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Clinical Image

A Pancreatic Tumor with Central Calcification

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2. Clinical Image

A 49-year-old woman visited our hospital with a history of epigastralgia, high fever and general malaise. Her temperature was 37.6 ℃. There was no significant past medical history. On physical examination, she had tenderness at the upper abdomen. Laboratory data were as follows: white blood cell count of 3000/μL, C-reactive protein of 6.77mg/dL, carcinoembryonic antigen of 1.9ng/mL, and carbohydrate antigen 19-9 of 33.8U/mL. Contrast-enhanced computed tomography scanning showed a poorly enhancing mass approximately 4.5cm in diameter at the head of the pancreas, which contained calcification in the middle of the mass (Figure 1). There were multiple lymphadenopathies around the stomach and mesenterium, peritoneal disseminations and ascites. Magnetic resonance imaging revealed a pancreatic mass with moderately high intensity on the T1 weighted image. Endoscopic ultrasound showed a hypoechoic mass which had central calcification at the head of the pancreas (Figure 2). Doppler examination demonstrated no blood flow in the mass. We performed endoscopic ultrasound fine needle aspiration, but histology revealed only fibrosis. To make a histological diagnosis, the patient underwent laparoscopic resection of white nodules around the omentum. Histologically, hematoxylin and eosin staining revealed cords of spindle cells and epithelioid cells within collagenous and fibrotic stroma (Figure 3). Immunohistochemistry revealed that the tumor was negative for AE1/AE3, desmin, S-100 protein, CD34 and STAT6, but was positive for MUC4 (Figure 4). Based on these findings, she was diagnosed with sclerosing epithelioid fibrosarcoma (SEF). She received doxorubicin therapy followed by eribulin therapy, but the tumor size enlarged 10 months after diagnosis. Then, she started to receive pazopanib therapy. SEF is a very rare variant of soft tissue sarcoma. SEF is clinically an aggressive tumor with a high rate of local and metastatic recurrence [1]. SEF mainly occurs in the extremities, limb girdles, trunk and head and neck area [1]. Its occurrence is exceedingly rare in visceral organs, with only one case reported in the liver, lower gastrointestinal tract, and pancreas, respectively [2]. Typical histology of SEF is a proliferation of neoplastic oval or round cells arranged in cords or a nest-like distribution against a collagen background with prominent features of sclerosis [3]. The immunohistochemical expression of MUC4 is seen in 78% of SEF. EWSR1-CREB3L1 gene fusions were distinguishing features between SEF and the morphologically similar low-grade fibromyxoid sarcoma [3]. Complete surgical resection, with or without adjuvant radiotherapy, remains the mainstay of therapy [1]. There is a limited response to chemotherapy and/or radiotherapy as sole treatment modalities [1]. In conclusion, we herein report a case of sclerosing epithelioid fibrosarcoma of the pancreas. Even though extremely rare, SEF should be considered as a differential diagnosis for a pancreatic tumor with central calcification.

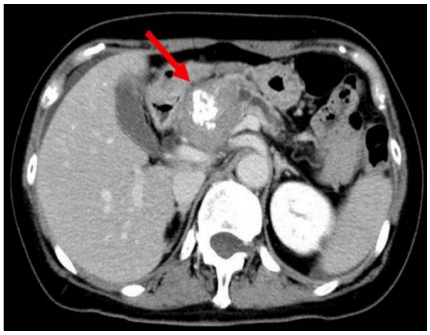

Figure 1: Contrast-enhanced computed tomography showed a poorly enhancing mass at the head of the pancreas, which contained central calcification. An arrow indicates calcification.

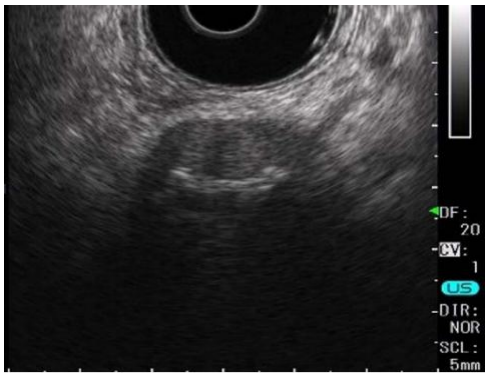

Figure 2: Endoscopic ultrasound showed a hypoechoic mass which had central calcification at the head of the pancreas.

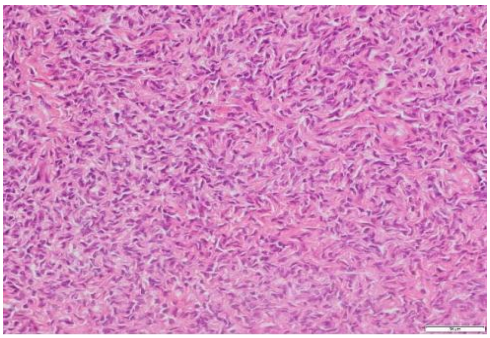

Figure 3: Histology of white nodules around the omentum revealed cords of spindle cells and epithelioid cells within collagenous and fibrotic stroma.

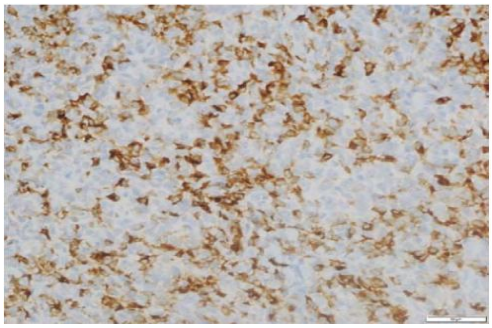

Figure 4: Immunohistochemical staining for MUC4 revealed that tumor was positive for MUC4.

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