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Case Report
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A Case Report of a Pancreatic Sarcoma Combined with Autoimmune Pancreatitis

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Abstract

Sarcomas, malignant tumors arousing from connective tissues, are rarely found within the pancreas, which is a mesenchymal supporting structure. This report describes a 77 year old male with a several month history of refractory epigastric pain. A low-attenuation lesion at the pancreatic head was identified on abdominal computed tomography. The Whipple procedure (pancreaticoduodenectomy) with pancreaticogastrostomy reconstruction was performed. Histology revealed high-grade sarcoma and lymphoplasmacytic sclerosing pancreatitis. Although the operative course of treatment was smooth, the patient died of cerebrovascular accident postoperatively.

Keywords: Pancreatic sarcoma; Autoimmune pancreatitis; Chronic pancreatitis; Pancreatic mass lesion

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Introduction

Sarcomas represent a relatively rare malignant entity, and primary sarcomas of the pancreas are particularly uncommon. Among pancreatic sarcomas, leiomyosarcomas are most commonly reported. This report describes a case of pancreatic sarcoma combined with autoimmune pancreatitis in an elderly man who was treated with surgical resection and steroids.

Case Presentation

A 77 year old male presented with acute-onset upper abdominal pain. The patient had hypertension, type II diabetes mellitus, and peptic ulcer disease, and he had previously been treated conservatively for acute pancreatitis. At that time, abdominal ultrasound revealed a "shadow" in the pancreas, and a biopsy *via* Endoscopic Retrograde Cholangiopancreaticography (ERCP) was unsuccessful. The patient refused surgical intervention initially and was discharged after the symptoms of pancreatitis subsided. Since that time, the patient suffered from several relapses over the following months. Since the recurrent abdominal sharp pain bothered him so much that he change his mind to the surgical intervention. Laboratory studies revealed a normal white blood cell count and liver function test, but elevated amylase and lipase (793 U/L and 1634 U/L respectively). The IgG4 was 169 mg/dL, and the tumor markers CA199 and carcinoembryonic antigen were within normal limits. Abdominal Computed Tomography (CT) disclosed a 1.5 cm low-attenuation lesion at the pancreatic head without dilatation of the main pancreatic duct (Figure 1). Surgical resection was elected under the impression of refractory chronic pancreatitis with a pancreatic head mass lesion. An ulcerative tumor over the pancreatic head with compression of the common bile duct was identified. The Whipple procedure with pancreaticogastrostomy reconstruction was performed. Gross examination identified one tumor measuring 1.2 cm × 1.0 cm × 0.8 cm in the head of the pancreas. Microscopic examination revealed epithelioid to spindle tumor cells with pleomorphic nuclei and prominent nucleoli arranged in solid pattern (Figure 2A and 2B). The mitotic count was

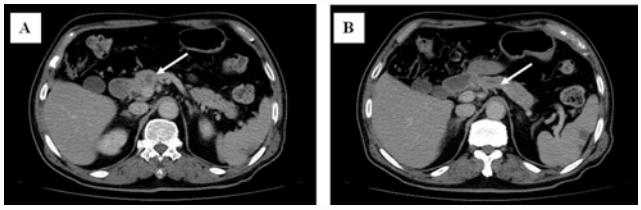

Figure 1: A) Abdominal computed tomographic scan revealing a 1.5 cm low-attenuation lesion (arrow) at pancreatic head. B) The main pancreatic (arrow) duct was not dilated.

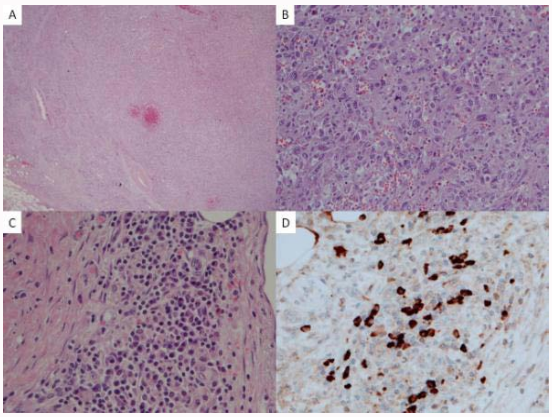

Figure 2: A) The tumor (right) involved in the pancreas. B) Epithelioid to spindle tumor cells with pleomorphic and prominent nucleoli. C) Chronic lymphoplasmacytic cell infiltration and fibrosis within the pancreas. D) Some plasma cells highlighted by IgG4 stain were noted.

22/10 high-power fields, and tumor necrosis was present. The margins of the pancreas, common bile duct, and duodenum were tumor-free, and the lymph nodes were negative for metastasis. Histology also revealed pancreatic lobules with chronic lymphoplasmacytic cell infiltration and fibrosis (Figure 2C). Some plasma cells were highlighted by IgG4 stain (Figure 2D), and immunohistochemic staining demonstrated the tumor cells were focally positive for CD31 (Figure 3A) and smooth muscle actin, but negative for other markers such as cytokeratin, S-100, CD-34, CD-117, and HMB-45 (Figure 3B-3D). The final pathologic diagnoses were high-grade sarcoma and lymphoplasmacytic sclerosing pancreatitis. It could not, however, be determined whether the malignant tumor was either a primary or secondary neoplasm of the pancreas due to well-defined tumor border with many tumor emboli in the vessel lumens. Although the surgery was successful, the patient died of a cerebrovascular accident postoperatively.

Discussion

Primary sarcomas of the pancreas are exceedingly rare. Baylor et al. [1] reported an incidence of 0.1% of pancreatic sarcoma after reviewing 5,000 cases of pancreatic cancer. Among pancreatic sarcomas, leiomyosarcomas tend to occur relatively frequently, whereas metastatic sarcomas of the pancreas are uncommon [2]. Distant metastases occur in almost 25% to 30% of patients with soft-tissue sarcomas as a disseminated disease, with the vast majority preferentially metastasizing to the lungs (which is the first site of recurrence in 80% of patients) [3,4]. Cerebellar metastases and pancreatic metastases in the absence of pulmonary metastases are exceptional events in the natural history of mesenchymal neoplasms. The patient described in this report had no evidence of sarcoma other

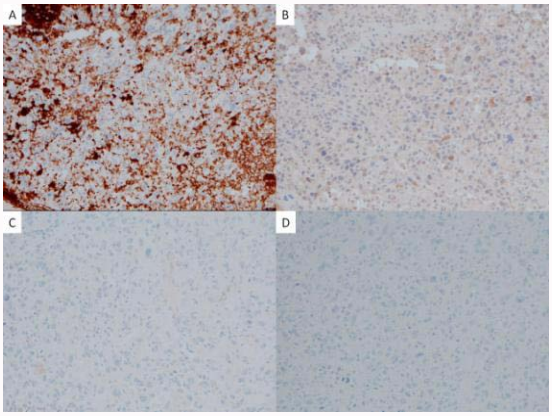

Figure 3: A) Immunohistochemistry of tumor cells showed positive staining for CD31. B) Negative staining for cytokeratin. C) Negative staining for CD34. D) Negative staining for HMB45.

than the pancreatic head lesion, and pathology could not determine whether the malignant tumor was the either a primary or secondary neoplasm of the pancreas due to the presence of a well-defined tumor border with many tumor emboli in the vessel lumens. Pancreatic mass lesions are typically identified using various imaging studies including ultrasound, CT, magnetic resonance imaging, and ERCP [5]. Diagnostic biopsy of a suspected pancreatic malignancy is only indicated for treatment planning if there is systemic spread of the disease, local evidence of an unresectable tumor, if the patient is unfit for surgery, or if neoadjuvant treatment is being considered. Preoperative biopsy of potentially resectable pancreatic tumors is not generally advisable because a benign sample does not exclude the presence of a neighboring malignancy [6,7]. Surgical resection is the only possible cure for pancreatic malignancies [8]. In this case, a history of pancreatitis and elevated serum IgG4 were suggestive of autoimmune pancreatitis; however, the abdominal CT did not reveal evidence of diffuse enlargement of the pancreas or dilatation of the main pancreatic duct; however, a low-attenuation lesion at the pancreatic head was identified. It is the responsibility of the physician to judge the probability of autoimmune pancreatitis and to weigh the benefits of medical treatment and risks associated with delayed therapy when malignancy is present. In this case, our surgical team thoroughly discussed the possible preoperative diagnoses and the corresponding treatment choices with the patient and his family. Surgical intervention was elected in consideration of both the intolerable abdominal pain due to refractory chronic pancreatitis and the suspicion of malignancy, which were both demonstrated pathologically.

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