

# High-grade malignant pancreatic neoplasm with sarcomatoid features

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**Abstract:** Sarcomatoid carcinoma is a rare and aggressive form that occur at diverse locations in the body such as upper respiratory tract, upper and lower digestive tracts, genitourinary tract, breast and thyroid glands. However, its occurrence in pancreas has been rarely reported. Sarcomatoid carcinoma of pancreas (SCP) is a high-grade epithelial malignancy composed predominantly of spindle cells often having features suggestive of epithelial derivation without features indicative of a specific line of mesenchymal differentiation. Its pathogenesis has not been elucidated. Microscopically, SCP comprises mostly anaplastic cells and is strikingly sarcoma-like in appearance. Confirmation of this disease is often based on the pathological diagnosis. We report a case that was incidentally found after a CT was done for worsening chronic back pain and the patient was found to have a pancreatic mass and a liver lesion. Endoscopic ultrasound (EUS)-guided liver biopsy revealed high grade malignant pancreatic neoplasm with sarcomatoid features. Further CT chest revealed bilateral lung nodules and PET scan revealed prominent bony metastases within vertebral bodies at L1, L2, and L3. The patient refused definitive treatment and succumbed to illness within 3 months.

**Keywords:** Sarcomatoid carcinoma of pancreas (SCP); spindle cell; undifferentiated pleomorphic sarcoma of the pancreas (UP-PS)

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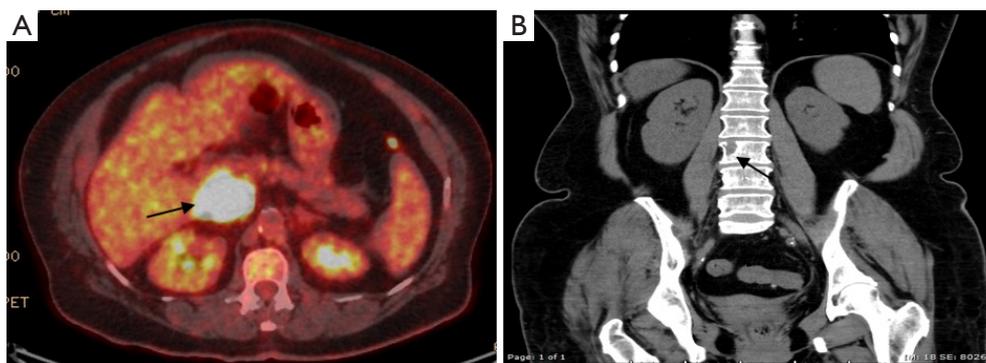
## Introduction

Sarcomatoid carcinoma arising in pancreas is extremely rare, with only few cases reported in the literature (1-4). Histologically, it is a subtype of undifferentiated pancreatic carcinoma composed predominantly of malignant spindle cells, with or without a coexisting epithelial cell component. Its pathogenesis remains unclear. Microscopically, sarcomatoid carcinoma of pancreas (SCP) is comprised mostly of anaplastic cells and is strikingly sarcoma-like in appearance. Spindle cells appeared to be epithelial in some tumors and mesenchymal in origin in other tumors. Confirmation of this disease is often based on the pathological diagnosis and are often more aggressive with poor prognosis. Despite aggressive surgical management, median postoperative survival has been consistently reported at less than 1 year, with many succumbing to

early carcinomatosis. We discuss a fatal case of SCP with metastasis to lungs, liver and vertebrae.

## Case presentation

A 64-year-old female with medical history significant for chronic obstructive pulmonary disease (COPD), diabetes and long-term cigarette smoking. The patient was incidentally found to have a pancreatic mass, and liver lesion after undergoing compute tomography (CT) for worsening chronic back pain. Accompanying symptoms included a loss of appetite and pruritus. Imaging with abdominal CT revealed a pancreatic head mass (3.7 cm × 3.6 cm) along with a liver mass (1 cm) in the lateral right lobe (*Figure 1A*). Endoscopic ultrasound (EUS)-guided liver biopsy was performed and revealed high-grade spindle cell sarcomatoid cancer. Microscopically, poorly differentiated



**Figure 1** Images depicting the metastases of the tumor. (A) PET-CT scan depicting right hepatic lesion (arrow); (B) PET scan showing L-3 metastatic lesion (arrow). PET, positron emission tomography; CT, computed tomography.

neoplastic cells showed marked nuclear pleomorphism. Immunohistochemistry was positive for vimentin and CD56 while negative for estrogen receptors or epithelial markers. Given the clinical and radiological findings, it is felt the liver lesion was most likely a metastatic lesion from the pancreatic mass. Positron emission tomography (PET) scan revealed prominent bony metastases within vertebral bodies at L1, L2, and L3 (*Figure 1B*). The patient refused surgical treatment, however she was treated with palliative radiotherapy for spinal metastasis. Despite treatment, her condition deteriorated, and she passed away.

## Discussion

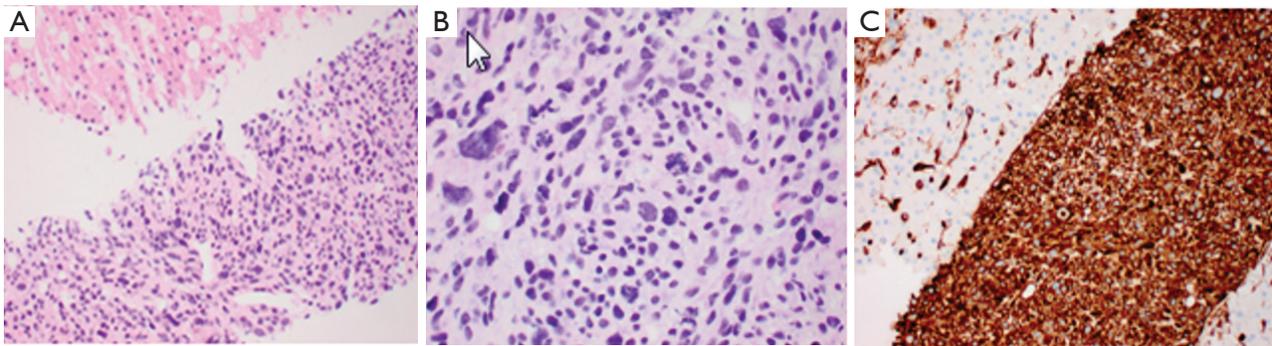
SCP are rare anaplastic variants which comprise two percent of pancreatic exocrine tumors. According to the 2010 World Health Organization (WHO) classification of tumors, the sarcomatoid carcinoma of the pancreas is classified under the undifferentiated carcinomas of pancreas together with spindle cell carcinoma and carcinosarcoma (5). SCP is an aggressive form of carcinoma composed of malignant spindle cells and undifferentiated cells without morphologic evidence of glandular, squamous or urothelial differentiation. In some cases, the malignant spindle cell proliferation can be admixed with discohesive, bizarre, and multinucleated giant cells without any ultrastructural or immunohistochemical features suggestive of a specific line of mesenchymal differentiation (6). However, the diagnosis of SCP should only be made after demonstrating epithelial derivation either immunohistochemically or at the ultrastructural level (5).

Imaging techniques such as ultrasound, CT and MRI may help to identify the nature of the tumor. A clear

bordered, cystic-solid mass with heterogeneous density is the most common imaging feature of undifferentiated carcinoma of the pancreas (5). Pathological confirmation is a reliable way to prove the diagnosis of undifferentiated carcinoma of the pancreas. However, in practice, it is hard to differentiate subtypes, since most of these types of tumor possess a spindle element.

### *SCP or primary pancreatic undifferentiated pleomorphic sarcoma (UP-PS)*

The histopathologic diagnosis of SCP includes the presence of poorly differentiated or anaplastic cells with a predominance of spindle cells, sarcomatoid features, and epithelial derivation (6). In the present case, given the clinical history of a pancreatic head mass with liver and lung nodules, and although the morphology and immunophenotype was non-specific, the overall findings were most consistent with metastatic pancreatic sarcomatoid carcinoma when the clinical picture was taken into account. Additionally, microscopic examination suggests SCP and revealed a highly cellular spindle cell proliferation arranged in a fascicular growth pattern and characterized by pleomorphic spindle cells with occasional giant cells (*Figure 2A,B*). In most cases of SCP, spindle cells are positive for cytokeratin and vimentin, however in this case the spindle cells were only positive for vimentin (*Figure 2C*), raising the possibility of a primary pancreatic undifferentiated pleomorphic sarcoma (UP-PS). UP-PS is an extremely rare entity which represents 0.1% of all pancreatic tumors and is usually a diagnosis of exclusion. Although there was no confirmatory evidence of epithelial differentiation in our case, the reactivity of SCP with



**Figure 2** Histopathological characteristics and immunostaining of the tumor. (A) Tumor showing fascicular growth pattern characterized by highly cellular and pleomorphic spindle cells with occasional giant cells 2 (HE staining,  $\times 10$ ); (B) atypical mitotic figures (center) and bizarre pleomorphic giant cells 2 (HE staining,  $\times 20$ ); (C) tumor positive for vimentin immunostain ( $\times 20$ ).

epithelial markers can be focal especially in small biopsies, as was in our case.

The clinical course and surgical outcomes of SCP are poorly characterized owing to its rarity. Further, the impact of adjuvant chemotherapy on survival of SCP has not been well defined. Given its aggressive biological behavior and poor prognosis, it is of prime importance to make early diagnosis for patients with SCP.

### Conclusions

Conclusively, in the present case the absence of epithelial differentiation morphologically or by immunostains confined the differential diagnosis to SCP and UP-PS. SCP is very rare pathological type of carcinoma with poor prognosis. It should be included in the differential diagnosis of pancreatic mass lesions with appropriate image characteristics. Many of the reported cases have described rapidly fatal outcomes, thus highlighting its aggressive behavior.

### Acknowledgements

None.

### Footnote

*Conflicts of Interest:* The authors have no conflicts of interest to declare.

*Informed Consent:* The patient passed away and exhaustive attempts have been made to contact the family and that the

paper has been sufficiently anonymized not to cause harm to the patient or their family.

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