no deaths attributed to perioperative complications. Both long-term survivors had disease in the body/tail of the pancreas and received adjuvant radiotherapy. One also received adjuvant gemcitabine-based chemotherapy.

Conclusions: SCP is a rarely appreciated subset of pancreatic malignancy that does not necessarily portend to a uniformly dismal prognosis. Although some have rapid recurrence and an early demise, long-term survival may be possible. Future studies are needed to better define the cohort with potential for long-term survival so that aggressive therapies may be tailored appropriately in this patient subset.

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Introduction

Pancreatic cancer is an aggressive malignancy that continues to be a leading cause of mortality. An estimated 53,070 new cases are expected to occur in the United States in 2016, with approximately 42,000 deaths.[1] Sarcomatoid carcinoma of the pancreas (SCP) is a rarely observed malignant subtype characterized by a predominance of spindle cells and sarcomatous morphologic features with epithelial derivation.[2-4] This histologic feature is distinct from sarcoma, which is broadly defined as a tumor of mesenchymal differentiation. The World Health Organization (WHO) classification of gastrointestinal tumors identifies SCP as a subset of the broad class of undifferentiated carcinomas. Other undifferentiated carcinomas that can occur in the pancreas include giant cell tumors and pleomorphic undifferentiated carcinomas (often termed anaplastic carcinoma).[5] Although undifferentiated carcinoma of the pancreas has a reported incidence of 5%, SCP is exceptionally rare. Most examples are found in the literature only as case reports.[6]

Treatment for pancreatic adenocarcinoma as a whole remains challenging. Complete surgical extirpation is the only chance of cure, with 5-year survival rates of approximately 20%.[7,8] Although no direct comparisons exist, SCP has historically been thought to have a long-term prognosis that is worse than ductal adenocarcinoma.[2,9,10] Despite aggressive surgical management, median postoperative survival has been consistently reported at less than 1 year, with many succumbing to early carcinomatosis.[2,9-12] Owing to the rarity of disease, the clinical course, surgical outcomes, and optimal treatment strategies for SCP are poorly characterized. In this study, we report our institution's experience with sarcomatoid carcinoma to further characterize the clinicopathological features, surgical outcomes, and survival data in the individuals that undergo resection for this rare diagnosis.

Methods

A retrospective cohort analysis of a prospectively managed database was conducted of patients who underwent pancreatectomy with curative intent at the Johns Hopkins Hospital from October 1991 to December 2015. Internal Institutional Review Board approval was obtained for the creation and use of this deidentified database for research purposes with waiver of informed consent.

Cases were initially identified by searching our pathology database for the terms "sarcomatoid", "sarcomatous" and "spindle cell". Patients identified had a primary diagnosis consistent with SCP and were reviewed by our pancreatic pathologists. Available cases had slides rereviwed and diagnosis confirmed. The histopathologic diagnosis of SCP was defined as the presence of poorly differentiated or anaplastic cells with a predominance of spindle cells, sarcomatoid features, and epithelial derivation. Pathology suggestive of pancreatic sarcoma or anaplastic carcinoma without sarcomatoid features was excluded.

Patient demographics, characteristics, operative, and perioperative outcomes were assessed via chart review. Individuals who obtained initial workup or neoadjuvant/adjuvant treatment at outside institutions were included. Perioperative mortality was defined as death within 30 days of the operative date or mortality before hospital discharge from index admission. Complications were graded by the Clavien-Dindo scale.[13] Overall survival was calculated from the date of surgery. Date of death was obtained from medical records, social security death index, or local obituaries.

Results

From October 1991 to December 2015, 7009 patients underwent pancreatic resections, eight (0.11%) patients had a final pathologic diagnosis of SCP. The median patient age was 66 years (range: 54-80). Three patients were male (37.5%). The majority of patients presented with vague back or abdominal pain (62.5%) and the remaining were asymptomatic (37.5%) on initial preoperative visit with tumor found incidentally. Additional findings on presentation included jaundice (12.5%) and weight loss (12.5%). Preoperative workup included multiphase CT scan. Preoperative imaging was unavailable for review in two patients; however, of the images available, pancreatic ductal dilatation was appreciated in four cases (66%), atrophy of remaining pancreas in three cases (50%), a hypodense lesion in five cases (83%), cystic appearance in two cases (33%), heterogenous enhancement in two cases (33%), and venous phase peripheral enhancement in two cases (33%). Tumor size in greatest dimension was a median of 5 cm with range of 3-15 cm. Preoperative tissue sampling by needle biopsy was obtained in four patients: two (50%) were identified as pancreatic adenocarcinoma, one (25%) as pleomorphic adenocarcinoma, and one (25%) as atypical epithelioid cells insufficient for diagnosis. Patient cohort characteristics are represented in Table 1.

Pathologic analysis revealed a diagnosis of SCP in all cases. Two cases included the presence of mixed osteoclast-like giant cells (OCGCs). Pathologic grade ranged from poorly differentiated (43%) to anaplastic undifferentiated (57%). Five (62%) revealed lymphovascular invasion, and perineural invasion was present in four (50%) cases. Two (25%) subjects did not have any lymph node metastasis appreciated, whereas the remaining subjects had at least one lymph node positive for metastatic carcinoma. Of note, the longest survivor did have lymph node metastasis in one of 22 nodes at the time of operation.

An R0 resection was achieved in 75% of cases, one of which required a vascular resection of the superior mesenteric vein.

Table 1 – Characteristics of patients with sarcomatoid carcinoma of the pancreas.

Sarcomatoid cancer patients ($n = 8$)	Value
Mean age (years), range	67.1 (54-80)
Tumor size greatest dimension (cm) median, range	5 (3-15)
R0 resection	6 (75%)
Node involvement (≥1)	6 (75%)
>5-year survival	2 (25%)