

Pancreatic Undifferentiated Pleomorphic Sarcoma Mimicking a Gastric Gastrointestinal Stromal Tumor

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Received: April 20, 2021

Accepted: May 17, 2021

Published: May 18, 2021

Citation: Ehrhart M, Hanson JA, Boehler C, Runde RE, Thompson WM. 2021. Pancreatic Undifferentiated Pleomorphic Sarcoma Mimicking a Gastric Gastrointestinal Stromal Tumor. *J Med Imaging Case Rep* 5(1): 22-24.

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Abstract

Undifferentiated pleomorphic sarcoma (UPS), previously known as malignant fibrous histiocytoma, is the most common type of soft tissue sarcoma. Commonly seen in the extremities, UPS is an uncommon primary tumor of the GI tract. We report a case of an elderly female with a known renal neoplasm who incidentally was found to have a malignant lesion in the epigastrium. Given exophytic character abutting the stomach and pancreas, the mass initially thought to represent a gastrointestinal stromal tumor arising from the stomach. Biopsy of the mass showed atypical and pleomorphic malignant cells of unknown differentiation and was nonreactive with various immunohistochemical markers including both stromal and epithelial cell lines. A poorly differentiated sarcomatoid neoplasm of the pancreas was diagnosed.

Keywords

Pleomorphic sarcoma, GIST, Pancreas, Histopathology

Case Report

A 74-year-old female was undergoing active surveillance for solid mass in the right kidney compatible with renal cell carcinoma. She had multiple comorbidities and was a poor surgical candidate. She was hospitalized for gross hematuria and severe anemia (hemoglobin 6.5 g/dL, hematocrit 23%) requiring blood transfusion. During that admission, she had a contrast-enhanced CT abdomen and pelvis which showed the known renal mass, as well as a large mass in the epigastrium which abutted both the pancreas and stomach (Figure 1). Ultrasound (US) demonstrated a mixed echogenicity, ovoid mass with mildly lobulated margins situated in the epigastric region, measuring approximately 13.1 cm in longest dimension (Figure 2).

To better assess the mass, the patient underwent a multiphasic abdominal CT. Unfortunately, this study did not reveal diagnostic enhancement characteristics, and biopsy was planned; a gastric gastrointestinal stromal tumor (GIST) was the favored diagnosis. A transcutaneous US-guided biopsy using an 18 gauge Core Biopsy needle passed through 17 gauge needle guide of the epigastric mass was performed. Pathology revealed atypical and pleomorphic cells with vacuolated cytoplasm (Figure 3). The reported cytomorphology was not characteristic of epithelial neoplasms and tissue was nonreactive for numerous epithelial markers. A diagnosis of poorly differentiated sarcomatoid neoplasm was favored.

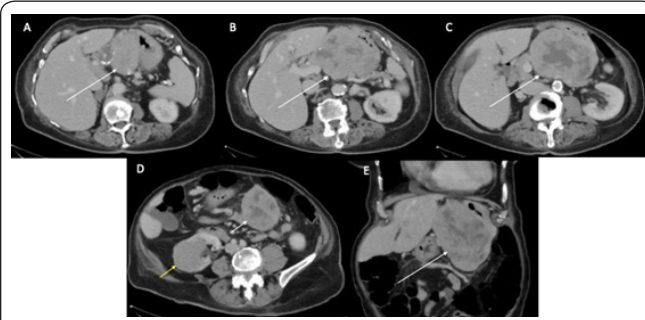


Figure 1: Multiple contrast enhanced axial (A-D) and coronal (E) CT images through the epigastrium. There is heterogenous density retrogastric mass (white arrows, A-E) measuring 12.5 cm with areas of increased and decreased attenuation. Lesion exhibits mass effect on various structures including the stomach, pancreas, small bowel, colon, and gastroduodenal artery. Patient's known presumed renal cell carcinoma in the right kidney is also seen (yellow arrow, D).

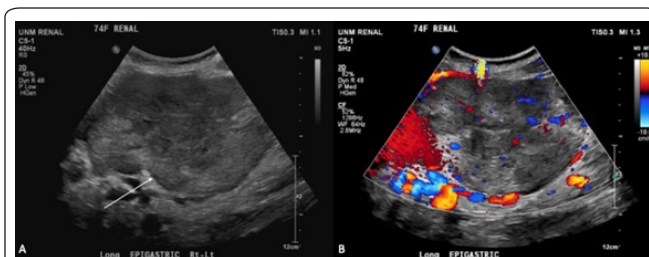


Figure 2: Longitudinal greyscale (A) and color doppler (B) ultrasound images. There is a mixed echogenicity ovoid mass with mildly lobulated margins in the epigastrium (arrow, A). The lesion exhibits internal vascularity (B).

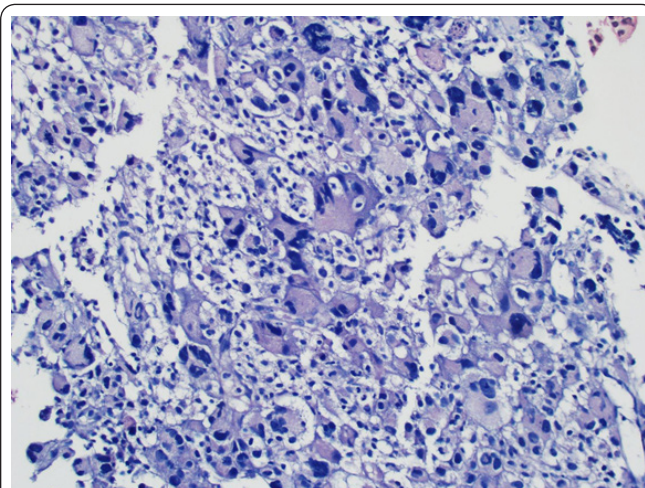


Figure 3: The tumor consists of highly atypical rhabdoid to epithelioid cells. Bizarre tumor giant cells are also seen. There is no specific line of morphologic differentiation and immunohistochemical stains also failed to reveal a specific diagnosis. The tumor was ultimately classified as an undifferentiated sarcomatoid neoplasm.

Subsequently, she again presented to the emergency department with extreme fatigue and weight loss, and her skilled nursing facility was unable to give her the level of care she required. Her condition continued to deteriorate rapidly. The patient was seen by surgical oncology but, based on her continued poor performance status, she was not a surgical candidate. After discussion with patient and her family, a

decision was reached to move forward with palliative care and hospice.

Discussion

Undifferentiated pleomorphic sarcoma (UPS), previously known as malignant fibrous sarcoma (MFH), is the most common type of soft tissue sarcoma [1, 2]. These types of tumors occur commonly in the soft tissues of the extremities, less likely in the retroperitoneum, and are infrequently seen elsewhere. UPS has only rarely been reported as a primary pancreatic malignancy, and is one of the rarest pancreatic neoplasms of non-epithelial origin [1, 3].

UPS commonly grow relatively large in size. Pancreatic UPS usually presents clinically with epigastric pain, nausea and vomiting, weight loss, and, depending on location, jaundice [1, 3]; these symptoms are also common of other pancreatic neoplasms.

Sarcomas as primary pancreatic neoplasms represent only 0.1% of all pancreatic neoplasms. It should be noted that UPS are far more likely to arise from mesenchymal elements in the retroperitoneum and grow into the pancreas than they are to arise as a sarcoma of actual pancreatic origin [3]. Few pancreas UPS have been reported in the literature, with a 2018 case report quoting only 21 cases [1]. Of those cases, mean age at diagnosis was 55, but ranged from 22-77 years. While males may be affected more than females [3], further studies are needed.

No specific imaging characteristics for UPS have been identified, and thus diagnosis relies on biopsy with pathologic evaluation. Biopsied tissue is initially assessed by microscopy, though immunohistochemistry is ultimately required for final diagnosis. In the presented case, microscopic evaluation showed that the tumor was comprised of highly atypical rhabdoid to epithelioid cells, with bizarre tumor giant cells also seen. It demonstrated both a cytomorphology and immunophenotype that was not characteristic of any specific differentiation. The pathologic specimen was negative for numerous epithelial markers (pancytokeratin, CK7, 20, and 19) as well as GIST markers (DOG-1, CD117) or muscle (SMA, Desmin) components, ruling out leiomyosarcoma. Additionally, immunohistochemistry (IHC) was negative for S100, HMB-45, and MDM2 ruling out MPNST, melanoma, and dedifferentiated liposarcoma. A diagnosis of poorly differentiated sarcomatoid neoplasm was therefore rendered.

Imaging findings in pancreatic UPS are nonspecific, with an enhancing solid mass seen on CT and a hypoechoic mass with internal vascularity seen on ultrasound. MRI exhibits similarly nonspecific findings, showing a solid mass exhibiting hypointensity on T1 weighted images with associated gadolinium enhancement. These masses usually exhibit FDG activity. Secondary features, including associated pancreatic and biliary ductal dilation, mass effect on other structures, or evidence of local invasion or metastasis are important features for the radiologist to note.

Typical differential considerations for pancreatic UPS include pancreatic ductal adenocarcinoma, lymphoma, pancreat-

ic neuroendocrine tumor, or less commonly intraductal papillary mucinous neoplasm (IPMN). In this case, GIST was also considered due to the exophytic character of the mass with mass effect on the stomach. It can sometimes be difficult to determine organ of origin of large masses only by imaging, as in the current case.

Ductal adenocarcinoma is the most common pancreatic malignancy, and commonly arise from the pancreatic head. Tumors in this location may cause a “double duct sign”, where there is dilation of the common bile and pancreatic ducts, best seen on US or MRCP. Pancreatic adenocarcinomas are hypovascular [4], and sometimes can be difficult to see on single-phase contrasted exams. Primary pancreatic lymphoma is rare, with pancreatic extension of a retroperitoneal lymphoma far more common (similar to pancreatic vs retroperitoneal UPS) [4]. There are two morphologic patterns of pancreatic lymphoma, including a focal form which presents as a solid mass typically in the pancreatic head, and a diffuse form which presents as an infiltrative lesion that may mimic pancreatitis. The focal form of pancreatic lymphoma is less likely to cause pancreatic duct dilation than pancreatic adenocarcinoma, and, in lymphoma, common bile duct dilation is more common than pancreatic ductal dilation. It will exhibit only faint contrast enhancement.

Neuroendocrine tumors typically exhibit hypervascularity, and thus more avidly enhance than other pancreatic neoplasms [4]. They do not reliably exhibit FDG activity, whereas they exhibit intense uptake on octreotide scan or Ga-68 DOTATATE PET/CT due to the presence of somatostatin receptors. In contrast, IPMNs present as cystic masses and may be considered in a differential diagnosis list for a centrally necrotic UPS. They have fluid density on CT, are anechoic on ultrasound (noting smaller IPMN may not be seen), and hyperintensity on T2 weighted images on MRI. Main duct communication is a key prognostic indicator, as main duct type IPMNs have a much higher malignant potential than

do branch duct type (i.e. those without main duct communication) [5]. FDG activity is somewhat variable, with mild FDG activity in IPMNs corresponding to a more malignant prognosis [6].

Due to its rarity, treatment of pancreatic UPS has been inadequately studied. While most cases go to surgery, there have been no studies suggesting one treatment algorithm (i.e. neoadjuvant or adjuvant chemotherapy, with or without radiation) over another, with current treatments likely dictated on an individual basis by tumor size and location, patient’s age, and functional status. The diagnosis of pancreatic UPS carries an overall poor prognosis, though has been inadequately studied [1, 2].

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