Review

Endoscopic Ultrasound

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Comments and illustrations of the European Federation of Societies for Ultrasound in Medicine guidelines: Rare pancreatic tumors, ultrasound and contrast-enhanced ultrasound features—Malignant mesenchymal tumors

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ABSTRACT

Rare malignant mesenchymal pancreatic tumors are systematized and reported in this review. The focus is on the appearance on imaging. The present overview summarizes the data and shows that not every pancreatic tumor corresponds to the most common entities of ductal adenocarcinoma or neuroendocrine tumor.

Key words: Malignant mesenchymal pancreatic tumors; Pancreatic EGIST; Imaging

INTRODUCTION

The World Federation for Ultrasound in Medicine and Biology has published guidelines on the use of contrast-enhanced ultrasound (CEUS) for the evaluation of focal liver lesions[1–5] and European Federation of Societies for Ultrasound in Medicine for the evaluation of nonhepatic indications.[6,7] Improved detection and characterization of common focal pancreatic lesions such as ductal adenocarcinoma, neuroendocrine tumors, and pancreatic metastases are the main topics of these guidelines. Mesenchymal neoplasms of the pancreas are rare.[8–10] Primary mesenchymal tumors account for approximately 0.3% to 0.5% of all histologically confirmed pancreatic tumors.[11,12] One-third of these are malignant mesenchymal tumors.[11,12] The present work focuses on the malignant mesenchymal tumors of the pancreas. Mesenchymal tumors occurring within the pancreas are listed in Table 1. It is important to distinguish malignant mesenchymal pancreatic tumors from anaplastic carcinomas and retroperitoneal tumors infiltrating the pancreatic tissue.[108] Mesenchymal tumors of the pancreas rarely demonstrate the typical imaging findings of ductal adenocarcinomas. Sarcomas are usually very large at diagnosis.[11] The most common primary malignant mesenchymal tumors (sarcomas) of the pancreas are leiomyosarcomas, Ewing sarcomas (ESs)/primitive neuroectodermal ESs/primitive neuroectodermal tumors (PNETs), and undifferentiated/unclassified sarcomas (also known as malignant fibrous histiocytomas).[11] Kim et al.[11] reported a 0.3% rate of primary pancreatic mesenchymal tumors in a single-center study of 7129 surgically resected or histologically confirmed pancreatic tumors, of which 30% were malignant. These were 3 cases of undifferentiated/unclassified sarcoma, 1 case of leiomyosarcoma, 1 case of ES/PNET, and 1 case of atypical lipomatous tumor/well-differentiated liposarcoma.[11] Zhang et al.[12] found 10 primary mesenchymal tumors (0.5%) among 1944 resected pancreatic tumors. Of these, 3 (33%) were primary malignant mesenchymal tumors. There were 1 case each of malignant gastrointestinal stromal tumor (GIST), malignant solitary fibrous tumor, and undifferentiated pleomorphic sarcoma.[12] Solitary fibrous tumor is usually benign, and approximately 12% take a malignant course. Preoperative diagnoses of all mesenchymal tumors in the study by Kim et al.[11] and Zhang et al.[12] were as follows: ductal adenocarcinoma, neuroendocrine tumor, mucinous cystadenoma, serous microcystic adenoma, Castleman disease, chronic pancreatitis, solid pseudopapillary tumor, and invasive intraductal papillary mucinous neoplasm. The diagnosis of mesenchymal tumor was made exclusively postoperatively.[12]

The aim of the present review is to demonstrate the complex presentation of rare malignant mesenchymal tumors of the pancreas apart from ductal adenocarcinoma and typical neuroendocrine tumors. The current published articles with criterion-standard histology

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Table 1

Benign/intermediate and malignant mesenchymal pancreatic tumors.

Benign or intermediate mesenchymal pancreatic tumor	Malignant mesenchymal pancreatic tumors
Lipoma[13–18]	Leiomyosarcoma[11,12,19–25]
Schwannoma[11,12,26]	Ewing sarcomas/primitive neuroectodermal Ewing sarcomas/PNETs[11,27–33]
Inflammatory myofibroblastic tumor (inflammatory pseudotumor)[11,34–45]	Undifferentiated/unclassified sarcomas (malignant fibrous histiocytoma)[11,12,46]
Solitary fibrous tumor[11,12,47–56]	Liposarcoma[11,57–59]
Hamartoma[11,60–66]	AS[67–71]
PEComa[72–77]	Fibrosarcoma[78–81]
Angiomyolipoma[11,82]	Kaposi sarcoma[83,84]
Ganglioneuroma[12,85–89]	Rhabdomyosarcoma[90,91]
Fibromatosis (desmoid tumor)[11,12,92–95]	EGIST[12,96–99]
Hemangioma[11,100–107]	
Myofibroblastoma[12]	

PEComa: perivascular epithelioid cell tumor; AS: angiosarcoma; PNETs: primitive neuroectodermal tumors; GIST: gastrointestinal stromal tumor.

include malignant pancreatic sarcomas: extragastrointestinal stromal tumors (EGISTs), leiomyosarcoma, ES, rhabdomyosarcoma, Kaposi sarcoma, angiosarcoma (AS), liposarcoma, sclerosing epithelioid fibrosarcoma, and kaposiform hemangioendothelioma. For some of the very rare entities, the engagement of a variety of authors can demonstrate the appearance on EUS with examples.

IMAGING AND DIAGNOSIS OF MALIGNANT MESENCHYMAL PANCREATIC TUMORS

Extragastrointestinal stromal tumor

Gastrointestinal stromal tumors are rare tumors, corresponding to less than 1% of tumors in the gastrointestinal tract. Gastrointestinal stromal tumors are the most common mesenchymal tumors in the gastrointestinal tract. Mostly they are located in the stomach or intestine, more rarely in the colorectum.[109] Less than 5% of GISTs are located outside the gastrointestinal tract, unrelated to its wall. These are referred to as EGISTs. These are located in the omentum, mesentery, and retroperitoneum and again rarely in less than 5%, in the pancreas.[110] Histopathologically, GISTs can be classified as spindle-cell (70%), epithelioid (20%), or mixed (10%) type. Most pancreatic EGISTs correspond to the spindle-cell–shaped type.[110] Approximately 95% of GISTs and EGISTs are CD117-positive. In addition, GISTs may stain positive for CD34 (60%–70%), heavy caldesmon (80%), smooth muscle actin (SMA) (30%–40%), S100 (5%), and desmin (<5%).[111] Liu et al.[110] researched 45 pancreatic EGISTs and compared them with 297 gastric GISTs. The most common location was the pancreatic head (38%). They are often incidental findings, and patients typically present with nonspecific symptoms. Nevertheless, the tumors were usually large at initial diagnosis with a diameter of greater than 5 cm in 74% of cases. Fifty-six percent were cystic or had mixed imaging features; 44% were solid. Mitotic index was the only risk factor for disease-free survival (DFS) of pancreatic GISTs. More than 42% had a mitotic index greater than 5 of 50 per high-power field. The 5-year DFS and disease-specific survival rates were 66.1% and 95.8%, respectively. The DFS of pancreatic GISTs was lower than that of gastric GISTs.[110] All solid pancreatic EGISTs were of the spindle-cell type, whereas only half of the cystic and mixed type. Sonography plays an important role in the primary diagnosis of pancreatic EGISTs.[97] Preoperative diagnosis has been predominantly by computed tomography (CT). The use of EUS was reported only occasionally.[97,111,112] Usually, the definite diagnosis was made by surgical resection and histological workup.[98,113–117] Careful descriptions exist for the endosonographic appearance of GISTs. Those with low risk were characterized by a smooth contour, low echogenicity, and distinct vascularity. Those with higher risk may be hypoechoic and heterogeneous with necrosis, cystic spaces, echogenic foci, and/or calcifications and are heterogeneously hypervascularized except for necrosis. The contour may be irregular.[96,118–121] EUS-guided sampling with either fine-needle aspiration (FNA) or fine-needle biopsy (FNB) is recommended for the diagnosis of GIST and other subepithelial lesions in the stomach.[122–124] In a meta-analysis of 10 studies comparing EUS-FNA and EUS-FNB, EUS-FNB outperformed EUS-FNA in all diagnostic criteria, including adequate specimen collection rate, optimal histologic core collection rate, diagnostic accuracy, and number of passes required to obtain diagnostic specimens.[125] Because of the limited data available, it is unclear whether these data can also be applied to pancreatic EGISTs. Endosonographically, pancreatic EGISTs appear in the case reports as smoothly circumscribed hypoechoic lesions, occasionally with cystic portions.[99,111,126] Cases with EUS-FNB of pancreatic EGISTs have been described.[111,126–128] In a single case report of a pancreatic EGIST at the pancreatic body, CEH-EUS with SonoVue demonstrated hyperenhancement of the tumor in the arterial phase.[129] A pancreatic GIST with complex endosonographic diagnosis is demonstrated in Figure 1.

Leiomyosarcoma

Leiomyosarcomas are rare, malignant tumors arising from smooth muscle cells. As such, it can involve any organ where smooth muscle is present, with the most frequent localization being the uterus[130] followed by the retroperitoneum.[131] Both primary pancreatic leiomyosarcoma and metastatic leiomyosarcoma of the pancreas are extremely rare.[20,132] It is postulated that primary pancreatic leiomyosarcoma originates from the smooth muscle located within the pancreatic ducts or the wall of intrapancreatic vessels.[133,134] Histologically, the tumor is characterized by intersecting groups of spindle cells with a typical appearance of elongated and blunt-ended nuclei.[11] Aleshawi et al.[23] conducted a comprehensive review of 87 cases of primary pancreatic leiomyosarcomas that have been reported in the literature. They demonstrated that there was no specific gender predisposition, and the mean age at diagnosis was 53.8 years. Ethnicity may play a role, and more patients of East Asian origin have been reported than patients from other geographical origins. Because of its rarity, the optimum treatment regimen remains in doubt, but surgical resection remains the mainstay of treatment, and doxorubicin-based chemotherapy has been used in

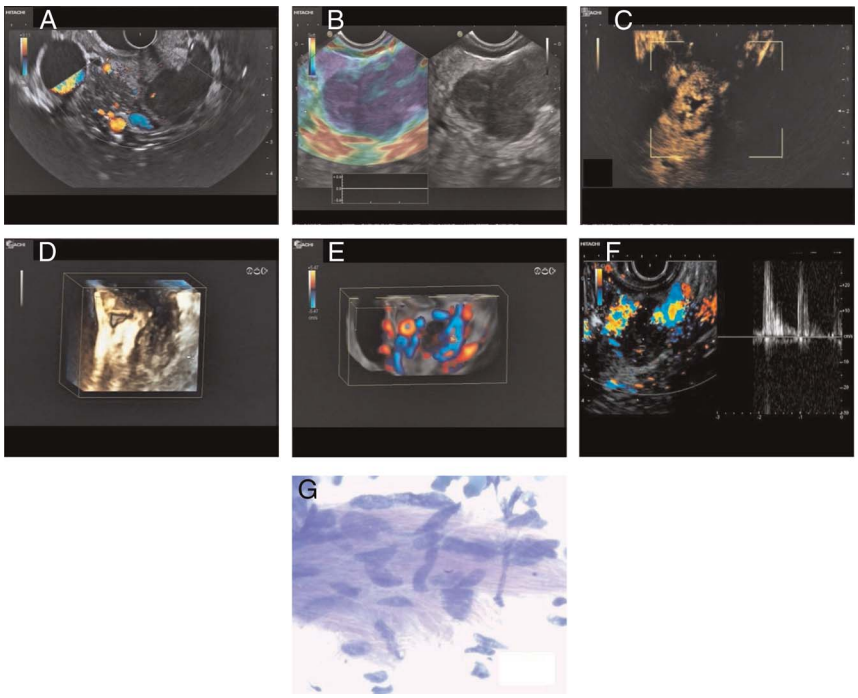

Figure 1. Extragastrointestinal stromal tumor at the pancreatic head. Display of the GIST tumor on the right side and the normal pancreatic head on the left side. The scan was taken from the descending duodenum. Unenhanced Doppler color display is overlaying the gray scale scan (A). The hypoechoic tumor is visible with the result of the elastographic scanning on the left side. The tumor appears blue that means harder than the surrounding tissue (B). Contrast-enhanced harmonic EUS with 4,8 mL SonoVue. The typical highly vascularized tumor and the central necrosis are visible (C). The tumor is displayed using 3D reconstruction after injection of 4,8 mL SonoVue in CEH-EUS (D). The same tumor in a 3D reconstruction of a color Doppler mode overlying gray-scale ultrasound. The surrounding vessels are clearly visible (E). Pulsed-Wave (PW)-Doppler mode after injecting of 4.8 mL SonoVue in high-mechanical-index EUS, only high-resistance arterial vessels can be detected within the tumor (F). Cytological result after endosonographic fine-needle puncture of the tumor. Typical mesenchymal tumor cells are displayed (H).

palliative settings. There are no imaging features that are specific to primary pancreatic leiomyosarcoma. The radiological descriptions of these tumors in the literature have shown variable attenuation and enhancement.[21,23,135] Of the 87 cases reported in the literature, cystic degeneration was observed in 32 patients. In the Pancreatic Multicenter Ultrasound Study, leiomyosarcoma is described as homogenous or heterogeneous hypervascular.[136] The mainstay of diagnosis in primary pancreatic leiomyosarcoma is acquisition of tissue for histological analysis. In addition, typical imaging features and treatment recommendations cannot be given [Figure 2].

Ewing sarcoma

Primitive neuroectodermal tumors (previously referred to as peripheral neuroepithelioma) are rare malignant tumors with various degrees of differentiation belonging to the Ewing family of

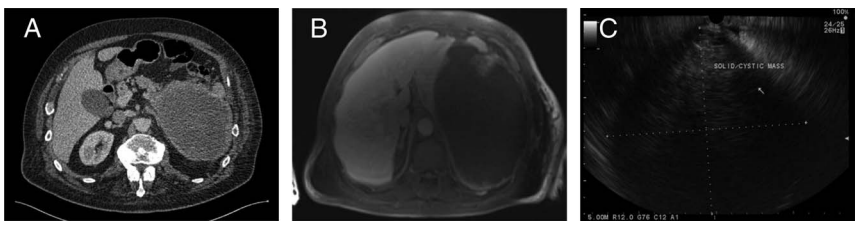

Figure 2. A 76-year-old man presented after a fall and a trauma series. Computed tomography scan (A) demonstrated a large, complex cystic mass between the pancreatic tail and the spleen. Initially felt to be a hemorrhagic lesion following the recent trauma, the complexity of the lesion prompted MRI evaluation. The MRI (B) demonstrated the lesion to arise from an expanded pancreatic tail. The fluid contents of the lesion appeared of intermediate high signal on T1 suggestive of hemorrhagic or proteinaceous characteristic and with more solid components within the peripheries of the lesion. Percutaneous ultrasound-guided drainage of the lesion yielded serous fluid for analysis, although cytological results did not demonstrate any malignancy. The patient underwent an EUS (C), which demonstrated a large mixed cystic/solid lesion and FNB of the solid components confirmed the diagnosis of high-grade leiomyosarcoma.

sarcomas.[137] Ewing sarcoma is a rare, small, round cell bone tumor, of presumed neuroectodermal origin, with a predilection for patients in the first or second decade of life.[138,139] It is the second commonest primary malignant bone neoplasm in children and adolescents, only preceded by osteosarcomas. Cytogenetically, ES is typically characterized by chromosomal translocation at 11:22 (q24:q12) in more than 80% of cases.[140] Although the majority of ESs present as localized disease in the axial or appendicular skeleton,[141] extraosseus soft tissue manifestations of ES have been described. Pancreatic ES is extremely rare, with only 25 reported cases in the literature until 2020.[137] Few cases are additionally reported.[142–145] These are usually very large tumors, but otherwise do not exhibit typical imaging features, and treatment recommendations cannot be given. Recommended treatment follows guidelines with a systemic approach. Local treatment is rarely necessary and might include endoscopic drainage procedures [Figure 3].

Rhabdomyosarcoma

Rhabdomyosarcoma is a rare, malignant tumor of mesenchymal origin arising from skeletal muscle cells.[146] It occurs more commonly in childhood and adolescence, and development of rhabdomyosarcoma in adulthood is rare.[91] The most commonly affected primary site at diagnosis is the chest/abdomen/pelvis, followed by the extremities, genitourinary tract, and the head and neck.[90] Rhabdomyosarcoma in adult patients is aggressive and typically metastasizes, with typical sites being the lungs, bones, omentum, and lymph nodes.[147,148] Pancreatic rhabdomyosarcoma is extremely rare, with most cases as a result of metastases from another primary site, although primary pancreatic rhabdomyosarcoma has been reported.[149] Typical imaging features and treatment recommendations cannot be given [Figure 4].

Kaposi sarcoma

Kaposi sarcoma is an angioproliferative low-grade neoplasm associated with human herpesvirus 8 (HHV-8). The clinical picture is named after the Hungarian dermatologist Moritz Kaposi, who first described it in 1872. There are 4 variants: the "classic" form is characterized by an indolent cutaneous course; the "African endemic" form of the disease shows lymphadenopathy and is usually fatal; the iatrogenic/posttransplant form is due to reactivation of HHV-8 in the context of drug immunosuppression; and the fourth variant develops in the context of HIV infection.[150] With the use of highly active antiretroviral therapy in HIV-infected patients, visceral manifestations have decreased.[151] The typical sites that are affected include the skin, mucous membranes, lymph nodes, and, among the visceral organs, the lungs and the gastrointestinal tract. Pancreatic manifestation is rare and usually occurs as one of the organ manifestations in extensive multifocal manifestations.[84] In an autopsy study of patients with HIV infection and Kaposi sarcoma (University California), visceral involvement was common.[152] The gastrointestinal tract was frequently affected with involvement of the pancreas (30%), spleen (40%), and liver (40%).[152] There was no direct correlation between manifestations in the gastrointestinal tract and the pancreas and other parenchymal organs. The parenchymal lesions corresponded to small nodules, although 1 patient had a 6-cm infiltrative retroperitoneal process involving the pancreas and infiltration of the surrounding organs and of the mesentery by small, firm purple masses.[152] In another autopsy study of patients with HIV infection and Kaposi sarcoma, the most common visceral involvement was observed in the lung (37%), the gastrointestinal tract (50%), and in lymph nodes (50%). Only 1 of 25 patients had pancreatic manifestations.[153] In another case, an HIV-positive patient with Kaposi sarcoma with skin, lymph node, and lung manifestations and HHV-8 developed occlusive jaundice as a result of an extensive infiltrative process in the area of the pancreatic head with vascular involvement. There was a double-duct sign with distal stenosis of the common bile duct and pancreatic duct. The findings exhibited similar characteristics as a ductal adenocarcinoma of the pancreas. An endoprosthesis was implanted into the bile duct. No sampling for cytology or histology was performed, but a mutation analysis of KRAS and HHV-8 in the pancreatic secretion was performed. KRAS mutation was negative and HHV-8 positive. In the overall context, the authors concluded from the constellation of findings that there was a pancreatic manifestation of Kaposi sarcoma. The patient was treated with paclitaxel again and started on additional antiviral therapy with foscarnet to eliminate HHV-8. Antiretroviral therapy was started with a triple regimen of indinavir, nevirapine, and stavudine. Under this regimen, the findings regressed, so that endoprosthesis therapy was no longer necessary.[83] In other cases, the pancreatic manifestation is described as a local infiltrative process that resembled pancreatic carcinoma in appearance. In the context of already existing other multilocular Kaposi manifestations in immunosuppressed patients with HHV-8 infection, such findings suggest that another Kaposi sarcoma manifestation should be suspected.[154,155]

Angiosarcoma

Angiosarcomas are highly malignant neoplasms with endothelial cell differentiation originating from blood or lymphatic vessels. Angiosarcomas account for 1% of all soft tissue sarcomas.[108] Localization to the pancreas is extremely rare. By 2022, a total of 10 cases had been published in the English-language literature.[67–71] Nguyen et al.[70] described the simultaneous diagnosis of ductal adenocarcinoma with AS of the pancreas in surgical histology.

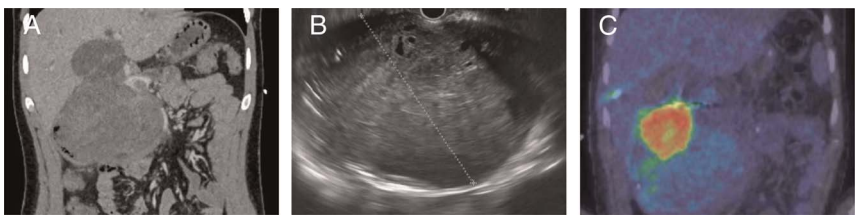

Figure 3. A 28-year-old man presented with general malaise, weight loss, and jaundice. A large palpable mass was discernible in the epigastrium on abdominal examination. Computed tomography scan (A) demonstrated a 12-cm lobulated mass arising from the head of the pancreas causing biliary and pancreatic duct obstruction. EUS (B) demonstrated a heterogeneous soft tissue mass lesion with some cystic foci, and FNB was performed. PET-CT (C) showed intense tracer (^{18}F-FDG) uptake.

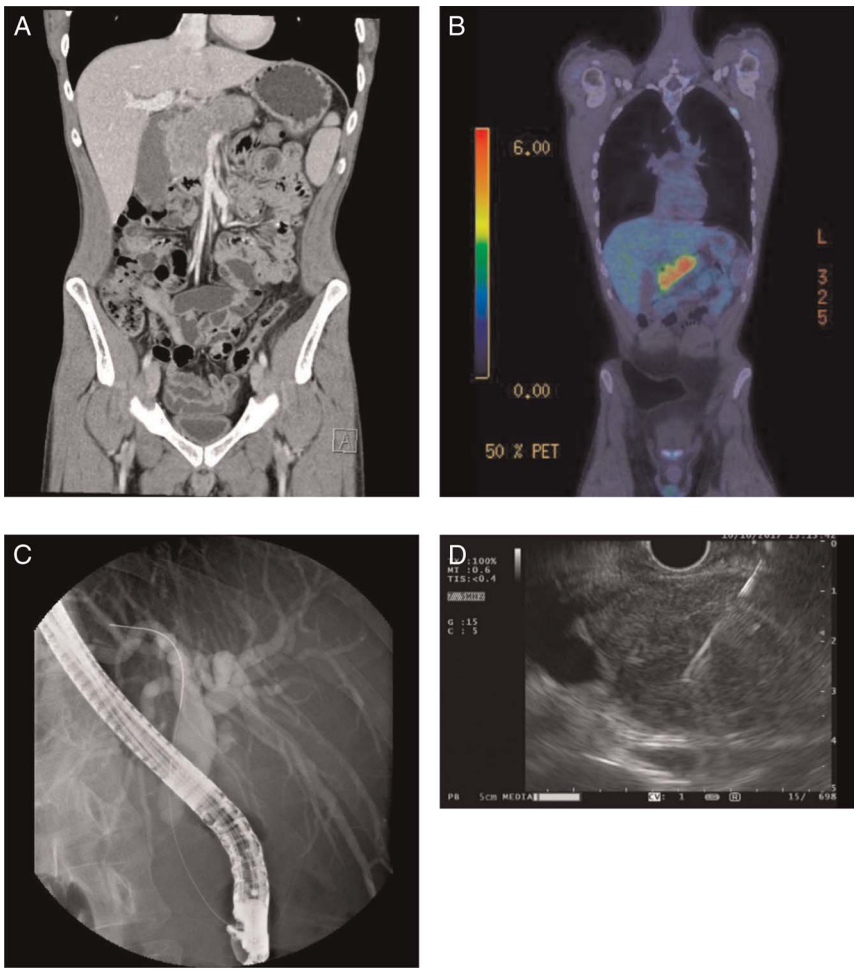

Figure 4. A 30-year-old man who was diagnosed with rhabdomyosarcoma of the skull base with bone and lymph node metastases 2 years previously presented with jaundice. He underwent a CT (A) scan demonstrating a diffusely abnormal pancreas with biliary obstruction with no focal pancreatic lesions seen and no adjacent peripancreatic fluid or inflammatory stranding. A PET-CT (B) showed diffuse ^{18}F-FDG uptake throughout the pancreatic parenchyma. An ERCP and biliary stenting (C) were performed, and cholangiogram demonstrated stricturing of the entire intrapancreatic portion of the extrahepatic common bile duct. EUS-FNB (D) was performed concurrently with ERCP and demonstrated the pancreas gland to be diffusely abnormal and expanded in its entirety with multiple hyperechoic foci. Histology confirmed diagnosis of metastatic rhabdomyosarcoma to the pancreas.

Angiosarcoma can be localized in all areas of the pancreas. The clinical symptoms can be nonspecific. Abdominal discomfort, weight loss, and icterus may occur. Gastrointestinal bleeding has been reported because of hemobilia[156] or hemosuccus pancreaticus.[70] The bleeding is due to the fact that hemorrhages occur in ASs. Autopsy revealed central necroses with surrounding hemorrhage.[70,156] Imaging features are nonspecific. Findings with[156] and without[70] pancreatic duct dilatation have been described. The descriptions of imaging are very straightforward. There are no specific criteria on ultrasonography for pancreatic ASs because of the rarity of the diagnosis. Computed tomography scan provides limited diagnostic utility because sarcomas of the pancreas range from highly homogeneous solid masses to highly inhomogeneous enhancing lesions with necrosis.[156] The finding of acute hemorrhagic necrotizing pancreatitis with peripancreatic fluid collections has also been described on CT. This appearance could be attributed to the reported hemorrhages in ASs.[157] Meeks et al.[67] described the difficulties of EUS-FNP in diagnostic assignment by cytology. Features of FNA of epithelioid AS are similar to those of adenocarcinomas. Without immunohistochemical staining, AS may also be difficult to distinguish histopathological from poorly differentiated adenocarcinomas. A histopathological examination and immunohistochemical analysis are required for a definitive diagnosis of AS.[67] The prognosis of AS is generally poor. There are only a few cases on the pancreas. Surgery was the treatment of choice. The prognosis was fatal in most patients. One case with survival within 1 year without evidence of recurrence is described[158] and 1 patient with a follow-up of 4 months without tumor recurrence after surgical resection and therapy with paclitaxel-based chemotherapy.[68]

Liposarcoma

Liposarcoma is the most common soft tissue sarcoma. Fifteen percent to 20% of all mesenchymal malignancies are liposarcomas. On the pancreas, however, liposarcoma is extremely rare.[11,108] Up to 2019, 7 cases have been described in a review in the English-language literature.[57–59,159–162] Descriptions of the appearance in imaging are very manageable. Tanabe et al.[163] reported that the

unenhanced CT at the time of first detection showed that the tumor was heterogeneous, with low-attenuation areas indicating fat components. As the tumor grew, the fat components became unclear.[163] Surgical resection is the treatment of choice.[58] The first description of pancreatic liposarcoma on magnetic resonance imaging (MRI) was by Machado et al.[58] Magnetic resonance imaging demonstrated a multilobulated, well-demarcated, and heterogeneous mass within the pancreatic head-neck interface with thick wall. The pattern of enhancement was marked hypovascular and signal predominantly hyperintense in T2-weighted sequences.[58] The pancreatic duct was dilated, but the common bile duct was not affected.[58] Therapy of choice is surgical resection.[58] Both conventional chemotherapy and radiotherapy have limited efficacy. A positive response to apatinib and a crossline rescue therapy combined with paclitaxel in an advanced pancreatic liposarcoma was reported.[162]

Sclerosing epithelioid fibrosarcoma

Sclerosing epithelioid fibrosarcoma is an extremely rare fibrosarcoma variant of the pancreas, with only a few cases being described.[80,164] The even rarer variant of the association of characteristics of a low-grade fibromyxoid sarcomas and sclerosing epithelioid fibrosarcomas in a pancreatic tumor has been described and analyzed histologically in detail.[81] The affected patients were middle-aged.[80,164] In the case reports described, there was no information on the sonographic appearance. In the CE-CT, the tumor appeared as a well-defined regular nodular mass with mild enhancement in the arterial phase and moderate heterogeneous enhancement in the portal phase and late phase. On unenhanced MRI, the lesion was hypointense on T1-weighted images, hyperintense on T2-weighted images, and slightly hyperintense on diffusion-weighted images.[80,81] Surgical resection is the treatment of choice.

Kaposiform hemangioendothelioma

Kaposiform hemangioendothelioma (KHE) is an aggressive, locally infiltrative tumor that primarily affects the skin. Visceral, retroperitoneal, and especially pancreatic manifestations are very rare. Kaposiform hemangioendothelioma is a vascular tumor with intermediate malignant potential. Kaposiform hemangioendothelioma shows histological features overlapping with benign hemangioma and Kaposi sarcoma.[165,166] Histologically, it is composed of infiltrating nodules, sheets of spindled endothelial cells, and slit-like vascular channels.[167] Predominantly infants, but also children are affected.[168] However, there are also cases among young adults.[166] In a review of 10 cases with pancreatic KHE, all tumors were located at the head of the pancreas, the tumors were 1.5 to 9.9 cm in size, and all patients presented with obstructive jaundice.[168] Fifty percent of the patients had severe thrombocytopenia and coagulopathy (Kasabach-Merritt phenomenon).[168] Kaposiform hemangioendothelioma is often associated with the Kasabach-Merritt phenomenon.[169] Diagnosis is based on histologic findings. Descriptions of imaging appearance of KHE are accordingly rare. On abdominal ultrasound, 1 case of KHE was described as a hyperechoic lesion,[168] and another case as a smoothly bordered, heterogeneous, and hypoechoic lesion.[170] There are no data on CEH-EUS. As it is a vascular tumor, hyperenhancement would be expected, but this is speculative. Radiological imaging describes the KHE as infiltrative lesions with solid, well-defined central regions.[168,171] Contrast-enhanced CT showed heterogeneous enhancement. Nonenhanced MRI scans showed heterogeneous and hyperintense enhancement compared with muscles on T2-weighted imaging, whereas enhanced MRI scans mostly showed intense heterogeneous enhancement.[168,171] The optimal therapy is surgical resection. For cases with a noncurative surgical treatment, variable responses to vincristine, corticosteroids, and interferon have been reported.[168] A new treatment approach is inhibitor of the mammalian target of rapamycin, sirolimus.[168,172–174]

CONCLUSION

It is important to recognize that not all pancreatic tumors correspond to a ductal adenocarcinoma, and not all well-vascularized tumors correspond to a neuroendocrine tumor. Transabdominal ultrasonography, CEUS, EUS, CEH-EUS, as well as EUS-guided sampling, are powerful diagnostic methods to obtain a variety of individual information for the diagnosis of rare pancreatic tumors. In the research of the rare pancreatic tumors described here, it was found that ultrasound played only a minor role and was used at most in primary diagnosis. We were able to demonstrate an example of a pancreatic EGIST with complex endosonographic diagnostics including CEH-EUS. In this example, the EGISTs proved to be well vascularized, similar to the known data of GISTs. This may allow differentiation from pancreatic ductal adenocarcinoma. In contrast, the derived arterial Doppler curves resemble those of pancreatic ductal adenocarcinoma. Sarcomas in the pancreas are rare, usually very large at diagnosis, but the paucity of reports allows no generalization of the observed imaging features. If the primary diagnosis is made safely by ultrasound, there are no differentiated reports on complex ultrasound diagnostics. The literature describes that the diagnosis of sarcomas is usually made only by histological confirmation. In our case studies, mesenchymal cells were described in EGISTs of the pancreas by a gastroenterologist experienced in on-site cytology (M.H.). In all other cases, EUS-guided FNB was performed to obtain a histological diagnosis. In the literature on rare tumors, imaging only visualized the tumor, whereas diagnosis was usually achieved histologically.

Conflicts of Interest

Siyu Sun is the Editor-in-Chief of the journal, and Christoph F. Dietrich is a Co-Editor-in-Chief. Michael Hocke and Christian Jenssen are Editorial Board Members. This article was subject to the journal's standard procedures, with peer review handled independently of the editors and their research groups. The authors declare that they have no financial conflict of interest with regard to the content of this report.

Author Contributions

All authors contributed to the study conception and design. The concept was developed by [Christoph F Dietrich and Kathleen Möller]. Material preparation, data collection and analysis were performed by [Kathleen Möller]. Image collection was performed by: [Christoph F Dietrich, Michael Hocke, Wei On, Simon M. Everett, Kathleen Möller, Mihai Rimbas]. The first draft of the manuscript was written by [Kathleen Möller] and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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