

## OPEN

## Sclerosing Epithelioid Mesenchymal Neoplasm of the Pancreas

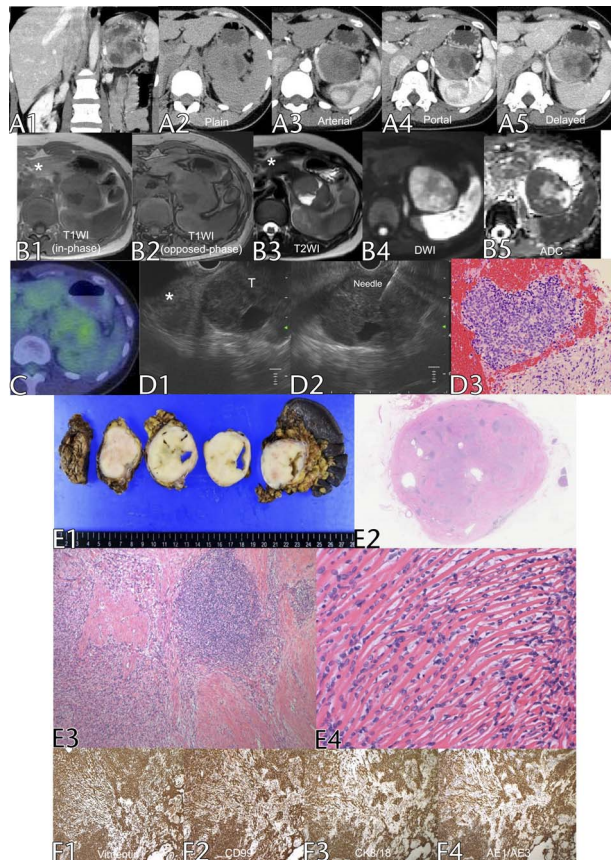
### Case Report and Literature Review of the Morphologic Characteristics

#### To the Editor:

Sclerosing epithelioid mesenchymal neoplasm (SEMN) of the pancreas was first reported as a new entity by Basturk et al<sup>1</sup> in 2020. It occurs most frequently in young women and has a favorable prognosis. Microscopically, it has a tendency to form “geographic” and “slit-like” patterns of enmeshed tumor cells. Immunohistochemically, the tumor has a mesenchymal nature, and molecularly, it demonstrates no recurrent somatic mutations but a distinct clustering of methylation profiling, compared with other mesenchymal neoplasms.

A 31-year-old woman was referred to our hospital with a suspected solid pseudopapillary neoplasm (SPN) of the pancreatic tail. Computed tomography (CT) revealed a well-demarcated hypovascular tumor approximately 5 cm in diameter in the pancreatic tail (Fig. 1A). Compared with the parenchyma of the rest of the pancreas, the solid part of the tumor exhibited a combination of hypointensity on T1-weighted images (T1WI) and hyperintensity on T2-weighted images (T2WI) on magnetic resonance imaging (MRI), whereas the cystic part showed remarkable hyperintensity on T2WI suggestive of fluid collection (Figs. 1B1–B3). Diffusion-weighted imaging revealed moderate diffusion restriction (Fig. 1B4), and the apparent diffusion coefficient values were attenuated in the solid part of the mass (Fig. 1B5). In the 18F-fluorodeoxyglucose positron emission CT, the tumor showed moderate focal uptake in the solid part of the mass (Fig. 1C). During endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA), the tumor showed a distinct hypoechoic large mass with a well-defined boundary with parenchyma of the rest of the pancreas (Figs. 1D1, D2). Biopsy specimens obtained by EUS-FNA revealed epithelioid cells with round to oval nuclei and scant cytoplasm (Fig. 1D3), and immunohistochemically, it was positive for vimentin and cytokeratin, but negative for a specific immunophenotype. Based on these features, we excluded SPN, pancreatic neuroendocrine tumor, acinar cell carcinoma, and ductal adenocarcinoma.

As we could not rule out a malignancy, we performed distal pancreatectomy.



**FIGURE 1.** A1, Coronal view of the CT shows a round hypovascular tumor. A2–A5, The tumor shows slight enhancement in the late phase, and calcification and rim enhancement are not evident in the axial view of the dynamic CT. B1–B5, Abdominal MRI. The solid part of the tumor is homogeneously hypointense on T1WI and hyperintense on T2WI compared with the body of the pancreas. Images of diffusion-weighted imaging (B4) and apparent diffusion coefficient (ADC) values (B5). C, Image of 18F-fluorodeoxyglucose positron emission CT. D1 and D2, Images obtained during EUS-FNA. D3, Hematoxylin and eosin staining of biopsy specimen obtained by EUS-FNA. E1, The resected specimen. E2, Loupe image of hematoxylin and eosin staining reveals sclerotic well-circumscribed structures of the tumor. E3, The tumor shows a geographic pattern. E4, Enmeshed tumor cells produce a slit-like pattern. F1–F4, The results of the immunohistochemistry. T indicates tumor. \*Indicates the body of the pancreas.

Macroscopically, the surgical specimen was a solid and round, well-circumscribed, and nonencapsulated tumor with a diameter of 5.4 cm, without calcification or hemorrhagic tissue (Figs. 1E1, E2). Microscopically, it was composed of epithelioid to spindle cells with mild to moderately atypical nuclei and scant cytoplasm and demonstrated hypocellular and hypercellular areas (a “geographic pattern”) of epithelioid cell nests (Fig. 1E3). Hypocellular and hypercellular areas containing collagenous stroma were evident, with a slit-like formation consisting of enmeshed tumor cells with dense fibrosis in some areas (Fig. 1E4). All tumor cells were positive for vimentin and cytokeratin and showed

no definitive direction of differentiation to any known pancreatic neoplasm, in agreement with the reported previously immunohistochemical characteristics of SEMN (Fig. 1F, Table 1). The patient was diagnosed with SEMN of the pancreas and has been followed up as an outpatient without adjuvant chemotherapy. She is alive and disease free at 19 months after surgical treatment.

Among the pancreatic neoplasms, SPN of the pancreas is a major differential diagnosis from the perspective of age and sex because it generally affects young females. Solid pseudopapillary neoplasm comprises a large well-encapsulated mass with solid and cystic components from

**TABLE 1.** Results of Immunohistochemistry and Genomic Analysis Compared With the Original Report

IHC and Molecular Features	Our Case	Basturk et al, 2020
		+/-
AE1/AE3	+	8/0
CK18	+	8/0
CD99	+	8/0
Vimentin	+	8/0
Chromogranin A	-	0/8
Synaptophysin	-	1/8
PR	-	0/8
CD10	-	0/8
β-Catenin	-	0/8
Trypsin	-	0/8
TTF1	-	0/8
MCU4	-	0/8
Desmin and myogen	-	0/8
INI1 (BAF-47)	+	3/3
CD117 and DOG1	-	0/8
S-100	-	1/8
HMB-45 and Melan A	-	0/8
CD31 and ERG	-	0/8
CD21	-	0/8
CD45	-	0/8
BCL-2	-	0/8
ALK	-	0/8
CD34	-	0/8
STAT6	-	0/3
EWSR1 gene fusion	-	0/5
SYT-SSX fusion gene	-	0/5

ALK indicates anaplastic lymphoma kinase; IHC, immunohistochemistry; PR, progesterone receptor.

hemorrhagic degeneration; in some cases, calcifications and fibrous thickened capsules are seen at the periphery of the mass.<sup>2</sup> On MRI, SPN is a well-defined lesion with a mixture of high and low signal intensities on T1WI and T2WI that are characteristic of intratumoral blood products and necrotic tissues.<sup>3</sup> It is noteworthy that the present case showed a different distribution of T1 and T2 signal intensities between the solid and cystic parts of the tumor. Undifferentiated carcinoma should be considered in the differential diagnosis because of its mesenchymal entities such as spindle cells and vimentin and cytokeratin expression, and its poor prognosis.<sup>4,5</sup> The histological diagnosis of SEMN by EUS-FNA may be difficult because SEMN does not have a specific immunophenotype and diagnostic molecular features. In the current situation, it would be difficult to obtain the exact diagnosis if a patient suspected of having SEMN did not undergo surgical resection.

Supplemental Table 1 (<http://links.lww.com/MPA/A877>) summarizes the characteristics of various pancreatic tumors that show hypoattenuation and well-demarcated lesions morphologically or a mesenchymal nature histopathologically in the differential diagnosis of SEMN.

Further accumulation of similar cases is required to elucidate its clinicopathological and imaging characteristics.

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## The Role of Daily Physical Activity on Pancreatic Cancer Survival A Retrospective Cohort Study

#### To the Editor:

The incidence of pancreatic cancer increases worldwide, and despite the advances in diagnosis and therapy, the improvement in survival rate is regarded as marginal.<sup>1</sup> Surgery remains the cornerstone of a multidisciplinary approach to cure this lethal disease. Pancreatectomy remains a challenging operation characterized by high morbidity.<sup>2,3</sup> The quality of life and survival are critical outcomes. Exercise helps patients to not only live longer but also better; it is known that it can improve mental and emotional functioning and boost productivity. The aim of this single-center, retrospective cohort study was to assess the way that daily physical activity and cancer staging can influence survival.