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. 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. 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. 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.

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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 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.

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.

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hemorrhagic degeneration; in some cases, calcifications and fibrous thickened capsules are seen at the periphery of the mass. On MRI, SPN is a well-defined lesion with hypoattenuation and well-demarcated characteristics of various pancreatic tumors that show hypointensity 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.

**ACKNOWLEDGMENT**

We gratefully acknowledge the assistance of Dr Akihiko Yoshida, Department of Diagnostic Pathology, National Cancer Center Hospital, Tokyo, for the confirmation of the histological diagnosis.

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**Authors’ declaration of conflict of interest:** The authors declare no conflict of interest.

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**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. Surgery remains the cornerstone of a multidisciplinary approach to cure this lethal disease. Pancreatectomy remains a challenging operation characterized by high morbidity. 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.

**REFERENCES**

1. Basturk O, Weigelt B, Adsay V, et al. Sclerosing epithelial mesenchymal neoplasm of the pancreas—a proposed new entity. Mod Pathol. 2020;33:456–467.

2. Anil G, Zhang J, Al Hamar NE, et al. Solid pseudopapillary neoplasm of the pancreas: CT imaging features and radiologic-pathologic correlation. Diagn Interv Radiol. 2017; 23:94–99.

3. Choi JY, Kim MJ, Kim JH, et al. Solid pseudopapillary tumor of the pancreas: typical and atypical manifestations. AJR Am J Roentgenol. 2006;187:W178–W186.

4. Hoshimoto S, Matsui J, Miyata R, et al. Anaplastic carcinoma of the pancreas: case report and literature review of reported cases in Japan. World J Gastroenterol. 2016;22:8631–8637.

5. Xie Y, Xiang Y, Zhang D, et al. Sarcomatoid carcinoma of the pancreas: a case report and literature review. Med Mol Rep. 2018; 18:4716–4724.

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

### 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                        | –        | 1/8                 |
| CD10                      | –        | 0/8                 |
| J-Catenin                 | –        | 0/8                 |
| Trypsin                   | –        | 0/8                 |
| TTF1                      | –        | 0/8                 |
| MCU4                      | –        | 0/8                 |
| Desmin and myogen         | –        | 0/8                 |
| INII (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                 |
| EWSRI gene fusion         | –        | 0/5                 |
| SYT-SSX fusion gene       | –        | 0/5                 |

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