Sarcomatoid carcinoma of pancreas with liver metastases – A case report with review of literature

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Abstract

Sarcomatoid carcinoma is a rare, high-grade epithelial malignancy composed exclusively or predominantly of spindle cells often having features suggestive of epithelial derivation without features indicative of a specific line of mesenchymal differentiation. Sarcomatous carcinomas occur in diverse locations throughout the body, including the upper respiratory tract, upper and lower digestive tracts, genitourinary tract, breast and thyroid glands, among others. Sarcomatoid carcinoma rarely presents in the pancreas. The current study presents the case of a 41-year-old male with a tumor mass in the uncinate of the pancreas and liver metastases. Histopathology and immunohistochemistry of the liver metastases were confirmatory of metastatic sarcomatoid carcinoma. To the best of our knowledge, only nine cases of pancreatic sarcomatoid carcinoma have been reported in literature.

Introduction

Sarcomatoid carcinoma is an aggressive form of carcinoma composed of malignant spindle cell elements, with or without a coexisting epithelial cell component. Ultrastructural analyses of sarcomatoid carcinoma have shown a spectrum of differentiation ranging from epithelial to mesenchymal-type features. Sarcomatoid carcinoma occurs very rarely in the pancreas. We describe here a 41-year-old male patient with sarcomatoid carcinoma of the pancreas with liver metastases with the supporting immunohistochemical (IHC) and histological findings. The current study represents only the tenth case of sarcomatoid carcinoma of the exocrine pancreas as defined above that has described in literature [1-11].

Case report

A 41-year-old man presented with vague abdominal discomfort and a five-kg weight loss over one month. His Eastern Cooperative Oncology Group performance score was one. Physical examination revealed a large, firm, nontender mass in the left upper quadrant of abdomen. All blood investigations were within normal limits, including those for tumor markers (CEA, CA 19-9, AFP, and CA 125). Abdominal computed tomography with contrast enhancement showed an ill-defined hypodense lesion of size 22 × 21 mm in the uncinate process of pancreas with multiple small rounded lesions in various segments of liver (suggestive of hepatic metastases) (Figure 1). CT guided biopsy of the liver lesion showed spindle cells having abundant eosinophilic cytoplasm and pleomorphic nuclei admixed with inflammatory cells (Figure 2). IHC staining was strongly positive for cytokeratin (Figure 3). Histology and IHC results confirmed the diagnosis of sarcomatoid carcinoma.

Discussion

Sarcomatoid carcinoma is characterized by proliferation of malignant spindle cell that in some cases demonstrates epithelial derivation without any ultrastructural or immunohistochemical features suggestive of a specific line of mesenchymal differentiation, but in some other cases demonstrate mesenchymal-type differentiation. The microscopically nondescript spindle cells typically express keratin or other epithelial-related markers consistent with an epithelial pathogenesis. Carcinosarcoma is an epithelial malignancy associated with sarcomatoid carcinoma with an equally aggressive clinical course that by definition demonstrates biphasic epithelial and mesenchymal differentiation. Therefore, for practical diagnostic purposes, carcinosarcoma is used interchangeably with sarcomatoid carcinoma. In the present case report, carcinosarcomas with heterologous mesenchymal elements demonstrating light microscopic and/or immunohistochemical evidence of specific mesenchymal (lipogenic, smooth or skeletal muscle, peripheral nerve sheath, vascular or osteo-/cartilaginous) differentiation were excluded. World Health Organization classification of exocrine pancreatic tumors assigns spindle cell carcinoma, sarcomatoid carcinoma and carcinosarcoma under the category of undifferentiated (anaplastic) carcinoma [11,12], since the majority of these types of tumor harbors a spindle element that demonstrates an epithelial immunohistochemical profile and/or genetic alterations in pancreatic ductal adenocarcinomas [1,13].

In addition to the tumor of the present case study, nine additional examples of pancreatic sarcomatoid carcinoma with confirmed epithelial derivation of the spindle component and/or absence of specific mesenchymal differentiation have been identified in a comprehensive review of literature. Of the previously reported nine cases (Table 1), only two patients had metastatic disease at presentation.

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similar to our case and both patients had liver metastasis. Of the patients with adequate follow-up, six out of seven patients succumbed to their condition within nine months of surgery. Longest survival was twenty-six months, documented in a 85 year old male who underwent distal (near-total) pancreatectomy, splenectomy and partial gastrectomy [8]. In a previous review of pancreatic tumors classified as ‘carcinosarcomas’, including two cases that are included in the present case report as examples of sarcomatoid (spindle cell) carcinoma [3,4],
average post-operative survival interval was six months and that the longest living patient survived for fifteen months. In conclusion, the current case study documents the tenth case of sarcomatoid (spindle cell) carcinoma of the pancreas with data substantiating the epithelial derivation of the nondescript malignant spindle cell element.

References

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