Review

Endoscopic Ultrasound

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Comments and illustrations of the European Federation of Societies for Ultrasound in Medicine contrast-enhanced ultrasonography guidelines: multiparametric imaging and EUS-guided sampling in rare pancreatic tumors. Mesenchymal pancreatic tumors of intermediate biological behaviour

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ABSTRACT

The focus of the review is on mesenchymal pancreatic tumors with intermediate biological behavior and their imaging appearance. Similar to benign and malignant mesenchymal pancreatic tumors, these tumors are extremely rare. The diagnosis is often confirmed only by postoperative histology. The very limited data on abdominal ultrasound and EUS findings including contrast-enhanced techniques of these pancreatic lesions are summarized here.

Key words: Mesenchymal pancreatic tumors; Contrast-enhanced ultrasonography (CEUS); EUS; Imaging

INTRODUCTION

The World Federation for Ultrasound in Medicine and Biology has published guidelines on the use of contrast-enhanced ultrasound for the evaluation of focal liver lesions,[1–5] and the European Federation of Societies for Ultrasound in Medicine for the evaluation of nonhepatic indications.[6,7] More recently, the guidelines have been commented and illustrated.[8–19] The Asian Federation of Societies of Ultrasound in Medicine and Biology has established guidelines for contrast-enhanced EUS.[20] Improved detection and characterization of common focal pancreatic lesions like ductal adenocarcinoma, neuroendocrine tumors, pancreatic metastases, and autoimmune pancreatitis are the main topics of these guidelines. The special features of small pancreatic tumors[21] and various intrapancreatic metastases[22] in ultrasound- and EUS-guided imaging are described. Mesenchymal tumors of the pancreas are very rare. Mesenchymal tumors occurring in the digestive tract and their biological potential are reported in the 2019 World Health Organization (WHO) classification.[23] A possible pancreatic manifestation is mentioned there. Primary mesenchymal tumors account for approximately 0.3%–0.5% of all histologically confirmed pancreatic tumors.[24,25] One-third of these are benign, intermediate, or malignant mesenchymal tumors, respectively.[24,25] The biological behavior is classified as intermediate if the tumor is either locally aggressive or rarely metastasizes (<2%).[23]

Primary manifestations of mesenchymal tumors must be differentiated from intrapancreatic sarcoma metastases, but also from infiltrations of mesenchymal tumors of the peripancreatic environment, the peripancreatic soft tissue, and retroperitoneum and aging processes.[26,27] Half of the primary mesenchymal tumors reported in a large surgical cohort study were located in the peripancreatic region.[24] Soft tissue tumors are classified by their specific lineage of differentiation, for example, adipocytic, fibroblastic/myofibroblastic, vascular, and smooth muscle neoplasms.[28] With the inclusion of new molecular genetic aspects, new tumor types have been designated in the WHO 2020 classification of soft tissue tumors.[28] Primary pancreatic mesenchymal tumors reported in the literature are listed in Table 1. The frequencies in the larger surgically resected or biopsied patient population are listed in Table 2. Preoperative diagnosis is a major challenge.

Preoperative diagnoses of all mesenchymal tumors in the retrospective studies of Kim et al.[24] and Zhang et al.[25] were as follows:

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Table 1

Mesenchymal tumors with primary pancreatic manifestation.

Intermediate mesenchymal pancreatic tumors	Benign mesenchymal pancreatic tumors	Malignant mesenchymal pancreatic tumors
Solitary fibrous tumor[24,25,29–42]	Lipoma[43–51]	Leiomyosarcoma[24,25,52–58]
Fibromatosis (desmoid tumor)[24,25,59–62]	Schwannoma[24,25,63–68]	Ewing sarcomas/primitive neuroectodermal Ewing sarcomas/primitive neuroectodermal tumors[24,69–75]
PEComa[76–82]	Hamartoma[24,83–90]	Undifferentiated/unclassified sarcomas (malignant fibrous histiocytoma)[24,25,91]
Inflammatory myofibroblastic tumor (inflammatory pseudotumor)[92–96]	Hemangioma[24,97–105]	Liposarcoma[24,106]
	Angiomyolipoma[24,107]	Angiosarcoma[108–112]
	Ganglioneuroma[25]	Fibrosarcoma[113–116]
	Myofibroblastoma[25]	Kaposi sarcoma[117,118]
		Rhabdomyosarcoma[119,120]
		Extragastrointestinal stromal tumor[25,121–124]

pancreatic ductal adenocarcinoma (PDAC), pancreatic neuroendocrine tumor, mucinous cystadenoma, serous microcystic adenoma, Castleman disease, chronic pancreatitis, solid pseudopapillary neoplasm (SPN), and invasive intraductal papillary mucinous neoplasm. The diagnoses of mesenchymal tumors in these series were probably all only made postoperatively on the resected specimen.[25]

The following work gives an overview of the pancreatic mesenchymal tumors of intermediate biological behavior, with data on imaging and the difficulties in preoperative diagnosis. These are perivascular epithelioid cell neoplasm, solitary fibrous tumors (SFTs), fibromatosis (desmoid tumors), and the inflammatory myofibroblastic tumor (IMT; inflammatory pseudotumor). Benign and malignant mesenchymal pancreatic tumors are not described in this article but in separate reviews.

PERIVASCULAR EPITHELIOID CELL NEOPLASM

A perivascular epithelial cell tumor (PEComa) is a rare mesenchymal tumor composed of histologically and immunohistochemically characteristic perivascular predominantly epithelial cells with variable expression of smooth muscle (actin, desmin) and melanocytic markers (HMB-45 and Melan-A).[23] PEComa is listed as benign in

Table 2

Prevalence of mesenchymal tumors in surgically resected or biopsy-proven pancreatic neoplasms.

Study	Mesenchymal tumors	Primary mesenchymal pancreatic tumors
Kim et al. (n = 7129), surgically resected or biopsy-proven pancreatic tumors[24]	**Mesenchymal tumors (n = 47)** • Metastatic sarcoma (n = 6) • Peripancreatic mesenchymal tumor (peripancreatic soft tissues, mesentery retroperitoneum) (n = 21) • **Primary mesenchymal pancreatic tumors (n = 30) (0.3%)**	**Benign and borderline mesenchymal pancreatic tumors (70%)** • Fibromatosis (n = 4) • Cavernous hemangioma (n = 2) • Schwannoma (n = 2) • Solid and cystic hamartoma (n = 2) • Solitary fibrous tumor (n = 2) • Inflammatory myofibroblastic tumor (n = 1) • Angiomyolipoma (n = 1) **Malignant mesenchymal tumors (30%)** • Undifferentiated/unclassified sarcoma (n = 3) • Leiomyosarcoma (n = 1) • Ewing sarcoma/primitive neuroectodermal tumor (n = 1) • Atypical lipomatous tumor/well-differentiated liposarcoma (n = 1)
Zhang et al. (n = 1944), surgically resected pancreatic tumors[25]	**Primary mesenchymal tumors (n = 10) (0.5%)**	**Benign and borderline tumors (70%)** • Solitary fibrous tumor (n = 2) • Fibromatosis/desmoid (n = 1) • Ganglioneuroma (n = 1) • Myofibroblastoma (n = 1) • Schwannoma (n = 1) • Uncertain malignant potential extragastrointestinal stromal tumor (eGIST) (n = 1) **Malignant mesenchymal tumors (30%)** • Malignant eGIST (n = 1) • malignant solitary fibrous tumor (n = 1) • undifferentiated pleomorphic sarcoma (n = 1)

the WHO classification of mesenchymal tumors. However, there are both benign (most common) and malignant (rare) forms.[23] For this reason, the PEComa is discussed here as a mesenchymal tumor of intermediate biological potential.

Bonetti et al.[125] were the first to use the term "perivascular epithelioid cells." The "family" of PEComas also includes angiomyolipoma, clear cell "sugar" tumor of the lung, and lymphangioleiomyomatosis.[126] A pancreatic PEComa was first reported by Zamboni et al. in 1996.[127] The term "sugar tumor" was given because the perivascular epithelioid cells of this tumor have a clear cell cytoplasm and are rich in glycogen.[127] Angiomyolipoma is a PEComa subtype, containing adipocytes and thick-walled tortuous blood vessels in addition to perivascular epithelioid cells.[23] Sixto et al. performed a systematic review of the literature and reviewed an own case and 34 pancreatic PEComas from the literature.[128]

Predominantly women are affected (male-to-female ratio 1:4) with a mean age of 48.4 years. All age groups may be affected (range, 17–74 years). Abdominal pain was present in 51.4% of patients, 22.9% were incidental findings, 14.3% presented as a mass lesion, and the remaining patients had various, mostly nonspecific complaints. The mean tumor size was 3.88 cm (range, 6–15 cm). The majority of pancreatic PEComas were located in the pancreatic head (42.8%), 28.6% in the body, and 17.1% in the tail, and 8.6% involved more than 1 part of the pancreas.[128] PEComas are classified in benign/uncertain malignant and malignant. For prognostic assessment of the malignant potential, Folpe et al. formulated "worrisome features" for all localizations. These include a size greater than 5 cm, infiltrative margins, high nuclear grade, cellularity, mitotic rate of ≥1/10 high-power field, necrosis, and vascular invasion.[129] Presence of more than 2 of these characteristics classifies the tumor as malignant,[129] and 14% of pancreatic PEComas reviewed by Almousa et al. were malignant.[79]

On abdominal ultrasonography, the pancreatic PEComa is described as smooth-bordered, homogeneous,[80,82] and on color Doppler imaging as highly vascularized.[80] In EUS, pancreatic PEComas are well defined and homogeneously or heterogeneously hypoechoic.[76–78,80,81,130,131] A small cystic portion is described in only 1 case.[131] On computed tomography (CT), the pancreatic PEComa appears to be well defined and heterogeneously enhanced almost to the same degree as the surrounding pancreatic tissue in both arterial and portal venous phases.[78] Tumors were slightly hypodense on delayed CT imaging, whereas some presented delayed enhancement.[78] On MRI, it was slightly hyperintense on T2-weighted magnetic resonance imaging (MRI). Diffusion-weighted imaging showed the lesion was markedly hyperintense.[78] Uno et al. reported a well-defined hypoechoic mass region in the pancreatic tail on EUS and contrast-enhanced harmonic EUS with Sonazoid showing the tumor to be isoenhanced compared with surrounding pancreatic tissue.[78] A complex endosonographic description was given by Ulrich et al. A 25-mm smooth-bordered tumor was heterogeneous, hypoechoic with lateral shadowing. The pancreatic duct was not tangentially involved. On EUS, the tumor was stiffer. On native power Doppler, tumor hypervascularity was suspected. In contrast-enhanced harmonic EUS with SonoVue, the tumor was hyperenhanced and showed a long-lasting enhancement in contrast to pancreatic neoplasms (PanNEN). It was confirmed by EUS-guided fine-needle aspiration (FNA) using a 22-gauge needle.[76] Pancreatic PEComa data are summarized in Table 3.

EUS-guided sampling (EUS-FNA or EUS-guided fine-needle biopsy [FNB]) has been performed in 19 of 35 (54.3%) pancreatic PEComa cases reported so far in the literature.[128] Definite diagnosis was possible in only 13 of these 19 cases (diagnostic yield 63.2%).[128] On EUS-guided sampling, cytology was characterized by epithelioid spindle-shaped cells with abundant granular eosinophilic cytoplasm and distinct prominent nucleoli. In immunohistochemistry, pancreatic PEComas express simultaneously melanocytic (HMB-45, Melan-A) and smooth muscle markers (desmin, actin), whereas staining for neuroendocrine markers (synaptophysin, chromogranin A), CD-117, S-100, and cytokeratin markers remains negative. This immunohistochemical pattern is diagnostic.[76,128] Cytology smears alone are not conclusive, and immunohistochemical studies must be performed to prove the diagnosis.[76–79,128,131]

Important differential diagnoses in imaging include other well-vascularized tumors such as PanNEN or renal cell carcinoma metastases and finally an intrapancreatic accessory spleen. Due to the malignant potential of pancreatic PEComa, treatment of choice is surgical resection.

INFLAMMATORY MYOFIBROBLASTIC TUMOR

Inflammatory myofibroblastic tumor is a marked fibroblastic/myofibroblastic neoplasm with intermediate biological potential.[23] Inflammatory myofibroblastic tumor is composed of fibroblasts and myofibroblasts, usually arranged in a storiform pattern, with moderate to marked inflammation. Focally, IMT may have the same morphologic appearance as that of immunoglobulin G4 (IgG4)–related sclerosing disease. However, the majority of IMTs are distinguished from IgG4-related lesions by ALK (anaplastic lymphoma kinase) gene expression, a low infiltration of $IgG4^+$ cells, and the absence of obstructive phlebitis.[92] In the literature, the tumor is alternatively referred to as plasma cell granuloma, plasma cell pseudotumor, inflammatory

Table 3

Pancreatic PEComa on imaging.

Method	Appearance
Ultrasound	Well-defined, hypoechoic on color Doppler imaging (CDI) highly hypervascularized[80,82]
Computed tomography	Well-defined, heterogeneously enhanced almost to the same degree as the surrounding pancreatic tissue in both arterial and portal venous phases. Tumors were slightly hypodense on delayed computed tomography imaging, whereas some presented delayed enhancement[78]
Magnetic resonance imaging	Slightly hyperintense on T2[78]
EUS, CH-EUS	Well-defined, hypoechoic, heterogeneous, stiffer on elastography, hypervascularization on CDI[76–78,80,81,130,131] CH-EUS with Sonazoid: isoenhancement (single case)[78] CH-EUS with SonoVue: hyperenhancement on CH-EUS with long-lasting enhancement (single case)[76]

CH-EUS: Contrast Harmonic-EUS.

pseudotumor, inflammatory fibroxanthoma, and histiocytoma.[96] The official name in the WHO classification is IMT.[23] There are 29 cases described in the pancreas.[96] Men (69%) are more frequently affected than women (31%). Mean age was 42 years with range from 6 month to 82 years. The most common location was in the head of the pancreas (72%). Tumor size ranged from 1.5 to 15.0 cm. Most patients had abdominal pain. When localized in the pancreatic head, jaundice due to bile duct obstruction was common. Symptoms due to pancreatic duct stenosis or vascular infiltration were observed. The appearance on imaging depends on the composition of the tumor.

Sonographically, IMT is described as well demarcated, lobulated, and both hyperechoic and hypoechoic, also with cystic parts.[93–96] Color Doppler imaging showed focal macrovessels.[95] In CT and MRI, IMT can exhibit variable attenuation and signal intensity with variable heterogeneous enhancement.[95] On contrast-enhanced CT scan, a case of IMT showed lower attenuation in the precontrast phase, and heterogeneous hyperenhancement in the arterial and portal venous phase with relatively homogeneous enhancement in the delayed phase.[95] On MRI, IMT was hypointense or hyperintense on the T1-weighted image and mildly hyperintense on the T2-weighted image.[95,96] A centripetal enhancement pattern was observed in the delayed phase of contrast enhancement.[96]

Differential diagnosis

Typically suspected preoperative diagnoses include PDAC, PanNEN, SPN, other mesenchymal tumors, or IgG4-related pseudotumor, depending on the extent of tumor vascularization, bile duct stenosis, and pancreatic duct dilatation. Pancreatic IMT is regarded as a low-grade malignancy with a generally favorable prognosis. Pulmonary metastases occurred in 1 of 29 patients after 6 years.[132] Twenty of 29 patients were free from recurrence during the observation period. No data are available for the remaining patients.[96] Patients must be followed up carefully, as recurrences and metastases may occur even in the long term.

SOLITARY FIBROUS TUMOR

Solitary fibrous tumors (formerly hemangiopericytoma) are characterized by an intermediate biological potential. Most tumors have a benign course; however, 5% to 10% of tumors recur or metastasize, typically to the lungs, liver, and bones, including occasional cases with benign histology.[28] Apparently malignant SFTs have a metastasis rate of 20% to 30%.[28] Most frequent localizations are the abdominal cavity, 31%; limbs, 29%; pleura, 22%; trunk, 11%; and others, 7%.[133] The incidence of all SFTs is very low with about 1 case/1 million people per year.[133] Risk stratification of SFT is based on age (<55 or >55 years), tumor size (<5 to ≥15 cm in increments of 5 cm), and mitotic count (0, 1–3, or ≥4/10 high-power fields), and presence of necrosis. Accordingly, the tumor is classified as low/intermediate and high risk.[28,134] Immunohistochemical markers commonly expressed in SFTs are CD34, bcl2, CD99, and STAT6 (signal transducer and activator of transcription 6).[133] Pancreatic localization is extremely rare. In a review, a total of 29 cases were researched by 2020.[39] The median age of the patients was 55 years, and both sexes were equally affected. The most frequent location was the pancreatic head (59%). Thirty-eight percent were incidental findings. The remaining patients had abdominal complaints or nonspecific complaints.[39] Refractory hypoglycemia due to increased secretion of a prohormone form of the insulinlike growth factor II is seen in the context of Doege-Potter syndrome and is usually an expression of a malignant SFT.[40]

Sonographically, a pancreatic SFT is described as a well-defined heterogeneous mass.[39] Kwon et al. described a smooth bordered, ovoid, heterogeneous lesion with cystic portion with hypoechoic heterogeneous content on EUS.[42] Yamashita et al. described a circumscript well-encapsulated, multicystic solid SFT with hypervascularization on EUS.[38] The only reported pancreatic metastasis from a primary SFT was smooth-bordered and hypoechoic on EUS.[135] Most pancreatic SFTs were well demarcated on imaging, hypervascular with internal heterogeneous contrast enhancing on CT, hypointensity on T1-weighted imaging, and hyperintensity on T2-weighted imaging on MRI.[39–41] These features are not typical and do not distinguish SFT from other pancreatic mesenchymal tumors. Only 1 of 29 (3.4%) pancreatic SFTs was confirmed preoperatively. Another 3 of 29 (10.3%) were considered as differential diagnosis.[39] Sufficient material for immunohistochemistry is required when EUS-guided sampling is performed. Nevertheless, diagnosis may not be definitive if the material is not adequate or the immunohistochemistry is not typical.[33,39]

Differential diagnosis

The most often suspected diagnoses preoperatively were PanNEN, occasionally SPN, mesenchymal tumor, extragastrointestinal stromal tumor (eGIST), Acinar cell carcinoma (ACC), and cystic adenocarcinoma. A diagnosis of malignant pancreatic SFT was ultimately made in 6 of 29 (20.7%) cases. Of these, 3 of 6 (50%) malignant SFTs and 3 of all 29 (10.3%) pancreatic SFTs showed recurrence, respectively.[39] Even though most SFTs are not recognized as such before surgical resection, this is also the standard therapy of choice[28,39,133] due to their malignant potential. SFT is sensitive to radiotherapy. A retrospective series of patients treated with definitive radiotherapy (60 Gy) reported an overall response rate of 67% with a 5-year local control of 81.3% and 5-year overall survival of 87.5%.[133,136] In case of metastasis, anthracycline-based chemotherapy regimens and antiangiogenesis inhibitors are used[133] [Figure 1].

FIBROMATOSIS/DESMOID

Desmoid fibromatosis (desmoid tumor) is a rare benign tumor, but often with locally aggressive, infiltrative, fibrous growth.[137,138] In the WHO classification, desmoid fibromatosis is classified as intermediate (locally aggressive) fibroblastic and myofibroblastic tumors.[23] Familial adenomatous polyposis/Gardner syndrome increases the lifetime risk of desmoid fibromatosis up to 10%–30%.[139] In the pancreas, 32 cases have been researched up to 2021.[60] The tumors were usually surgically resected under suspicion of a malignant tumor, and the actual diagnosis was only established postoperatively. Desmoid tumors manifest as dense fibrous masses. Microscopically, spindle cells are detectable with minimal atypia within a dense collagenous background that infiltrates normal tissue. Extragastrointestinal stromal tumors are important differential diagnoses. Positive β-catenin staining and mutational analysis of CTNNB1 are important in establishing the diagnosis.[60,137,140]

In a review of 32 cases of pancreatic desmoid fibromatosis, men and women were affected in equal proportions. The median age was 39 years. Patents were either asymptomatic or had epigastric discomfort and nonspecific symptoms such as weight loss. The pancreatic tail was the most common site (40%). Forty-two percent each were solid or cystic/solid, and 16% were cystic. Fifty-three percent had infiltration of surrounding organs.[60] Additional 7 cases were reported in 2022 with a clear preponderance of females (male-to-female ratio 1:6) and a mean age of 54 years.[61] In another review, the mean

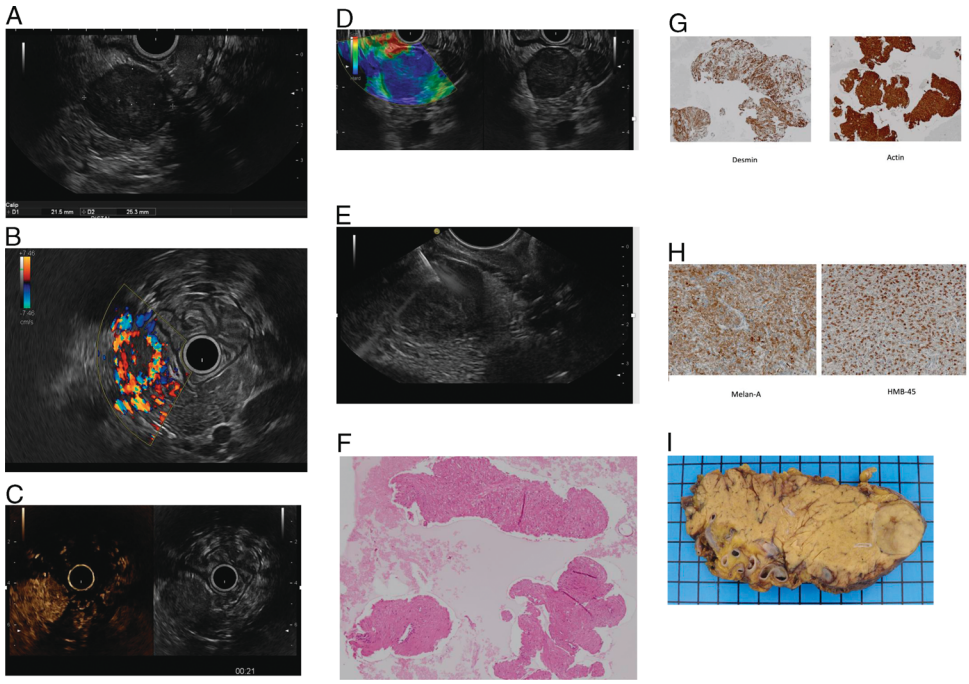

Figure 1. A 43-year-old woman underwent upper EUS, which showed a well-defined pancreatic neck mass measuring 25 × 20 mm (A). The mass showed a whorled-like pattern of hyperechogenicity that was hypervascular (B, C) and stiffer than the surrounding parenchyma (D). Biopsy (22-gauge) confirmed the diagnosis of PEComa (E). Histologic sections of fragments of the pancreatic lesion consisting of sheets of uniform epithelioid cells with abundant granular eosinophilic cytoplasm and distinct prominent nucleoli (F). Immunohistochemistry showed positivity for desmin and actin (G) and melanocytic markers including melanoma-associated antigen (HMB-45) (H) and smooth muscle antigen (SMA [not shown]) and negativity for cytokeratin (CK), synaptophysin (SYN), and chromogranin (CG [not shown]). Pancreaticoduodenectomy was performed due to the malignant potential (I).

tumor size was 7.99 cm.[141] On CT, they may appear as well-defined or poorly defined masses with variable attenuation. On MRI, they show low signal intensity compared with muscle on T1-weighted images. T2 signal is variable (some desmoid tumors have been described as cystic). Dallaire et al. described pancreatic desmoid fibromatosis as hyperintense with a hypointense nodular capsule on T2-weighted images and heterogeneous, predominantly hypointense on T1-weighted images. After gadolinium injection, the lesion showed progressive enhancement in the venous and late phases, which was maximal in the latest phase.[62] In an endosonographic case report, the lesion presented oval, smooth bordered, hypoechoic, and heterogeneous and with patchy acoustic shadowing. EUS-guided sampling (22-gauge) was able to diagnose a pancreatic desmoid tumor preoperatively. The aspirates showed a low-grade spindle cell lesion. On immunohistochemistry using the cell block technique, the cells were positive for β-catenin. At the cell block technique, it was possible to perform a mutation analysis, which detected the diagnostic *CTNNB1* gene mutation (*T41A*)[60] [Figure 2].

Differential diagnosis

Preoperative suspected diagnoses were mucinous neoplasia, intraductal papillary mucinous neoplasm, SPN, PanNEN, eGIST, PDAC, and IMT.[60]

The treatment strategy of desmoid tumors includes both a watch-and-wait strategy and surgical resection. In a nonrandomized single-center study of nonpancreatic desmoid tumors, the progression-free survival rate after 3 years (88.9%) in the watch and wait-group was higher than the postoperative recurrence-free rate in the surgical group (77.1%).[138] Because the diagnosis of pancreatic desmoid tumors is usually made postoperatively, there is no corresponding experience. In a 2022 review by Litchinko et al, tumor recurrence was absent in 18 of 31 cases (58%) after pancreatic resection. For all other cases, data were not available.[141]

Particularly, in patients with familial adenomatous polyposis, desmoid fibromatosis should be considered in case of a pancreatic tumor.

CONCLUSION

Mesenchymal pancreatic tumors with intermediate biological behavior like all pancreatic mesenchymal tumors are very rare. At best, there are case series in double digits for the different entities. Although contrast-enhanced abdominal ultrasound and EUS are powerful methods to characterize these tumors, there are only isolated case reports available that do not allow generalizations. Mesenchymal tumors of intermediate biological behavior are usually well defined and hypoechoic on ultrasound and EUS. Fibromatosis may have cystic parts. The PEComa is vascularized in contrast enhancement. Based on the limited data and isolated case reports only, no general imaging features can be derived. Predominantly, patients underwent CT and MRI. Contrast-enhanced ultrasound or EUS was performed only occasionally. The diagnosis is often made only postoperatively. Surgical resection is indicated for all mesenchymal tumors with intermediate biological behavior and the usually unclear preoperative diagnosis. Only in the case of fibromatosis (desmoid) a watch-and-wait strategy can be an alternative. In cases where EUS-guided sampling was performed, most of the results did not lead to the final diagnosis. The type of sampling, whether EUS-FNA or EUS-FNB, and needle size were not

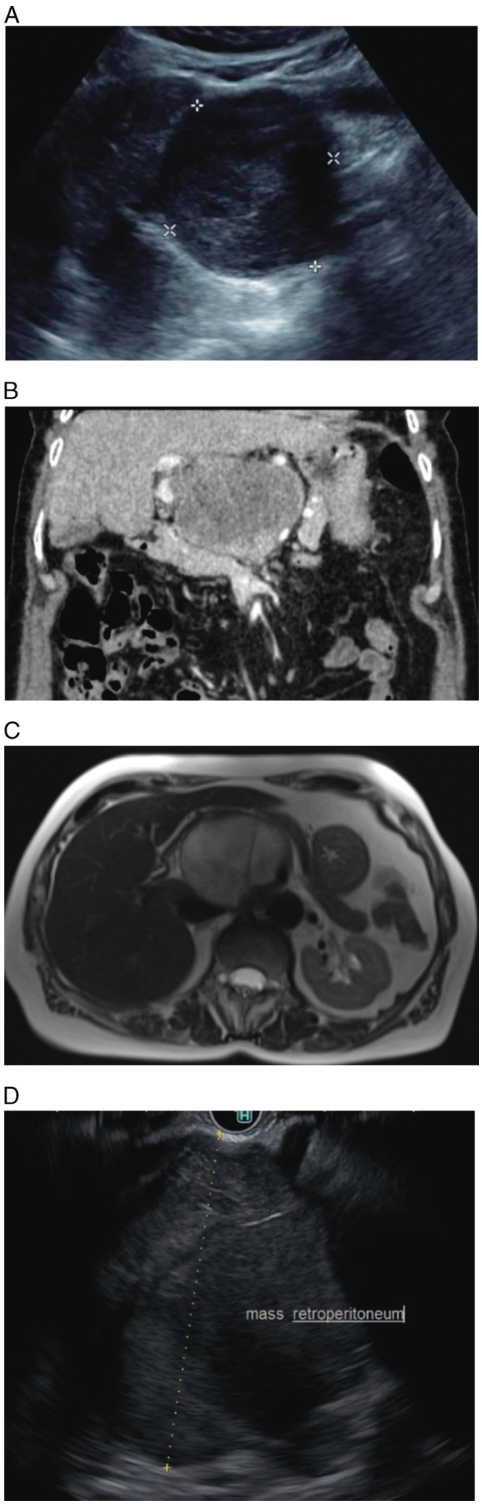

Figure 2. A 74-year-old woman underwent a TUS (A) following complaints of renal colic. TUS demonstrated a solid, well-circumscribed pancreatic lesion next to the caudate lobe of the liver. A computed tomography (B) showed a large mass with areas of hyperenhancement appearing completely separate to the adjacent organs and displacing the mesenteric vasculature. Magnetic resonance imaging (C) demonstrated multiple internal septations in the lesion with homogenous high T2 signaling throughout. EUS (D) demonstrated a large, solid, hypoechoic mass in the retroperitoneum. Fine-needle biopsy confirmed the diagnosis of SFT. TUS: Transabdominal Ultrasonography.

always clearly designated. However, the review articles clearly show that definitive preoperative diagnosis of these very rare tumors is possible only if the specimens are sufficient for extensive immunophenotyping. Therefore, we recommend the use of EUS-FNB needles and the performance of multiple needle passages to obtain a suitable quantity of tissue cylinders when rare pancreatic tumors are suspected by the examiner.

Conflicts of Interest

Siyu Sun is the Editor-in-Chief of the journal, and Christoph F. Dietrich is a Co-Editor-in-Chief. Christian Jenssen and Michael Hocke 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.

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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, Christoph Schlag, Wei On, Simon M. Everett]. 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.

References

1. Dietrich CF, Averkiou M, Nielsen MB, et al. How to perform contrast-enhanced ultrasound (CEUS). *Ultrasound Int Open* 2018;4:E2–E15.
2. Claudon M, Dietrich CF, Choi BI, et al. Guidelines and good clinical practice recommendations for contrast enhanced ultrasound (CEUS) in the liver—update 2012: a WFUMB-EFSUMB initiative in cooperation with representatives of AFSUMB, AIUM, ASUM, FLAUS and ICUS. *Ultraschall Med* 2013;34:11–29.
3. Claudon M, Dietrich CF, Choi BI, et al. Guidelines and good clinical practice recommendations for contrast enhanced ultrasound (CEUS) in the liver—update 2012: a WFUMB-EFSUMB initiative in cooperation with representatives of AFSUMB, AIUM, ASUM, FLAUS and ICUS. *Ultrasound Med Biol* 2013;39:187–210.
4. Dietrich CF, Nolsøe CP, Barr RG, et al. Guidelines and good clinical practice recommendations for contrast-enhanced ultrasound (CEUS) in the liver—update 2020 WFUMB in cooperation with EFSUMB, AFSUMB, AIUM, and FLAUS. *Ultrasound Med Biol* 2020;46:2579–2604.
5. Dietrich CF, Nolsøe CP, Barr RG, et al. Guidelines and good clinical practice recommendations for contrast enhanced ultrasound (CEUS) in the liver—update 2020—WFUMB in cooperation with EFSUMB, AFSUMB, AIUM, and FLAUS. *Ultraschall Med* 2020;41:562–585.
6. Sidhu PS, Cantisani V, Dietrich CF, et al. The EFSUMB guidelines and recommendations for the clinical practice of contrast-enhanced ultrasound (CEUS) in non-hepatic applications: update 2017 (short version). *Ultraschall Med* 2018;39:154–180.
7. Piscaglia F, Nolsøe C, Dietrich CF, et al. The EFSUMB guidelines and recommendations on the clinical practice of contrast enhanced ultrasound (CEUS): update 2011 on non-hepatic applications. *Ultraschall Med* 2012; 33:33–59.
8. Zadeh ES, Schreiber N, Gorg C, et al. Comments on and illustrations of the WFUMB CEUS liver guidelines: rare malignant mesenchymal liver lesions. *Med Ultrason* 2023.
9. Safai Zadeh E, Gorg C, Huber KP, et al. Comments and illustrations of the WFUMB CEUS liver guidelines: rare malignant hematological liver lesions. *Med Ultrason* 2023.

10. Moller K, Hoffmann O, Zadeh ES, et al. Comments and illustrations of the WFUMB CEUS liver guidelines: peliosis hepatis and porphyria. *Med Ultrason* 2023.
11. Möller K, Friedrich T, Safai Zadeh E, et al. Comments and illustrations of the WFUMB CEUS liver guidelines: cystic fibrosis associated liver disease. *Med Ultrason* 2023.
12. Dong Y, Wang WP, Zadeh ES, et al. Comments and illustrations of the WFUMB CEUS liver guidelines: rare benign focal liver lesion, part I. *Med Ultrason* 2024;26(1):50–62.
13. Dong Y, Guggisberg E, Wang W-P, et al. Comments and illustrations of the WFUMB CEUS liver guidelines: rare benign focal liver lesion, part II. *Med Ultrason* 2023.
14. Moller K, Zadeh ES, Gorg C, et al. Comments and illustrations of the WFUMB CEUS liver guidelines: rare vascular pathology, part II. *Med Ultrason* 2023;25:189–200.
15. Moller K, Stock B, Ignee A, et al. Comments and illustrations of the WFUMB CEUS liver guidelines: rare focal liver lesions—non-infectious, non-neoplastic. *Med Ultrason* 2023;25:435–444.
16. Cekuolis A, Schreiber-Dietrich D, Augustiniene R, et al. Incidental findings in pediatric patients: how to manage liver Incidentaloma in pediatric patients. *Cancers (Basel)* 2023;15:2360.
17. Zander T, Zadeh ES, Moller K, et al. Comments and illustrations of the WFUMB CEUS liver guidelines: rare focal liver lesion—infectious parasitic, fungus. *Med Ultrason* 2023;25:423–434.
18. Ignee A, Moller K, Thees-Laurenz R, et al. Comments and illustrations of the WFUMB CEUS liver guidelines: rare focal liver lesions—infectious (bacterial). *Med Ultrason* 2023;25:312–324.
19. Moller K, Tscheu T, De Molo C, et al. Comments and illustrations of the WFUMB CEUS liver guidelines: rare congenital vascular pathology. *Med Ultrason* 2022;24:461–472.
20. Kitano M, Yamashita Y, Kamata K, et al. The Asian Federation of Societies for Ultrasound in Medicine and Biology (AFSUMB) guidelines for contrast-enhanced endoscopic ultrasound. *Ultrasound Med Biol* 2021;47:1433–1447.
21. Dietrich CF, Sahai AV, D'Onofrio M, et al. Differential diagnosis of small solid pancreatic lesions. *Gastrointest Endosc* 2016;84:933–940.
22. Moller K, Jenssen C, Braden B, et al. Comments on and illustrations of the EFSUMB CEUS guidelines: transabdominal and endoscopic ultrasound features of Intrapancreatic metastases and the role of multiparametric imaging and EUS-guided sampling in rare pancreatic tumors. *Cancers (Basel)* 2023;15:2546.
23. Fukayama M GJ, Miettinen M, Lazar AJ. Mesenchymal tumours of the digestive system. Digestive system tumours. Pp 436-498. In: WHO Classification of Tumours Editorial Board, ed. WHO Classification of Tumours. 5th ed. X, Lyon, France: World Health Organization, https://afkebooks.com.
24. Kim JY, Song JS, Park H, et al. Primary mesenchymal tumors of the pancreas: single-center experience over 16 years. *Pancreas* 2014;43: 959–968.
25. Zhang H, Yu S, Wang W, et al. Primary mesenchymal tumors of the pancreas in a single center over 15 years. *Oncol Lett* 2016;12:4027–4034.
26. Moller K, Jenssen C, Braden B, et al. Pancreatic changes with lifestyle and age: what is normal and what is concerning? *Endosc Ultrasound* 2023;12: 213–227.
27. Möller K, Jenssen C, Ignee A, et al. Pancreatic duct imaging during aging. *Endosc Ultrasound* 2023;12:200–212.
28. Kallen ME, Hornick JL. The 2020 WHO classification: what's new in soft tissue tumor pathology? *Am J Surg Pathol* 2021;45:e1–e23.
29. Tasdemir A, Soyuer I, Yurci A, Karahanli I, Akyildiz H. A huge solitary fibrous tumor localized in the pancreas: a young women. *JOP* 2012;13: 304–307.
30. Chen JW, Lu T, Liu HB, et al. A solitary fibrous tumor in the pancreas. *Chin Med J (Engl)* 2013;126:1388–1389.
31. Chetty R, Jain R, Serra S. Solitary fibrous tumor of the pancreas. *Ann Diagn Pathol* 2009;13:339–343.
32. Sugawara Y, Sakai S, Aono S, et al. Solitary fibrous tumor of the pancreas. *Jpn J Radiol* 2010;28:479–482.
33. Baxter AR, Newman E, Hajdu CH. Solitary fibrous tumor of the pancreas. *J Surg Case Rep* 2015;2015.
34. Paramythiotis D, Kofina K, Bangeas P, Tsiompanou F, Karayannopoulou G, Basdanis G. Solitary fibrous tumor of the pancreas: case report and review of the literature. *World J Gastrointest Surg* 2016;8:461–466.
35. Spasevska L, Janevska V, Janevski V, Noveska B, Zhivadinovik J. Solitary fibrous tumor of the pancreas: a case report and review of the literature. *Pril (Makedon Akad Nauk Umet Odd Med Nauki)* 2016;37:115–120.
36. Oana S, Matsuda N, Sibata S, Ishida K, Sugai T, Matsumoto T. A case of a "wandering" mobile solitary fibrous tumor occurring in the pancreas. *Clin J Gastroenterol* 2017;10:535–540.
37. Estrella JS, Wang H, Bhosale PR, Evans HL, Abraham SC. Malignant solitary fibrous tumor of the pancreas. *Pancreas* 2015;44:988–994.
38. Yamashita H, Fujino Y, Ohara T, et al. A rare case of metastatic solitary fibrous tumor of the pancreas manifesting as a cystic neoplasm: a case report. *Surg Case Rep* 2019;5:142.
39. Taguchi Y, Hara T, Tamura H, et al. Malignant solitary fibrous tumor of the pancreas: a case report. *Surg Case Rep* 2020;6:287.
40. Geng H, Ye Y, Jin Y, et al. Malignant solitary fibrous tumor of the pancreas with systemic metastasis: a case report and review of the literature. *World J Clin Cases* 2020;8:343–352.
41. Li J, Li J, Xiong Y, et al. Atypical/malignant solitary fibrous tumor of the pancreas with spleen vein invasion: case report and literature review. *Medicine (Baltimore)* 2020;99:e19783.
42. Kwon HJ, Byun JH, Kang J, Park SH, Lee MG. Solitary fibrous tumor of the pancreas: imaging findings. *Korean J Radiol* 2008;9(Suppl):S48–S51.
43. Beddi A, Merzem A, Harmak M, et al. Pancreatic lipoma: a pancreatic incidentaloma diagnosis with computed tomography. *Eur J Case Rep Intern Med* 2021;8:002252.
44. Deschner B, Gandhi J, Deneve JL, Dickson PV, Clark I, Glazer ES. Symptomatic pancreatic lipoma. *J Gastrointest Surg* 2019;23:1942–1943.
45. Butler JR, Fohtung TM, Sandrasegaran K, et al. The natural history of pancreatic lipoma: does it need observation. *Pancreatology* 2016;16: 95–98.
46. Di Matteo FM, Shimpi L, Pandolfi M, et al. EUS diagnosis of pancreatic lipoma: a case report. *Gastrointest Endosc* 2006;64:146–148.
47. Pausawasdi N, Apisarnthanarak P, Pongpaibul A, Charatcharoenwitthaya P. Pancreatic lipoma diagnosed by EUS-FNA. *Gastrointest Endosc* 2012; 76:668–669.
48. Suzuki R, Irisawa A, Hikichi T, et al. Pancreatic lipoma diagnosed using EUS-FNA. A case report. *JOP* 2009;10:200–203.
49. Jairo M, German T, Isabel B, Gloria M, Ruben A. Symptomatic pancreatic lipoma managed with a metallic biliary stent: case report. *Int J Surg Case Rep* 2022;93:106972.
50. Walker BS, Davis JL, Mayo SC. Large pancreatic lipoma causing duodenal obstruction. *J Gastrointest Surg* 2021;25:1070–1072.
51. Xiao RY, Yao X, Wang WL. A huge pancreatic lipoma mimicking a well-differentiated liposarcoma: a case report and systematic literature review. *World J Clin Cases* 2019;7:2352–2359.
52. Zhang H, Jensen MH, Farnell MB, Smyrk TC, Zhang L. Primary leiomyosarcoma of the pancreas: study of 9 cases and review of literature. *Am J Surg Pathol* 2010;34:1849–1856.
53. Hur YH, Kim HH, Park EK, et al. Primary leiomyosarcoma of the pancreas. *J Korean Surg Soc* 2011;81(Suppl 1):S69–S73.
54. Milanetto AC, Lico V, Blandamura S, Pasquali C. Primary leiomyosarcoma of the pancreas: report of a case treated by local excision and review of the literature. *Surg Case Rep* 2015;1:98.
55. Søreide JA, Undersrud ES, Al-Saiddi MSS, Tholfsen T, Søreide K. Primary Leiomyosarcoma of the pancreas—a case report and a comprehensive review. *J Gastrointest Cancer* 2016;47:358–365.
56. Aleshawi AJ, Allouh MZ, Heis FH, Tashtush N, Heis HA. Primary leiomyosarcoma of the pancreas: a comprehensive analytical review. *J Gastrointest Cancer* 2020;51:433–438.
57. Izumi H, Okada K, Imaizumi T, et al. Leiomyosarcoma of the pancreas: report of a case. *Surg Today* 2011;41:1556–1561.
58. Maarouf A, Scoazec JY, Berger F, Partensky C. Cystic leiomyosarcoma of the pancreas successfully treated by splenopancreatectomy: a 20-year follow-up. *Pancreas* 2007;35:95–98.
59. Rao RN, Agarwal P, Rai P, Kumar B. Isolated desmoid tumor of pancreatic tail with cyst formation diagnosed by beta-catenin immunostaining: a rare case report with review of literature. *JOP* 2013;14:296–301.
60. Stone AB, Mallery JS, Stewart J 3rd, Amin K. A rare sporadic pancreatic desmoid fibromatosis diagnosed by endoscopic ultrasound–guided fine-needle aspiration: case report and literature review. *Diagn Cytopathol* 2021;49:E49–E54.
61. Clarke-Brodber AL, Hartley CP, Ahmed F, Thangaiah JJ, Tiegs-Heiden C, Hagen CE. Desmoid fibromatosis involving the pancreas: a retrospective case series with clinical, cytopathologic and radiologic correlation. *Ann Diagn Pathol* 2022;60:152015.
62. Dallaire DFB, Dallaire DE, Perigny DM. Pancreatic desmoid tumor: a rare case with radiologic-pathologic correlation. *Radiol Case Rep* 2018;13: 1079–1083.
63. Zhang X, Siegelman ES, Lee MK 4th, Tondon R. Pancreatic schwannoma, an extremely rare and challenging entity: report of two cases and review of literature. *Pancreatology* 2019;19:729–737.
64. Ma Y, Shen B, Jia Y, et al. Pancreatic schwannoma: a case report and an updated 40-year review of the literature yielding 68 cases. *BMC Cancer* 2017;17:853.

65. Wang S, Xing C, Wu H, Dai M, Zhao Y. Pancreatic schwannoma mimicking pancreatic cystadenoma: a case report and literature review of the imaging features. *Medicine (Baltimore)* 2019;98:e16095.
66. Shi Z, Cao D, Zhuang Q, et al. MR imaging features of pancreatic schwannoma: a Chinese case series and a systematic review of 25 cases. *Cancer Imaging* 2021;21:23.
67. Hanaoka T, Okuwaki K, Imaizumi H, et al. Pancreatic schwannoma diagnosed by endoscopic ultrasound–guided fine-needle aspiration. *Intern Med* 2021;60:1389–1395.
68. Ntafam C, Miller AT, Beutler BD, Bamporiki J, Sahakian AB, Cheng PM. Pancreatic schwannoma: case report, clinico-pathologic correlation, and review of the literature. *Radiol Case Rep* 2022;17:3504–3510.
69. Bose P, Murugan P, Gillies E, Holter JL. Extraosseous Ewing's sarcoma of the pancreas. *Int J Clin Oncol* 2012;17:399–406.
70. Saif MW, Kaley K. Extraosseous Ewing's sarcoma of the pancreas: an uncommon but treatable disease. *Cureus* 2017;9:e1882.
71. Wu TT, Wang CF, Liu X, et al. Extraosseous Ewing's sarcoma of the pancreas: a clinicopathological analysis of 3 cases. *Zhonghua Bing Li Xue Za Zhi* 2023;52:49–51.
72. Welsch T, Mechtersheimer G, Aulmann S, et al. Huge primitive neuroectodermal tumor of the pancreas: report of a case and review of the literature. *World J Gastroenterol* 2006;12:6070–6073.
73. Nishizawa N, Kumamoto Y, Igarashi K, et al. A peripheral primitive neuroectodermal tumor originating from the pancreas: a case report and review of the literature. *Surg Case Rep* 2015;1:80.
74. Teixeira U, Goldoni M, Unterleider M, et al. Primitive neuroectodermal tumor of the pancreas: a case report and review of the literature. *Case Rep Surg* 2015;2015:276869.
75. Achufusi TG, Sohal R, Zamora E, Harne P, Russo R. Primitive neuroectodermal tumor of the pancreas. *Proc (Bayl Univ Med Cent)* 2020; 34:144–145.
76. Ulrich JD, Specht K, Schlitter AM, et al. A rare case of perivascular epithelioid cell tumor (PEComa) of the pancreas diagnosed by endoscopic ultrasound. *Endosc Int Open* 2020;8:E25–E28.
77. Collins K, Buckley T, Anderson K, Karasik M, Ligato S. Perivascular epithelioid cell tumor (PEComa) of pancreas diagnosed preoperatively by endoscopic ultrasound–guided fine-needle aspiration: a case report and review of literature. *Diagn Cytopathol* 2017;45:59–65.
78. Uno K, Shimizu S, Hayashi K, et al. Perivascular epithelial cell tumor of the pancreas diagnosed preoperatively by endoscopic ultrasound–guided fine-needle aspiration. *Intern Med* 2019;58:2515–2521.
79. Almousa MA, Alnashwan YA, Amr SS. Perivascular epithelioid cell tumor (PEComa) of the pancreas in a patient with ulcerative colitis: a case report and review of the literature. *Healthcare (Basel)* 2023;11:547.
80. Zhang S, Chen F, Huang X, et al. Perivascular epithelial cell tumor (PEComa) of the pancreas: a case report and review of literature. *Medicine (Baltimore)* 2017;96:e7050.
81. Jiang H, Ta N, Huang XY, et al. Pancreatic perivascular epithelioid cell tumor: a case report with clinicopathological features and a literature review. *World J Gastroenterol* 2016;22:3693–3700.
82. Mizuuchi Y, Nishihara K, Hayashi A, et al. Perivascular epithelial cell tumor (PEComa) of the pancreas: a case report and review of previous literatures. *Surg Case Rep* 2016;2:59.
83. Durczynski A, Wiszniewski M, Olejniczak W, Polkowski M, Sporny S, Strzelczyk J. Asymptomatic solid pancreatic hamartoma. *Arch Med Sci* 2011;7:1082–1084.
84. Kawakami F, Shimizu M, Yamaguchi H, et al. Multiple solid pancreatic hamartomas: a case report and review of the literature. *World J Gastrointest Oncol* 2012;4:202–206.
85. Katayama H, Azuma K, Koneri K, et al. A typical case of resected pancreatic hamartoma: a case report and literature review on imaging and pathology. *Surg Case Rep* 2020;6:107.
86. Zhou B, Li G, Xu S, Zhan C, Zheng X, Yan S. Pancreatic lipomatous hamartoma mimicking other pancreatic tumor: a case report and literature review. *Am J Transl Res* 2020;12:6682–6688.
87. Cui H, Lian Y, Chen F. Imaging findings for pancreatic hamartoma: two case reports and a review of the literature. *BMC Gastroenterol* 2020;20:37.
88. Noguchi T, Ryozawa S, Mizuide M, et al. Pancreatic hamartoma difficult to diagnose preoperatively. *Intern Med* 2021;60:2055–2059.
89. Matsushita D, Kurahara H, Mataki Y, et al. Pancreatic hamartoma: a case report and literature review. *BMC Gastroenterol* 2016;16:3.
90. Ahn AR, Song JS, Do Yang J, Moon WS. Pancreatic hamartoma mimicking neuroendocrine tumor. *Pathol Int* 2021;71:789–791.
91. Jarry J, Belleannee G, Laurent C, Coindre JM, Evrard S. Primary malignant fibrous histiocytoma of the pancreas: benefit of the multidisciplinary approach. *Eur J Gastroenterol Hepatol* 2010;22:765–768.
92. Basturk O, Askan G. Benign tumors and tumorlike lesions of the pancreas. *Surg Pathol Clin* 2016;9:619–641.
93. Liu HK, Lin YC, Yeh ML, Chen YS, Su YT, Tsai CC. Inflammatory myofibroblastic tumors of the pancreas in children: a case report and literature review. *Medicine (Baltimore)* 2017;96:e5870.
94. Baiao JM, Martins RM, Correia JG, et al. Inflammatory pseudotumor of the pancreas mimicking a pancreatic neoplasm. *Case Rep Gastroenterol* 2019;13:245–252.
95. Lim K, Cho J, Pak MG, Kwon H. Inflammatory myofibroblastic tumor of the pancreas: a case report and literature review. *Taehan Yongsang Uihakhoe Chi* 2020;81:1497–1503.
96. Chen ZT, Lin YX, Li MX, Zhang T, Wan DL, Lin SZ. Inflammatory myofibroblastic tumor of the pancreatic neck: a case report and review of literature. *World J Clin Cases* 2021;9:6418–6427.
97. Chang WT, Lee KT, Yang SF. Cavernous hemangioma of the pancreas: report of a case. *Pancreas* 2003;26:310–312.
98. Mondal U, Henkes N, Henkes D, Rosenkranz L. Cavernous hemangioma of adult pancreas: a case report and literature review. *World J Gastroenterol* 2015;21:9793–9802.
99. Kobayashi H, Itoh T, Murata R, Tanabe M. Pancreatic cavernous hemangioma: CT, MRI, US, and angiography characteristics. *Gastrointest Radiol* 1991;16:307–310.
100. Mundinger GS, Gust S, Micchelli ST, Fishman EK, Hruban RH, Wolfgang CL. Adult pancreatic hemangioma: case report and literature review. *Gastroenterol Res Pract* 2009;2009:839730.
101. Jin C, Mo JG, Jiang H, Wang LZ, Zou H, Wang KP. Adult pancreatic hemangioma: a rare case report and literature review. *BMC Surg* 2020; 20:118.
102. Zhou J, Chen Z. Rare adult pancreatic hemangioma: review of the literature with a case report. *Gland Surg* 2020;9:1596–1604.
103. Jarboui S, Salem A, Gherib BS, et al. Hemangioma of the pancreas in a 60-year-old woman: a report of a new case. *Gastroenterol Clin Biol* 2010;34:569–571.
104. Weidenfeld J, Zakai BB, Faermann R, Barshack I, Aviel-Ronen S. Hemangioma of pancreas: a rare tumor of adulthood. *Isr Med Assoc J* 2011;13:512–514.
105. Raymundo SRO, Hussain KMK, Hussein KG, Kuga ML. Rare case of adult pancreatic haemangioma and literature review. *BMJ Case Rep* 2018;2018:bcr2018226456.
106. Elliott TE, Albertazzi VJ, Danto LA. Pancreatic liposarcoma: case report with review of retroperitoneal liposarcomas. *Cancer* 1980;45:1720–1723.
107. Heywood G, Smyrk TC, Donohue JH. Primary angiomyolipoma of the pancreas. *Pancreas* 2004;28:443–445.
108. Meeks M, Grace S, Veerapong J, et al. Primary angiosarcoma of the pancreas. *J Gastrointest Cancer* 2017;48:369–372.
109. Faria A, Lopes F, Figueira A, Miranda C, Coutinho J. Pancreas angiosarcoma—case report of a rare cause of abdominal pain. *Int J Surg Case Rep* 2020;76:116–120.
110. Supandi FN, Singh B, Thamutaram H, Lim KF, Yusoff AR. Pancreatic angiosarcoma: a case report. *Ann Hepatobiliary Pancreat Surg* 2021; 25(Suppl 1):S388.
111. Nguyen CL, Vu TK, Nguyen HH, et al. Pancreatic angiosarcoma with synchronous pancreatic ductal adenocarcinoma leading to hemosuccus pancreaticus: a surgical case report and review of literature. *Ann Med Surg (Lond)* 2022;76:103547.
112. Whitlock RS, Ebare K, Cheng LS, et al. Angiosarcoma of the pancreas in a pediatric patient with an activating KDR-internal tandem duplication: a case report and review of the literature. *J Pediatr Hematol Oncol* 2022; 44:e751–e755.
113. Nakamura Y, Inui K, Yoshino J, et al. Inflammatory myofibroblastic tumor (inflammatory fibrosarcoma) of the pancreas: a case report. *Hepatogastroenterology* 2005;52:625–628.
114. Nomelini RS, Jammal MP, Fernandes PC Jr., de Carvalho MM, Saldanha JC, Murta EFC. Myofibroblastic inflammatory breast tumor and fibrosarcoma: a case report. *Eur J Gynaecol Oncol* 2011;32:551–553.
115. Xia W, Yang Y, Huang Y. Imaging features of sclerosing epithelioid fibrosarcoma of the pancreas: a case report. *Front Oncol* 2020;10:901.
116. Kramer SP, Bowman CJ, Wang ZJ, et al. Hybrid low-grade fibromyxoid sarcoma and sclerosing epithelioid fibrosarcoma of the pancreas. *J Gastrointest Cancer* 2020;51:1025–1029.
117. Menges M, Pees HW. Kaposi's sarcoma of the pancreas mimicking pancreatic cancer in an HIV-infected patient. Clinical diagnosis by detection of HHV 8 in bile and complete remission following antiviral and cytostatic therapy with paclitaxel. *Int J Pancreatol* 1999;26:193–199.
118. Pantanowitz L, Dezube BJ. Kaposi sarcoma in unusual locations. *BMC Cancer* 2008;8:190.

119. Sultan I, Qaddoumi I, Yaser S, Rodriguez-Galindo C, Ferrari A. Comparing adult and pediatric rhabdomyosarcoma in the surveillance, epidemiology and end results program, 1973 to 2005: an analysis of 2,600 patients. *J Clin Oncol* 2009;27:3391–3397.
120. Amer KM, Thomson JE, Congiusta D, et al. Epidemiology, incidence, and survival of rhabdomyosarcoma subtypes: SEER and ICES database analysis. *J Orthop Res* 2019;37:2226–2230.
121. Pallio S, Crino SF, Maida M, et al. Endoscopic ultrasound advanced techniques for diagnosis of gastrointestinal stromal tumours. *Cancers (Basel)* 2023;15.
122. Akbulut S, Yavuz R, Otan E, Hatipoglu S. Pancreatic extragastrointestinal stromal tumor: a case report and comprehensive literature review. *World J Gastrointest Surg* 2014;6:175–182.
123. Elgeidie A, El-Magd EA, El-Maaty SRA, El-Hawary AK. Pancreatic gastrointestinal stromal tumor: a case report. *Int J Surg Case Rep* 2016; 29:67–70.
124. Kwon HJ. Extra-gastrointestinal stromal tumor of the pancreas: report of a case. *Ann Hepatobiliary Pancreat Surg* 2017;21:237–242.
125. Bonetti F, Pea M, Martignoni G, Zamboni G. PEC and sugar. *Am J Surg Pathol* 1992;16:307.
126. Hornick JL, Fletcher CD. PEComa: what do we know so far? *Histopathology* 2006;48:75–82.
127. Zamboni G, Pea M, Martignoni G, et al. Clear cell "sugar" tumor of the pancreas. A novel member of the family of lesions characterized by the presence of perivascular epithelioid cells. *Am J Surg Pathol* 1996;20:722–730.
128. Nogueira Sixto M, Carracedo Iglesias R, Estevez Fernandez S, Rodriguez Pereira C, Sanchez Santos R. Pancreatic PEComa, a not so uncommon neoplasm? Systematic review and therapeutic update. *Gastroenterol Hepatol* 2024;47:93–100.
129. Folpe AL, Mentzel T, Lehr HA, Fisher C, Balzer BL, Weiss SW. Perivascular epithelioid cell neoplasms of soft tissue and gynecologic origin: a clinicopathologic study of 26 cases and review of the literature. *Am J Surg Pathol* 2005; 29:1558–1575.
130. Al-Haddad M, Cramer HM, Muram T, Wang X, Pitt HA. Perivascular epithelioid cell tumor: an unusual pancreatic mass diagnosed by EUS-FNA. *Gastrointest Endosc* 2013;78:165; discussion 165-167–167.
131. Okuwaki K, Kida M, Masutani H, et al. A resected perivascular epithelioid cell tumor (PEComa) of the pancreas diagnosed using endoscopic ultrasound–guided fine-needle aspiration. *Intern Med* 2013;52:2061–2066.
132. Walsh SV, Evangelista F, Khettry U. Inflammatory myofibroblastic tumor of the pancreaticobiliary region: morphologic and immunocytochemical study of three cases. *Am J Surg Pathol* 1998;22:412–418.
133. Martin-Broto J, Mondaza-Hernandez JL, Moura DS, Hindi N. A comprehensive review on solitary fibrous tumor: new insights for new horizons. *Cancers (Basel)* 2021;13:2913.
134. Demicco EG, Wagner MJ, Maki RG, et al. Risk assessment in solitary fibrous tumors: validation and refinement of a risk stratification model. *Mod Pathol* 2017;30:1433–1442.
135. Colvin JS, Morris-Stiff G, Cruise M, Purysko A. Pancreatic metastasis from an osseous solitary fibrous tumour. *BMJ Case Rep* 2017;2017: bcr2017220114.
136. Haas RL, Walraven I, Lecointe-Artzner E, et al. Radiation therapy as sole management for solitary fibrous tumors (SFT): a retrospective study from the global SFT initiative in collaboration with the sarcoma patients EuroNet. *Int J Radiat Oncol Biol Phys* 2018;101:1226–1233.
137. Kasper B, Baumgarten C, Garcia J, et al. An update on the management of sporadic desmoid-type fibromatosis: a European consensus initiative between sarcoma PAtients EuroNet (SPAEN) and European Organization for Research and Treatment of Cancer (EORTC)/Soft Tissue and Bone Sarcoma Group (STBSG). *Ann Oncol* 2017;28:2399–2408.
138. Khan M, Almond M, Ford S, Desai A. Evaluation of outcomes in the management of primary sporadic desmoid-type fibromatosis at a specialist soft tissue sarcoma unit. *Eur J Med Res* 2022;27:123.
139. Nieuwenhuis MH, Mathus-Vliegen EM, Baeten CG, et al. Evaluation of management of desmoid tumours associated with familial adenomatous polyposis in Dutch patients. *Br J Cancer* 2011;104:37–42.
140. Tsukamoto Y, Imakita M, Nishitani A, Ito T, Izukura M, Hirota S. Pancreatic desmoid-type fibromatosis with beta-catenin gene mutation—report of a case and review of the literature. *Pathol Res Pract* 2016;212: 484–489.
141. Litchinko A, Brasset C, Tihy M, Amram ML, Ris F. Large Desmoid tumor of the pancreas: a report of a rare case and review of the literature. *Am J Case Rep* 2022;23:e937324.