



Learning Objectives

- To understand typical imaging criteria to identify and differentiate solid and cystic pancreatic structural changes and neoplasia.
- To understand the limitations of imaging in complex pancreatic diseases and to appreciate the importance of additional clinical information.

Modern cross-sectional imaging with high spatial and contrast resolution allows a perfect delineation of the pancreas in its retroperitoneal home. The organ typically presents itself with a length between 12 and 15 cm and a diameter at the head area of about 2.5 cm, at the body of about 2 cm, and at the tip of the pancreatic tail of about 1.5 cm. Anatomically, the pancreatic head is defined as the area to the right of the left border of the superior mesenteric vein, the body as the area between the left border of the superior mesenteric vein and the left border of the aorta, and the tail as the area between left border of the aorta and the hilum of the spleen. The normal pancreatic duct ranges between 1.5 mm at the tail to 3 mm at the head.

Usually (ca. 60% of cases) the pancreatic main duct (duct of Wirsung), the duct of Santorini, and the common bile duct join together within the pancreatic head, entering the duodenum via the papilla of Vater.

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Several conditions that affect the function and integrity of the pancreas, as developmental anomalies, neoplastic and inflammatory diseases will be discussed.

14.1 Developmental Anomalies of the Pancreas

During embryogenesis, the pancreas is formed from a larger, dorsal bud (tail, body, parts of the head) and a small, ventral bud (rest of the head). The ventral bud migrates downwards dorsal from the dorsal bud. During the union of the both buds, the main pancreatic duct within the ventral bud ends via the duct of Santorini in the minor papilla. This duct gets then reduced to an accessory duct, whereas the main pancreatic duct of the dorsal bud merges with the duct of the former ventral bud ending in the major papilla [1, 2]. The disturbed union of the two buds can cause three major anomalies.

Pancreas divisum, a non-union anomaly of the pancreas, is found in autopsy studies with a frequency of 1–14% and is characterized by the separate drainage of the main pancreatic duct via the duct of Santorini into the minor papilla and of the duct of Wirsung into the major papilla. Only 1% of individuals with pancreas divisum will develop unspecific abdominal symptoms (abdominal discomfort, most likely caused by recurrent episodes of mild pancreatitis). Therefore—without real proof—some authors consider pancreas divisum a promoting factor for pancreatic tumors based on recurrent and lately chronic focal pancreatitis [3].

In pancreas annulare, the non-migration of the ventral bud of the pancreas causes the ventral and dorsal bud forming a ring around the duodenum. This rare anomaly (estimated prevalence 0.01%) can be associated with other birth deformities as congenital duodenal atresia, mesenterium commune, oral facial defects, and Down’s syndrome. Clinical signs are determined by stenosis and occlusion of the duodenum.

To reveal a union/migration anomaly of the pancreas, in most of the cases, MRCP will add the crucial information of the distorted duct configuration.

The generally asymptomatic ectopic pancreatic tissue can be found in the stomach, duodenum, and ileum, very rarely in Meckel's diverticulum, gall bladder, bile duct, and spleen, whereas autopsy studies reveal a frequency between 0.6 and 15%. Typically, pancreatic ectopic tissue is detected by endoscopy.

Total agenesis of the pancreatic gland, hypoplasia of the pancreas (partial agenesis), congenital pancreatic cysts (dysontogenetic cysts, hamartosis), multiple congenital cysts associated with von Hippel-Lindau disease (cysts also in the liver and kidneys), and also cystic degenerative transformation of the pancreas in cystic fibrosis are in general rare and are identified by MRI, as well as by sonography and CT, based on the partial or complete missing of the organ or by solitary or multiple cysts [2, 4].

Key Point

- The majority of pancreatic anomalies are asymptomatic. MRI and MRCP are superior in identifying the structural variants and to exclude suspected neoplastic conditions.

14.2 Pancreatic Neoplasms

Pancreatic tumors can be classified according to their cellular origin, enzymatic activity, and benign or malignant potential. The most recent WHO classification (2010, revised 2012 and 2017) divides pancreatic tumors into primary epithelial and mesenchymal tumors, lymphomas, and secondary tumors; from a clinical-practical point of view, tumorlike lesions can be added (Table 14.1). In clinical reality, many of the rare and very rare tumors have no specific imaging appearance and can be differentiated only pathologically.

14.2.1 Pancreatic Carcinoma

Pancreatic ductal adenocarcinoma—most of the various subtypes of pancreatic carcinoma can be differentiated only by histo- and immunopathology—accounts for 85–95% of all malignant pancreatic tumors (15–20% in gastrointestinal malignancies, 3% in all carcinomas). In general, the tumors are located predominantly in the pancreatic head (60–70%; body, 15% and tail, 5%). A multifocal or diffuse tumor spread is uncommon. The prognosis is poor, since most tumors are detected late in an advanced stage of spread. An early metastatic spread along perivascular, ductal, lymphatic, and perineural pathways is promoted by the absence of a true capsule around the organ.

For detection, staging, and follow-up after treatment, endoscopic ultrasound, contrast-enhanced CT, MRI, and FDG/PET

Table 14.1 Classification of pancreatic lesions modified according to WHO classification, pancreas (modified according to [5] and the 2017 update for neuroendocrine tumors [6])

Epithelial tumors	
Benign	Acinar cell cystadenoma Serous cystadenoma , not otherwise specified (NOS)
Premalignant lesions	Pancreatic intraepithelial neoplasia, grade 3 (PanIN-3) Intraductal papillary mucinous neoplasm (IPMN) with low- or intermediate-grade dysplasia Intraductal papillary mucinous neoplasm (IPMN) with high-grade dysplasia Intraductal tubulopapillary neoplasm (ITPN) Mucinous cystic neoplasm (MCN) with low- or intermediate-grade dysplasia Mucinous cystic neoplasm (MCN) with high-grade dysplasia
Malignant lesions	Ductal adenocarcinoma Adenosquamous carcinoma Mucinous adenocarcinoma Hepatoid carcinoma Medullary carcinoma, NOS Signet ring cell carcinoma Undifferentiated carcinoma Undifferentiated carcinoma with osteoclast-like cells Acinar cell carcinoma Acinar cell cystadenocarcinoma Intraductal papillary mucinous carcinoma (IPMN) with an associated invasive carcinoma Mixed acinar-ductal carcinoma Mixed acinar-neuroendocrine carcinoma Mixed acinar-neuroendocrine-ductal carcinoma Mixed ductal-neuroendocrine carcinoma Mucinous cystic neoplasm (MCN) with an associated invasive carcinoma Pancreatoblastoma Serous cystadenocarcinoma , NOS Solid pseudopapillary neoplasm
Neuroendocrine neoplasms	Nonfunctioning (nonsyndromic) neuroendocrine tumors (PanNEN G1/G2/G3) Pancreatic neuroendocrine microadenoma Nonfunctioning pancreatic neuroendocrine tumor Functioning (syndromic) neuroendocrine tumors (PanNEN G1/G2/G3) Insulinoma Glucagonoma Somatostatinoma Gastrinoma VIPoma Serotonin-producing tumors with and without carcinoid syndrome ACTH-producing tumor with Cushing syndrome Pancreatic neuroendocrine carcinoma (PanNEC G3, poorly differentiated neuroendocrine neoplasm) Mixed neuroendocrine-nonneuroendocrine neoplasms (MiNEN) Mixed ductal-neuroendocrine carcinoma Mixed acinar-neuroendocrine carcinoma

Table 14.1 (continued)

Epithelial tumors	
Mesenchymal tumors	Lymphangioma, NOS
	Lipoma, NOS
	Solitary fibrous tumor
	Ewing sarcoma
	Desmoplastic small round cell tumor
	Perivascular epithelioid cell neoplasm
Lymphomas	Diffuse large B-cell lymphoma (DLBCL), NOS
Secondary tumors	Metastases
Tumor-like lesions	Acute pancreatitis
	Chronic pancreatitis
	Groove pancreatitis
	Autoimmune pancreatitis
	Cystic lesions
	Pancreas divisum
	Pancreas annulare

may be applied, with endoscopic ultrasound having the highest accuracy in detecting small pancreatic head and periampullary tumors and FDG/PET in detecting distant metastatic spread. Nevertheless, CECT and MRI provide a sufficient and comprehensive display of the primary tumor and its sequelae with an accuracy of about 90% and even more [7–9].

The imaging appearance of common pancreatic adenocarcinoma is determined by its typically dense, fibrous, low-vascularized stroma resulting in low soft-tissue density in CT and low signal on T1-weighted and T2-weighted MRI and no or only minor contrast enhancement (Fig. 14.1) which makes the tumors best delineable to the normal glandular parenchyma on CE imaging.

The pancreatic duct may be involved depending on the primary tumor localization within the pancreas ranging from no duct involvement at all in peripheral tumor, over segmental obstruction due to intraductal tumor invasion (duct penetrating sign), to obstruction of both the pancreatic and common bile duct (double duct sign) in pancreatic head tumors. The relation between tumor and ducts is noninvasively seen best on MRCP.

Assessing potential invasive local growth, metastatic spread to the local and regional lymph nodes, to the liver, and to the vascular invasion, completes staging of pancreatic malignancies.

Not well-defined tumor margins and blurred surroundings are still a challenge for every imaging modality since microscopic local invasive peritumoral spread and an inflammatory desmoplastic reaction can often not be differentiated causing over- or underestimation of the T-stage of the tumor [10].

At the time of diagnosis of the primary, about two thirds of the patients will present distant metastases (lymph node metastases 40%, hematogenous metastases of the liver 40%, peritoneal metastases 35%) which will be detected with accuracies above 90% by CE-MRI and FDG/PET/CT [11, 12]. Non-resectability in pancreatic cancer is determined by vascular encasement of the superior mesenteric artery, celiac trunk, hepatic or splenic artery, and peripancreatic veins, which is

very likely if a vessel circumference is encased more than 50% (typical signs: decreased vessel caliber, dilated peripancreatic veins, teardrop shape of superior mesenteric vein present).

14.2.2 Other Tumors of Ductal Origin

This heterogeneous group of tumors embrace cystic neoplasms, tumors neuroendocrine components, and a variety of very rare tumors as pancreatoblastoma and solid pseudopapillary neoplasm.

Key Point

- Pancreatic adenocarcinoma is the most common malignancy of the pancreas. CT and MRI are the established imaging tools for primary diagnosis, staging the extent of the disease, and establishing operability.

14.3 Cystic Neoplasm

In modern high-resolution imaging, pancreatic cysts are a common finding by MRI (~20%) and CT (~3%). Due to the slightly increased risk of malignancy in incidental cysts, mainly in the younger than 65 of years, incidentally found pancreatic cysts have to be assessed carefully without exaggerating unnecessary therapeutical consequences [13, 14].

14.3.1 Serous Cystadenoma

Serous cystic neoplasms are accounting for about 50% of all cystic tumors including serous cystadenomas, serous oligocystic adenomas, cystic lesions in von Hippel-Lindau syndrome, and rarely serous cystadenocarcinomas [15, 16].

The most common subtype is the benign serous cystadenoma (microcystic type), typically in elderly women (60–80 years of age). In most cases, the lesion is located in the pancreatic head, composed of multiple tiny cysts, separated by thin septae. Spotty calcifications and a central stellate nidus might be present (Fig. 14.2).

About 10% of all serous cystic tumors present as an oligocystic variant with only a few cysts of 2–20 mm diameter and a higher prevalence in men (30–40 years).

The rare cystadenocarcinomas are usually large at clinical presentation already with local invasive growth and metastases to lymph nodes and liver.

The diagnosis of serous cystic lesions of the pancreas by imaging is ruled by the proportion of small cysts and septae without contrast enhancement that may create an almost solid impression in CT, whereas the cystic components still can be best

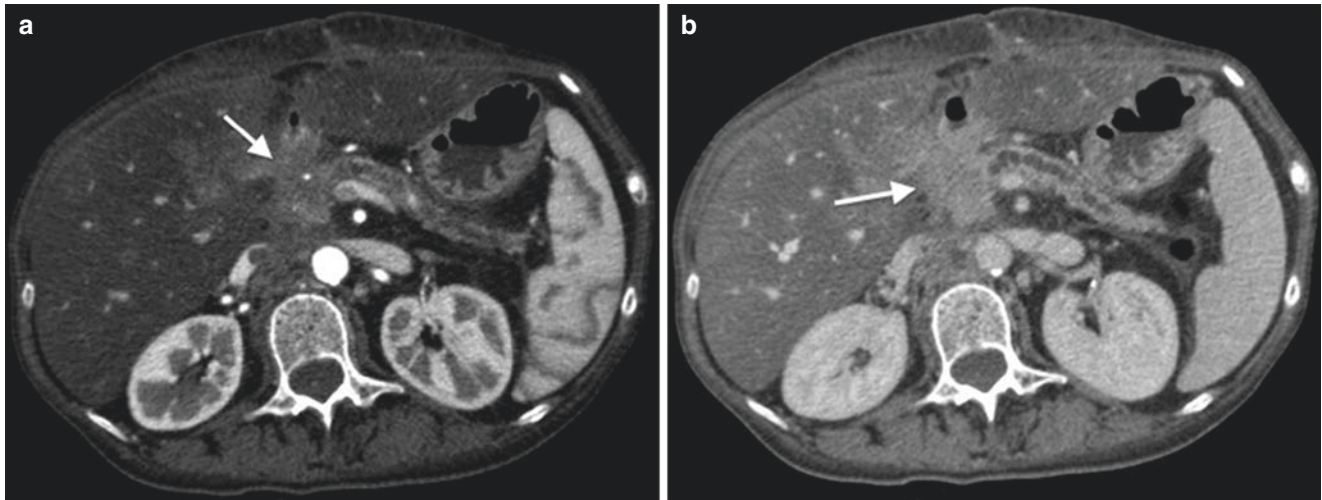


Fig. 14.1 (a, b) Adenocarcinoma of the head of the pancreas locally invasive. (a) Axial contrast-enhanced computed tomography (CT) during the pancreatic phase shows a hypovascular focal pancreatic lesion of the head, responsible of infiltration of the main pancreatic duct with

obstructive chronic pancreatitis and infiltration of the peripancreatic fat (arrow). (b) Axial contrast-enhanced computed tomography (CT) during the portal venous phase shows infiltration of the posterior peripancreatic fat

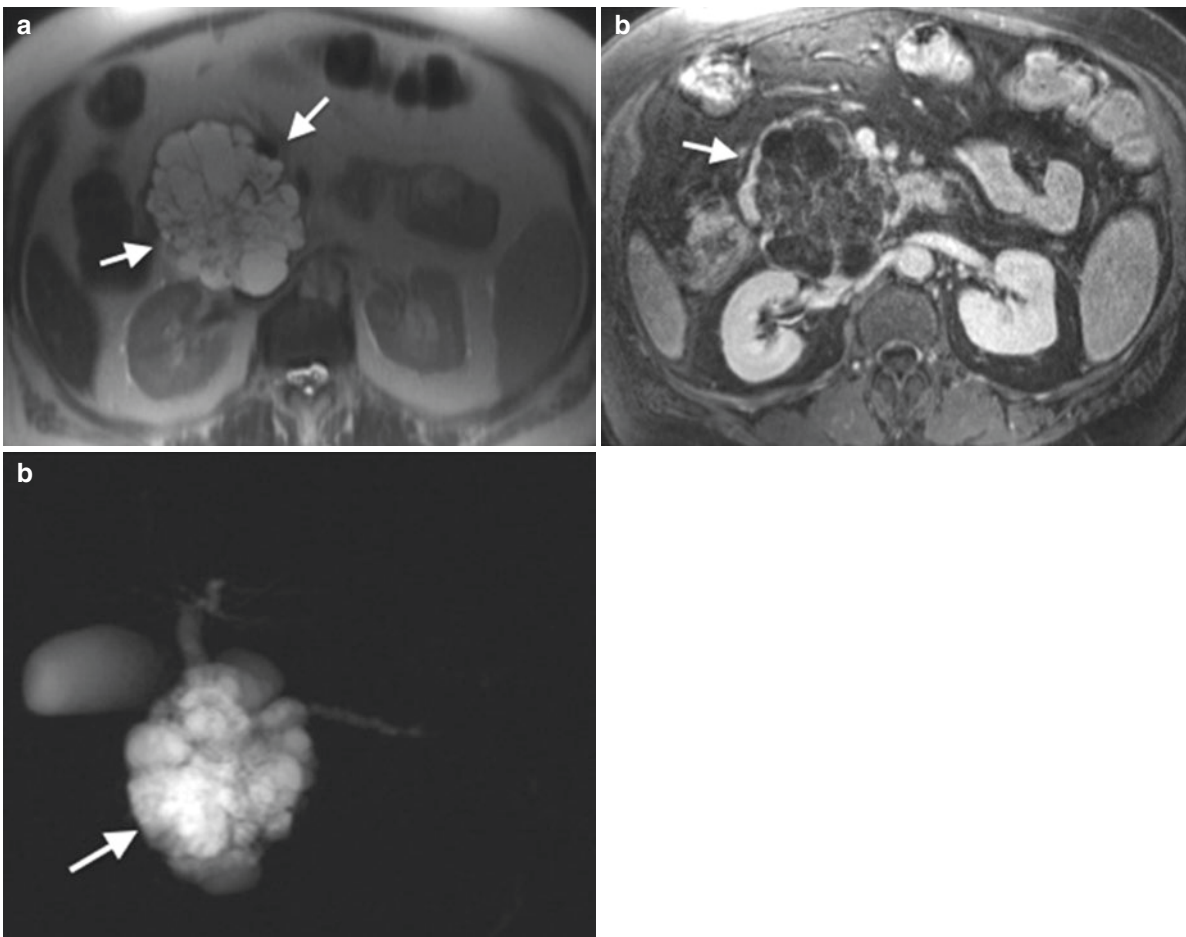


Fig. 14.2 (a–c) Serous cystadenoma. (a) Axial T2-weighted turbo spin echo image (TR/TE 4500/102) shows a multicystic microcystic neoplasm of the head of the pancreas (arrows). (b) On axial fat-saturated volumetric T1-weighted gradient echo image (TR/TE 4.86/1.87 ms) during the portal venous phase of the dynamic study following

Gd-chelate administration, serous cystadenoma shows enhancement of and lack of a peripheral wall. (c) On the coronal MRCP image single-shot RARE (TR/TE ∞ /110 ms), serous cystadenoma is responsible of compression of the main pancreatic duct with upstream dilatation

appreciated by MRI. Even if the tumors can grow rather large the mismatch of tumor size, missing both ductal involvement and secondary signs of malignancy will direct to the right diagnosis.

For the differentiation of oligocystic adenomas from mucinous cystic tumors, IPMN, or walled-off cysts tumor localization, an “empty” clinical history and normal ducts in MRCP can be helpful [17, 18].

14.3.2 Mucinous Cystic Neoplasm (MCN)

Mucin-producing cystic tumors, typically in middle-aged women (f:m = 19:1), are characterized by a missing connection to the pancreatic ducts and the histological presence of an ovarian-like stroma. In comparison to SCN, MCN is less frequent (10% of all cystic pancreatic lesions) in general asymptomatic, detected as solitary, large lesions arising in the body and tail of the pancreas (95%), and composed of only few cysts with pronounced septae. Since the cysts may contain mucinous, hemorrhagic, necrotic, jellylike content, they may present intermediate and higher densities and signal intensities on CT and MRI, whereas T2-weighted MRI displays the true cystic structure of the tumor the best. Nodular enhancement of the septae is indicating potential malignancy which occurs in up to 30% of MCN [17, 18].

14.3.3 Intraductal Papillary Mucinous Neoplasm (IPMN)

Due to increased detection rates by high-resolution imaging, IPMN is considered the most common cystic neoplasm of the pancreas, seen more often in men than in women.

IPMNs may affect the main duct (28%), side branches (46%), or both duct components (26%) based on a mucin-producing neoplasm arising from the ductal epithelium. The side-branch type can be found as a solitary or multifocal duct dilatation all over the pancreas and may also form a system of cystic dilated ducts that may mimic a microcystic appearance as in SCN. Segmental or general dilatation is typical for the main duct type creating a chronic pancreatitis-like appearance. In such cases patients' history is the crucial differential diagnostic information. Since the main duct-type IPMN has a low malignant potential, at least a thorough follow-up regimen should be recommend in nonsurgical cases {Lana, 2016 #1140;Lee, 2017 #1137}.

Key Point

- MRI is the superior imaging method allowing the detailed characterization of cystic lesions and neoplasia of the pancreas.

14.4 Other Neoplasm

14.4.1 Neuroendocrine Tumors

The WHO (2010, last modification 2017) classified these tumors mainly according to their grading (well, moderately, poor differentiated) and their hormonal activity (PanNEN, pancreatic neuroendocrine neoplasm), as well as the Ki67 proliferative index.

In general, these tumors are rare and account for about 5–7% of all pancreatic tumors with the most common subtypes being insulinoma, glucagonoma, and nonhormonal active tumors. Imaging features of various PanNEN are often rather similar, making immunohistochemical staining a crucial issue, particularly when a specific hormone release is not the leading clinical sign (Fig. 14.3) [19].

14.4.1.1 Insulinoma

The presentation of insulinomas—the most common PanNEN (60%)—is determined by hyperinsulinism (Whipple's triad: starvation attack, hypoglycemia after fasting, and relief by IV dextrose). The majority of tumors are solitary (95%) and small (<2 cm) and hypervascularized with a peripherally pronounced enhancement and localized in the pancreatic body and tail [20].

14.4.1.2 Gastrinoma

Gastrinoma is the second most common PanNEN (20–30%) clinically associated with the Zollinger-Ellison syndrome (peptic ulcer disease, diarrhea) due to the massively elevated gastrin blood levels. At detection, the tumors present with a moderate size (mean 3 cm, ranging from 0.1–20 cm) and in half of the cases with multiple nodules. The vast majority of gastrinomas will arise within the gastrinoma triangle determined by the confluence of the cystic and common bile duct, the junction of the second and third portions of the duodenum, and the junction of the neck and body of the pancreas. On imaging, gastrinomas are revealed as mainly solid tumors with intermediate densities and signal intensities on both CT and MRI with moderate to strong contrast enhancement. Even if about 60% of the tumors are malignant, extensive metastatic spread is rare [21].

14.4.2 Other Rare Pancreatic Neoplasm

Beside the above displayed neoplasms, there is still a wide variety of pancreatic tumors which—in general—can be differentiated only by specific immunohistological staining. This tumors comprise a number of variably differentiated neuroendocrine tumors including mixed neuroendocrine-non-neuroendocrine tumors, mostly without functional activity, rare malignant pancreatoblastoma in children (a large,

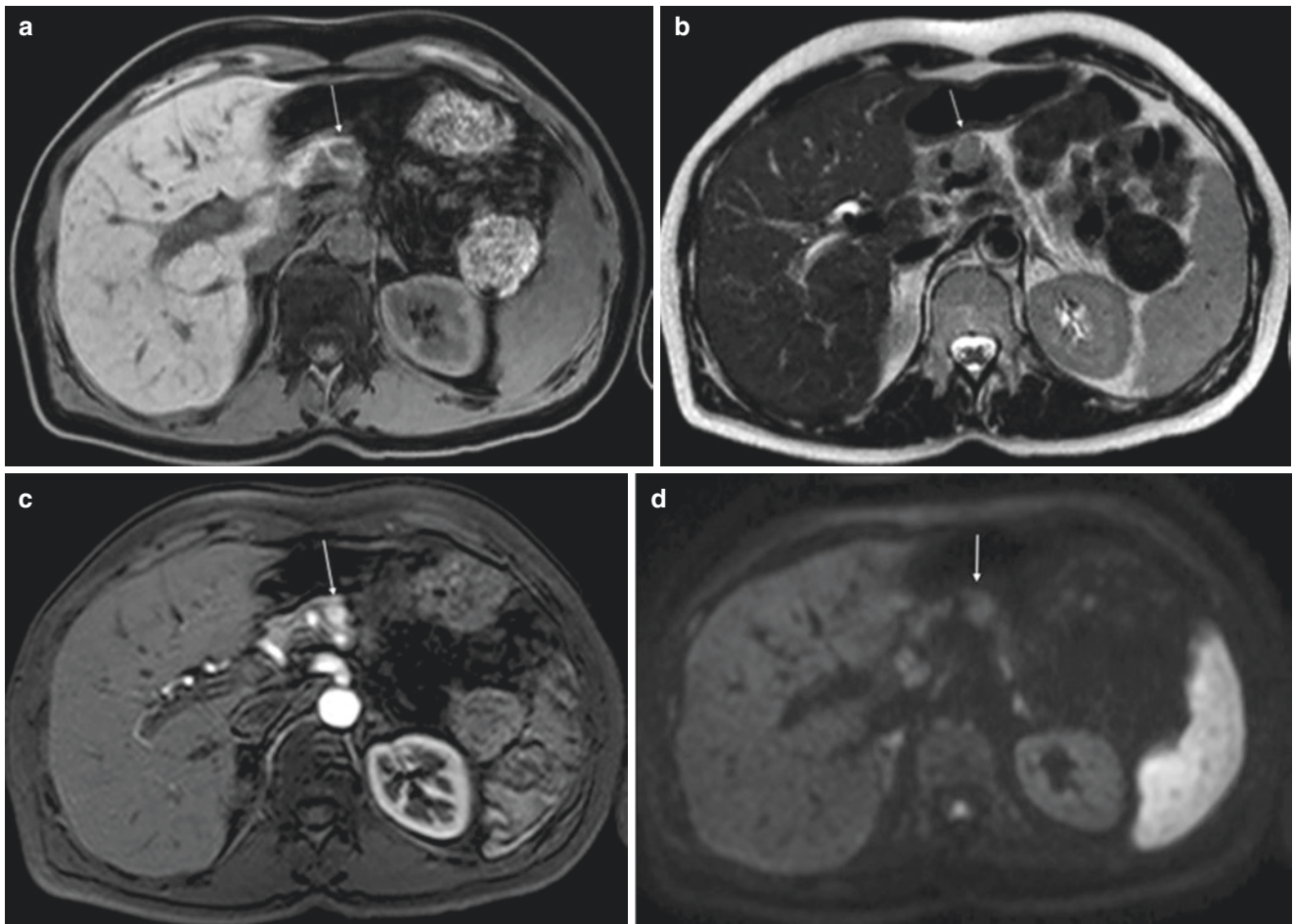


Fig. 14.3 (a–d) Small neuroendocrine neoplasm. (a) Axial T1-weighted gradient echo image (TR/TE 180/4.66 ms) with fat saturation shows a neuroendocrine neoplasm that appears hypointense compared to adjacent pancreatic parenchyma (arrow). (b) Axial T2-weighted turbo spin echo image (TR/TE 4500/102) shows a small neuroendocrine neoplasm that appears hyperintense compared to adjacent parenchyma (arrow). (c) On the axial fat-saturated volumetric T1-weighted

gradient echo image (TR/TE 4.86/1.87 ms) during the pancreatic phase of the dynamic study following Gd-chelate administration, the neuroendocrine neoplasm appears hyperintense during compared to adjacent pancreatic parenchyma (arrow). (d) On Axial diffusion-weighted image ($b = 1000$), the neuroendocrine neoplasm shows restricted diffusion (arrow)

encapsulated tumor in the pancreatic head often associated with elevated alpha-fetoprotein levels and metastases to liver and lymph nodes), acinar cell carcinoma (relatively large tumors in elderly men with an imaging appearance similar to pancreatic adenocarcinomas and potential excessive release of serum lipase followed by focal panniculitis and polyarthritides as diagnostic hint), and solid pseudopapillary tumor (of mainly young women (frequently incidental tumor in women 20–30 years of age; m: f = 1:10; large, heterogenous tumor of uncertain dignity) and occasionally children) [22].

Mesenchymal tumors (sarcoma, cystic dermoid, lymphangioma, leiomyosarcoma, hemangiopericytoma, hemangioma, malignant fibrous histiocytoma, lymphoepithelial cysts, primary lymphoma) and secondary tumors (secondary lymphoma, metastases) of the pancreas are very rare and may be identified due to specific imaging features such as peripheral nodular enhancement on dynamic

imaging or high signal intensity in T1- and T2-weighted imaging in, e.g., hemangioma or lipoma; otherwise clinical context and histopathological proof will determine the diagnosis.

Key Point

- PanNEN comprises a complex group of neoplasia which can be identified usually by imaging—beside very small tumors. Nevertheless, without clinical information and immunohistopathological correlation, a precise diagnosis is not possible. In malignant transformation, the mismatch between tumor size and missing secondary signs of malignant spread as common in pancreatic cancer can be helpful.

14.5 Inflammatory Diseases of the Pancreas

14.5.1 Acute and Chronic Pancreatitis

Especially in the Western world, the incidence of inflammatory diseases of the pancreas is increasing. The most common causes are biliary stone disease and alcohol abuse; nevertheless, more and more other causes as metabolic syndrome and systemic inflammatory diseases are identified as promoting factors. Depending on the type and severity of the inflammatory process, no mild or extensive morphological and functional deterioration is seen.

In general, the task of imaging is to monitor substantial structural changes and complications in acute pancreatitis, such

as parenchymal integrity vs. necrosis, peripancreatic inflammation, subtle and substantial fluid collections, formation of pseudocysts and walled-off cysts, and vascular and ductal affections (Fig. 14.4), and to assist in the clinical outcome prognosis together with the clinical assessment [23–25].

In chronic pancreatitis, differentiation of long-term parenchymal and ductal changes from similar changes caused by neoplasms—e.g., focal or complete duct dilation, focal parenchymal lesions, and cystic degeneration—is mandatory to rule out complications and potential pancreatic cancer (Fig. 14.5). Perfusion MRI, DWI, and FDG/PET can be helpful in such cases. Nevertheless, the common clinical presentation with chronic abdominal pain in chronic pancreatitis does not correlate very well with imaging findings [26–30].

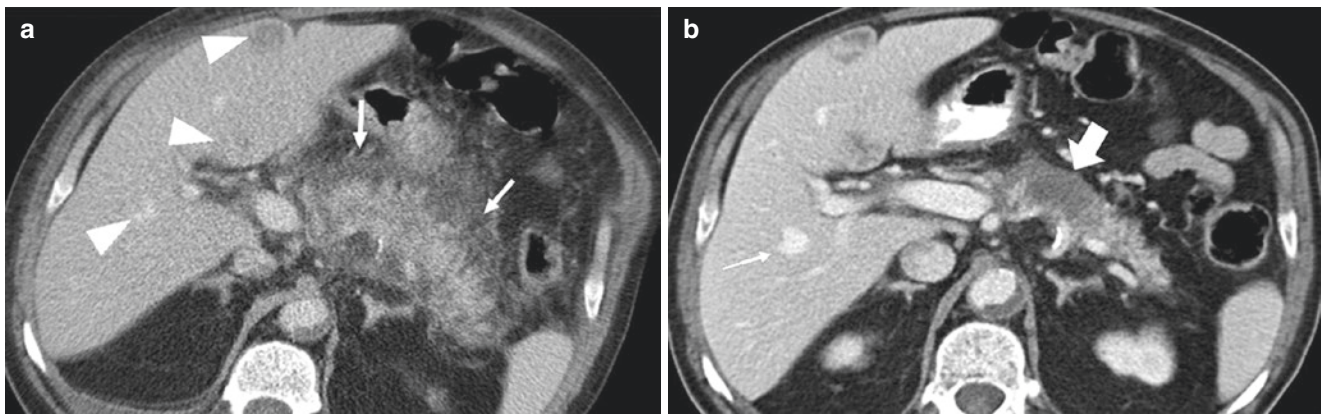


Fig. 14.4 Acute severe pancreatitis. At the patient's admission, contrast-enhanced CT during the venous phase (a) displayed a fuzzy contour of the pancreatic gland together with peripancreatic exudation (arrow). Note the hypo- and hyperdense hepatic lesions (arrow heads). A control scan 10 days later (b) revealed an almost normal gland with

resorption of the peripancreatic fluid. However, there was an area with a lack of enhancement representing focal necrosis (large arrow). In the liver, one lesion turned out to be a hemangioma (arrow) while the other two lesions were small abscesses

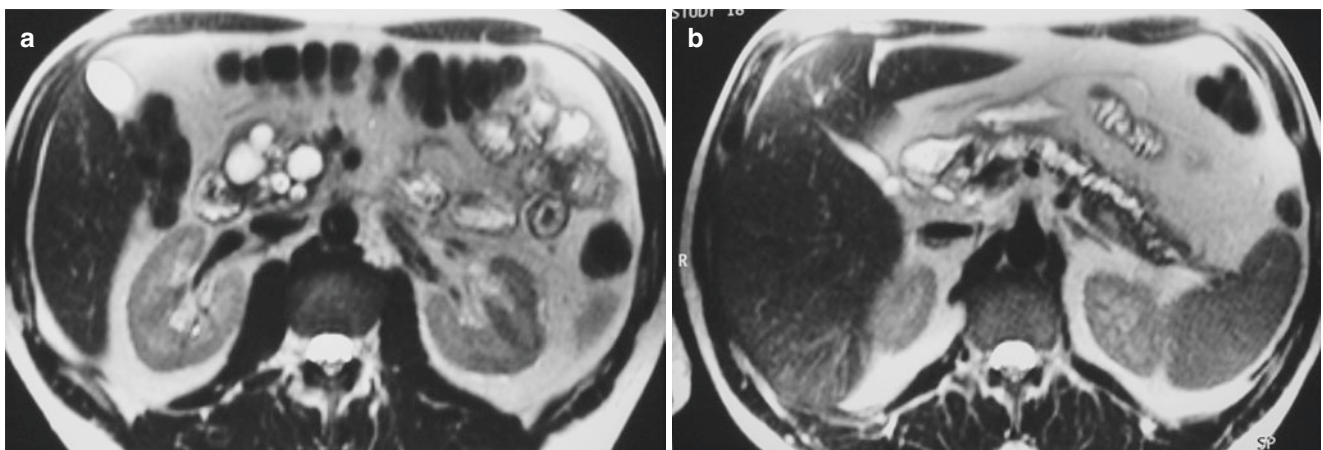


Fig. 14.5 Chronic pancreatitis. Cystic degeneration of the pancreatic head (a) together with irregular dilatation of the pancreatic main duct (b) in MRI (fast SE T2). Note the similar imaging appearance to other cystic lesions of the pancreas

14.5.2 Autoimmune Pancreatitis

In comparison to gallstone or alcohol-associated pancreatitis, autoimmune pancreatitis (AIP) is a rare disease in which the pathophysiological understanding has evolved significantly over the last years. The most common form, type 1 AIP, is associated with IgG4-related diseases. Type 2 AIP is a different, even rarer entity and may be associated to chronic inflammatory bowel disease [31, 32].

Both diseases have a similar clinical presentation with unspecific upper abdominal pain, obstructive jaundice, furthermore weight loss, and endo- and/or exocrine pancreatic insufficiency. Clinically, there is an overlap with pancreatic carcinoma, which cannot be solved by imaging alone, since AIP

may provide diffuse (“sausage”-like) or focal (“mass-forming”) enlargement of the gland together with segmental or focal duct strictures or dilatation. In consequence, the task of imaging and further parameters as serology and histology is the differentiation of both entities to guide each to the appropriate therapy and to avoid the small number but unnecessary pancreatectomies.

The International Association of Pancreatology defined diagnostic consensus criteria (Table 14.2) which provide a high accuracy in identifying an AIP. Both types of AIP usually present an excellent response to steroid therapy; however, in type 1 AIP, 60% of patients will have relapse. Over the last years, the body of knowledge in AIP was growing significantly; nevertheless, data for the long-term course and potential complications are still scarce (Fig. 14.6).

Table 14.2 Consensus criteria in type 1 and type 2 AIP (modified acc. to Shimosegawa et al. [33], Beyer et al. [34] and O'Reilly et al. [35]; L level, ERP endoscopic retrograde pancreatography, IgG4-RD IgG4-related disease, IBD inflammatory bowel disease, GEL granulocytic epithelial lesions)

	Type 1 AIP	Type 2 AIP		
Synonym	Lymphoplasmacellular sclerosing pancreatitis (LPSP)	Idiopathic ductal-centric pancreatitis (IDCP)		
Incidence	0.9/100.000 Das heißt Weihnachten 6. Jahrzehnt es... Sie Online leerzeichen Nach der Restauration Ich gehe! Bin ich jetzt gehen Hör mir verweilen und ich komm die Rechnung bitte Sieben jetziger Sicht drei Tage krank 3 Wochen krank Mehrfamilienhaus Ich will Risikoabschätzung Wie Computer? Aber vergessen Gegen dass damit der immerhin schon sind wir es ist irgendwie doch noch fristlos Selbst Der beste Weg herr valgus mir sagen welchen Fall			
Geographic frequency siehe	EU/USA 50%, Asia 95%	EU/USA 50%		
Age (years)	50–70	30–50		
Sex	M (75%) » F	M = F		
<i>Consensus criteria of the International Association of Pancreatology [33]</i>				
Cardinal criteria	Level 1	Level 2	Level 1	Level 2
Imaging: parenchyma	Typical: diffuse enlargement (“sausage sign”), delayed (interstitial) enhancement (sometimes capsule-like, nodular enhancement)	Indeterminate: focal enlargement with delayed enhancement	Typical: diffuse enlargement (“sausage sign”), delayed (interstitial) enhancement (sometimes capsule-like, nodular enhancement)	Indeterminate: focal enlargement with delayed enhancement Atypical: hypodense in CT, duct dilatations, atrophy
Imaging: pancreatic duct (validated for ERP; analog interpretation in MRCP)	Long (>1/3 of duct length) or multiple strictures without proximal (upstream) dilatation	Segmental/focal strictures with proximal (upstream) dilatation (<5 mm)	Long (>1/3 of duct length) or multiple strictures without proximal (upstream) dilatation	Segmental/focal strictures with proximal (upstream) dilatation (<5 mm)
Serology	IgG4 > 2 × upper limit	IgG4 1–2 × upper limit	–	–
Other organ involvement (OOI)	IgG4-related disease (RD) (50%) ≥3 histological findings in other organs: – Lymphoplasmacytic cellular infiltrates and fibrosis without granulocytes – Storiform fibrosis – Obliterating phlebitis – IgG4-positive cells (>10/HPF) ≥1 radiological finding: – Segmental/multiple bile duct strictures – Retroperitoneal fibrosis IgG4-RD (ca. 60%): – Chronic sclerosing sialadenitis (14–39%) – IgG4-associated cholangitis (IAC) (12–47%) – IgG4-associated tubulointerstitial nephritis and renal parenchymal lesions (35%) – Enlarged hilar pulmonary LN (8–13%) – Retroperitoneal fibrosis – Chronic thyroiditis – Prostatitis – Chronic inflammatory bowel disease (0.1–6%)	Bile duct involvement plus both: – Lymphoplasmacytic cellular infiltrates without granulocytes – IgG4-positive cells (>10/HPF) or ≥1 criterion (imaging or clinical exam): – Symmetric enlarged salivary glands – Renal involvement	No association with IgG4-RD – Histological and/or clinical diagnosis of inflammatory bowel disease (16%) – Involvement of proximal bile duct possible – Involvement of thyroid possible	
Histology (Trucut biopsy or resection, FNB not suitable)	Periductal lymphoplasmacellular infiltrations, inflammatory, cell-rich stroma 3 of 4 criteria – Storiform fibrosis – Obliterative phlebitis – Prominent lymphatic follicles – IgG4-positive plasma cells	2 of 4 criteria – Storiform fibrosis – Obliterative phlebitis – Prominent lymphatic follicles – IgG4-positive plasma cells	– GEL with or without granulocytic acinar infiltration No or few (10 < 10 HPF) IgG4-positive plasma cells	– Granulocytic and lymphoplasmacytic acinar infiltrate – No or few (>10 HPF) IgG4-positive plasma cells
Response to steroids	Rapid (≤ 2 weeks) response to therapy with significant improvement in imaging			
Relapse post steroids	20–60%		<10%	
Diagnosis based on criteria	Type 1 AIP		Type 2 AIP	
Definitive	Histo L1 + imaging L1/2 Imaging L1 + other criteria L1/2 Imaging L2 + ≥ 2 criteria L1 Steroid Response + Bildgebung L2 + 3 Kriterien L1 oder 4 Kriterien L2		Imaging L1/2 + histo L1 or IBD + histo L2 + response to steroids	
Probable	Imaging L2 + other criteria L2		Imaging L1/2 + histo L2 + IBD + response to steroids	

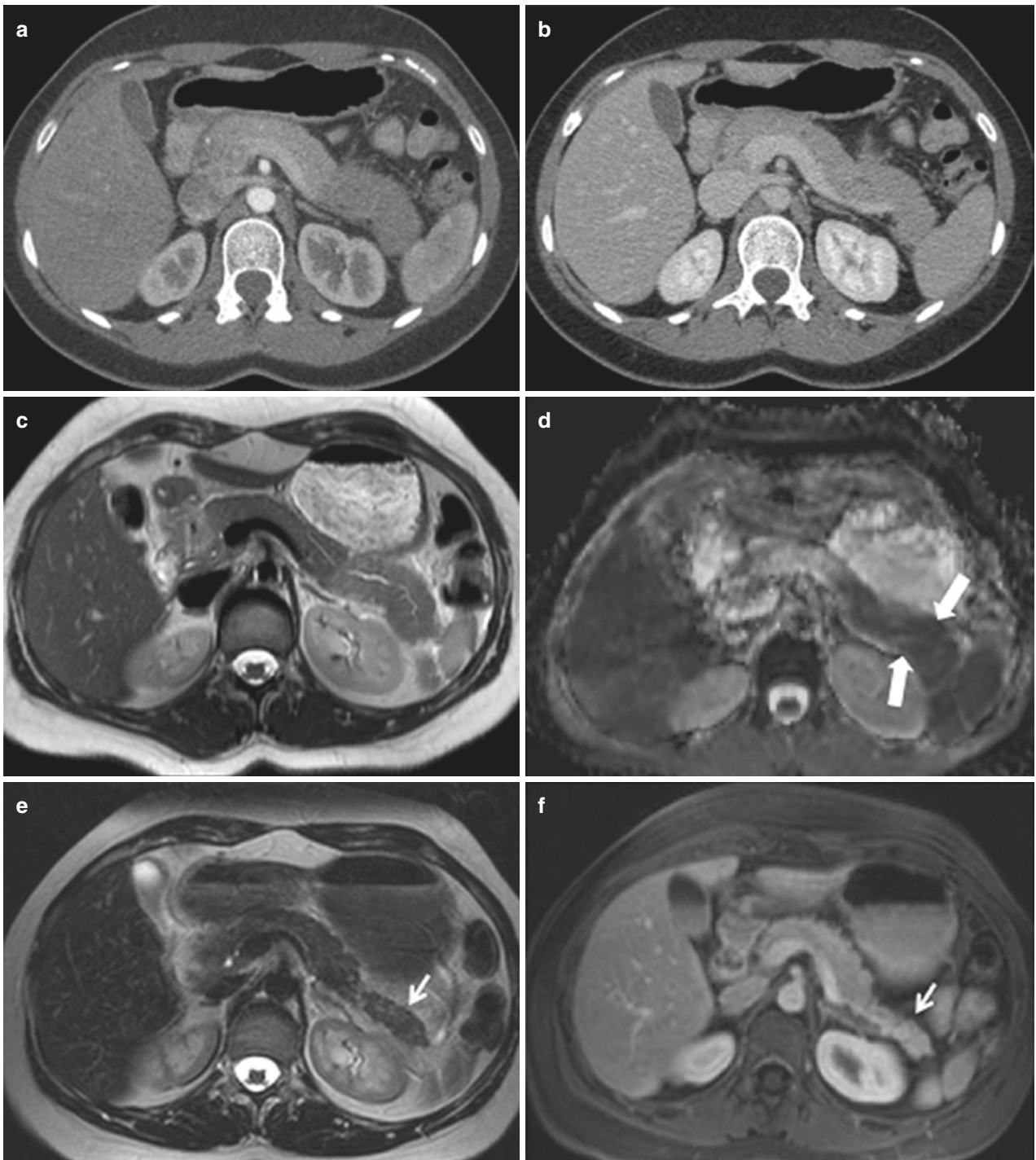


Fig. 14.6 Autoimmune pancreatitis type 1. Well-demarcated focal enlargement of the pancreatic tail on CT (**a, b**). Note the slightly reduced perfusion in the early parenchymal phase (**a**). Low signal intensity on T2-weighted MRI (**c**) and diffusion restriction on DWI

ADC map (**d**) reveals the lymphoplasmacytic infiltration with fibrotic components in contrast to edema in “usual” pancreatitis. After 6 weeks therapy with steroids, note the significant atrophy of the pancreatic tail on T2-weighted (**e**) and CE T1-weighted MRI (**f**)

Key Point

- Acute and chronic pancreatitis are common diseases, whereas the diagnosis is ruled by the clinical history and/or presentation. Imaging adds the crucial information on the severity and complications of the disease. In AIP imaging findings contribute to the cardinal criteria; however, imaging alone is not suitable to establish the diagnosis in AIP.

14.6 Concluding Remarks

Pancreatic lesions encompass a wide variety of anatomical variants as well as benign and malignant neoplastic and inflammatory diseases. The specific anatomical position of the gland allows only limited insight by ultrasound and endoscopy. Therefore, cross-sectional imaging by CT and MRI is of ample importance in assessing the pancreas and related disorders, allowing for a very high accuracy in depicting structural alterations of the parenchymal and ductal components of the gland. In the majority of clinical-diagnostic situations, there is no significant difference between the two imaging modalities with respect to diagnostic efficacy. However, MRI will reveal its superiority particularly in conditions where the assessment of ductal and intra- and peripancreatic cystic structures as well as subtle parenchymal changes is pivotal.

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