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PICTORIAL ESSAY

Imaging of congenital pancreatic lesions: emphasis on key imaging features

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Abstract Congenital pancreatic lesions are relatively uncommon, but they are frequently encountered during radiologic examination as an incidental finding in asymptomatic patients. However, some of these entities may produce symptoms such as abdominal pain, nausea, vomiting and gastric outlet obstruction. This article reviews normal pancreatic anatomy, imaging findings of congenital pancreatic lesions, including congenital pancreatic anomalies, congenital pancreatic ductal variants, pancreatic contour variants, congenital pancreatic cysts, and mimics of pancreatic lesions with emphasis on key imaging features.

Keywords Congenital pancreatic lesions · Pancreatic imaging · Imaging of congenital pancreatic disorders

Introduction

Congenital pancreatic lesions may be encountered in adulthood as well as in childhood and may be asymptomatic or significant. In this article we review normal pancreatic anatomy, imaging findings of congenital pancreatic anomalies, congenital pancreatic ductal variants, pancreatic contour variants, congenital pancreatic cysts and mimics of pancreatic lesions with emphasis on key imaging features (Table 1).

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Normal pancreatic anatomy

Pancreatic gland

The pancreas is a nonencapsulated gland that extends from the second part of the duodenum to the splenic hilar surface. The pancreas is typically coarsely lobulated, measures about 15–20 cm in length, is located in the retroperitoneal anterior pararenal space, and can be divided into four parts: uncinate process, head, body, and tail. The pancreatic neck is the constricted part between the pancreatic head and body. The pancreatic head is located to the right of the superior mesenteric vein (SMV). The uncinate process has a triangular shape with a straight or concave anteromedial border. The pancreatic body is arbitrarily divided from the tail using half of the distance between the pancreatic neck (about the level of the SMV) and the end of the pancreatic tail. There is a gradual decrease in size of the pancreas with age [1]. The normal diameter of the main pancreatic duct measures maximally 3.5 mm in the head, 2.5 mm in the body and 1.5 mm in the tail. There are numerous small side branches that enter at right angles from the pancreatic parenchyma. At the pancreaticobiliary junction, the duct of Wirsung joins with the common bile duct (CBD) and drains into the major papilla. The duct of Santorini or accessory duct drains the anterior and superior part of the pancreatic head via the minor papilla. Before entering the duodenum, the distal CBD and duct of Wirsung are encircled by the sphincter of Oddi, which typically measures 10–15 mm in length [2].

Vascular supply

The pancreas has a rich arterial supply from branches of celiac and superior mesenteric arteries. The pancreatic head receives arterial flow from two pancreaticoduodenal arcades

Table 1 Key imaging features of congenital pancreatic lesions

Pancreatic lesions	Key imaging features
Annular pancreas	Ring of pancreatic tissue encircling descending duodenum and presence of annular duct
Incomplete annular pancreas	Crocodile jaw appearance
Ectopic pancreas	Submucosal lesion with central umbilication with no anatomical connection to normal pancreas
Pancreatic hypoplasia	Short truncated pancreas, more frequently involved dorsal pancreas and dependent stomach or dependent intestine signs
Pancreatic divisum	Presence of dorsal duct passing anterior to the CBD
Anomalous pancreaticobiliary junction	Long common channel more than 15 mm
Duplication of pancreatic duct	Two main pancreatic ducts, common in the tail
Tuber omentale	Protuberance of pancreatic tissue, commonly at head-neck junction
Pancreatic cleft	Invagination of peripancreatic fat, mimics pancreatic fracture but asymptomatic
Congenital pancreatic cysts	Multiple pancreatic cysts, associated with ADPKD, VHL
Intrapancreatic accessory spleen	Well-defined nodule with similar appearance to the native spleen
Fatty replacement of pancreas	Diffuse or focal form with negative Hounsfield units

formed by anterior and posterior superior pancreaticoduodenal arteries from the common hepatic artery or gastroduodenal artery that joins the second pair of anterior and posterior inferior pancreaticoduodenal arteries from superior mesenteric artery. The neck, body and tail of the pancreas are supplied by dorsal pancreatic arteries that usually arise from the splenic artery [3]. All major arteries lie posterior to the pancreatic ducts. The pancreatic head drains into the superior mesenteric vein and portal vein. The rest of the pancreas drains into the splenic vein.

Congenital pancreatic anomalies

Annular pancreas

Annular pancreas is the second most common congenital abnormality of the pancreas after pancreas divisum and results from failure of the ventral bud to simultaneously rotate with the duodenum, resulting in envelopment of the duodenum by a ring of pancreatic tissue. The second part of the duodenum is encircled in 85 % of cases and the first or third part of the duodenum are encircled in the remaining 15 %.

Annular pancreas may be complete or incomplete. Complete annular pancreas usually presents early in life or in the neonatal period with vomiting owing to duodenal obstruction. Plain films show a double bubble appearance in typical cases. Approximately 70 % of infants with annular pancreas have associated congenital anomalies, such as duodenal stenosis or atresia, Down syndrome, and various congenital heart defects [4]. On computed tomography (CT) or magnetic resonance imaging (MRI), a ring of pancreatic tissue encircles the descending duodenum in continuity with pancreatic tissue (Fig. 1). Magnetic resonance cholangiopancreatography (MRCP) has taken the place of endoscopic retrograde cholangiopancreatography (ERCP) as it is a noninvasive tool which is able to identify anomalous pancreatic ducts and, where ERCP fails in particular, duodenal obstruction. On MRCP, diagnosis is confirmed by demonstration of an annular pancreatic duct encircling and extending to the right side of the duodenum. However, a complete ring of pancreatic tissue is not required to establish a diagnosis of annular pancreas, and this is termed incomplete annular pancreas. Incomplete annular pancreas is diagnosed by the presence of pancreatic tissue that extends in an anterolateral or posterolateral direction to the descending duodenum (crocodile jaw appearance) (Fig. 1), with the risk of gastric outlet or duodenal obstruction similar to that of complete annular pancreas.

Ectopic pancreas

Ectopic pancreas is pancreatic tissue lying outside its normal location with no anatomic or vascular connection to the normal pancreas. Ectopic pancreas occurs in 0.3–13.7 % of the population [5]. The ectopic pancreas usually measures 0.5–2 cm in its largest dimension (rarely up to 5 cm) and is commonly located in the submucosa of the gastric antrum, or in the proximal portions of the duodenum and jejunum, and can simulate tumors such as gastrointestinal stromal tumor (GIST) and leiomyoma. Ectopic pancreas in the gastrointestinal tract is usually asymptomatic. However, it can be involved with the same inflammatory or neoplastic disease as the normal pancreas. Surgery is not necessary unless complications occur, so diagnosis is important to avoid unnecessary surgical treatment.

With CT, there are no specific imaging findings to differentiate ectopic pancreas from other submucosal masses. It has a smooth broad-based submucosal lesion in the greater curvature of the gastric antrum or in the proximal duodenum (Fig. 2). A key feature is a central umbilication representing

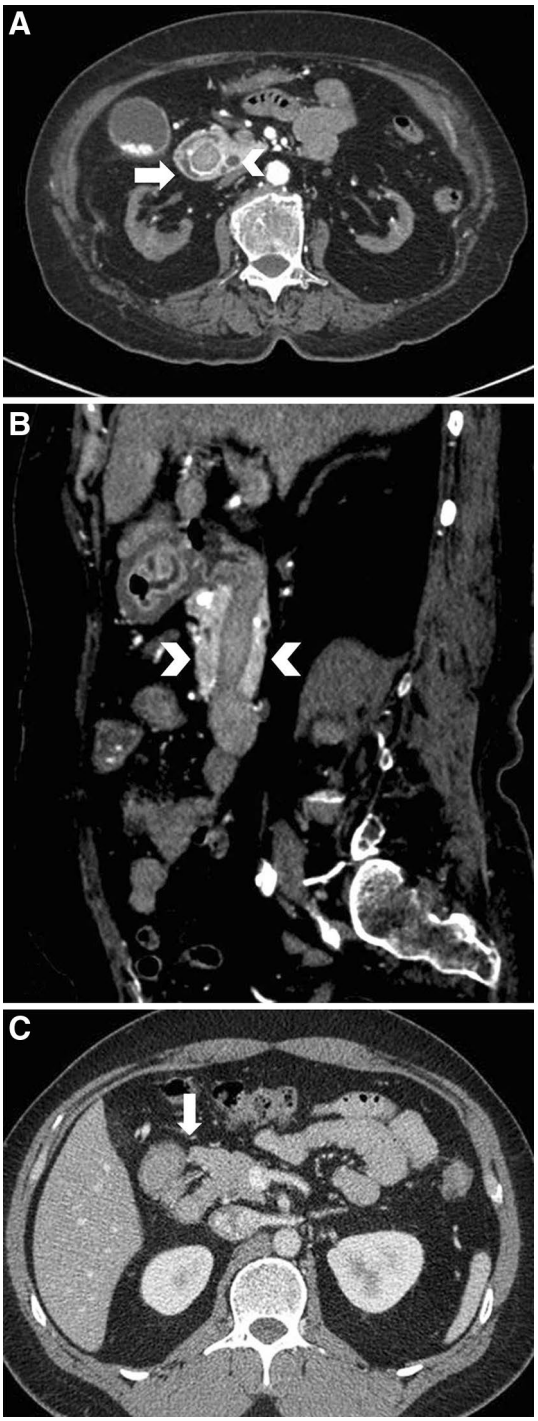

Fig. 1 Annular pancreas and incomplete annular pancreas. **a**, **b** A 35-year-old man with nausea and vomiting. Axial CT image, **a**, shows circular rim of pancreatic tissue with annular duct (*arrow*). *Arrowhead* indicates CBD. Sagittal CT image, **b**, shows pancreatic tissue around the second part of the duodenum (*arrowheads*). Findings are consistent with complete annular pancreas. **c** Axial CT image in 24-year-old man with abdominal discomfort shows pancreatic tissue semi-circumferentially extending around the second part of the duodenum, giving a crocodile jaw appearance (*arrow*), consistent with incomplete annular pancreas

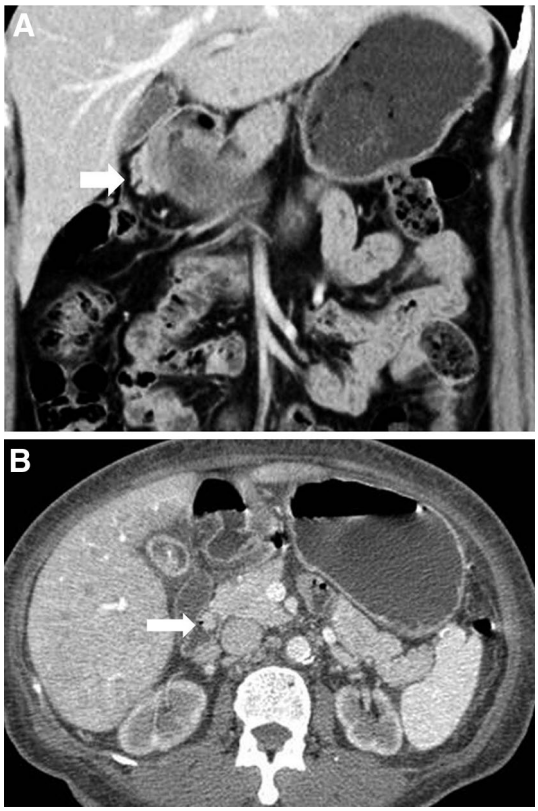

Fig. 2 Ectopic pancreas in two different patients. **a** A 24-year-old woman with abdominal pain. Coronal CT image shows lobulated broad-based submucosal lesion in the gastric antrum (*arrow*) resembling normal pancreatic tissue. Surgical specimen confirmed ectopic pancreatic tissue. **b** CT image of a 37-year-old man with colon cancer shows a small enhanced nodule with small central umbilication at the second part of the duodenum. *Arrow* indicates ectopic pancreatic tissue

the orifice of the rudimentary pancreatic duct containing a small collection of barium [6]. Ectopic pancreas may be mistaken as another submucosal mass such as GIST or leiomyoma on imaging. But GIST tends to grow with an exophytic or mixed pattern, globular contours and a better-defined border, while ectopic pancreas frequently grows with an endoluminal pattern, with flat contours and an ill-defined border.

Pancreatic hypoplasia

Hypoplasia or partial agenesis of the pancreas, also known as congenital short pancreas, results from absence of the ventral or dorsal anlage. Partial pancreatic agenesis more frequently involves the dorsal pancreas. Agenesis of the dorsal pancreas has two forms: complete agenesis and partial agenesis. Partial agenesis is more common than complete agenesis. In complete dorsal agenesis, the dorsal ductal system is missing. In partial dorsal agenesis, the dorsal ductal system

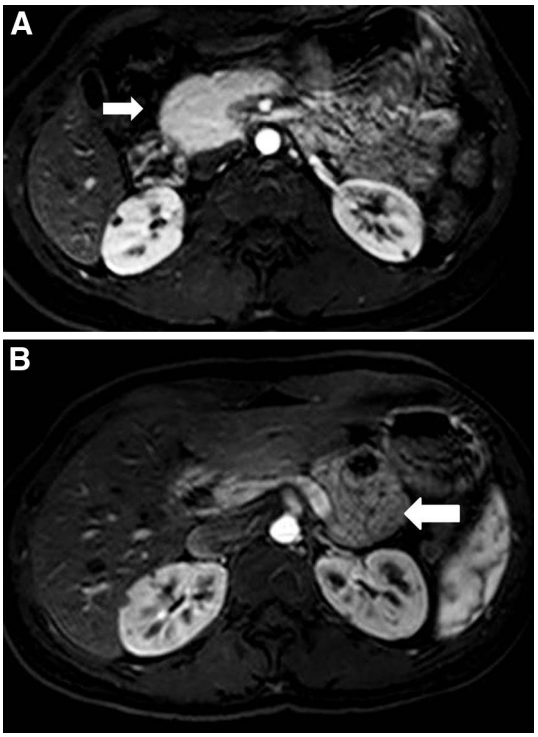

Fig. 3 Pancreatic hypoplasia. **a** Axial gadolinium-enhanced MR image shows truncated dorsal pancreas or partial agenesis of the pancreas with pseudotumoral enlargement of pancreatic head and uncinate (*arrow*). **b** Axial gadolinium-enhanced MR image at a different level shows absence of pancreatic neck, body and tail ventral to splenic vein with small bowel extending into this potential area (dependent intestine sign) (*arrow*)

is still identified. Dorsal agenesis may be an isolated finding, but has also been reported as part of heterotaxy syndrome [7]. Generally, patients remain asymptomatic but some of them present with nonspecific abdominal pain, pancreatitis and diabetes mellitus, as the bulk of beta cells are normally located in the pancreatic body and tail. Imaging shows a short pancreas with absent pancreatic neck, body and tail ventral to the splenic vein, with associated pseudotumoral enlargement of the pancreatic head and uncinate (Fig. 3). When agenesis of the dorsal pancreas is suspected, it is important to differentiate this condition from pancreatic cancer with upstream atrophy of the gland and pancreatic lipomatosis. Dependent stomach or dependent intestine signs are helpful imaging findings that allow differentiation of dorsal agenesis from lipomatosis.

Congenital pancreatic ductal variants

Pancreas divisum (PD)

PD is the most common congenital anomaly of the pancreas and results from failed fusion of the dorsal pancreatic

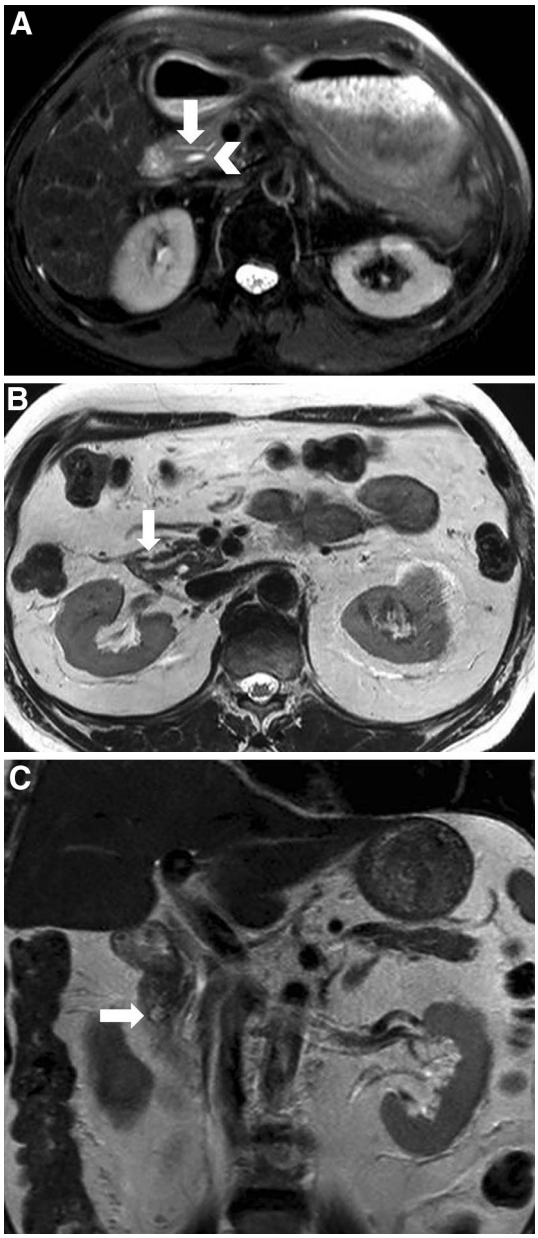

Fig. 4 Pancreas divisum in two different patients. **a** Axial T2-weighted MR image in 43-year-old woman shows main pancreatic duct (*arrow*) passing anterior to common bile duct (*arrowhead*). **b**, **c** Axial and coronal MR images show separate draining of dorsal and ventral duct consistent with pancreatic divisum. There is a small focal dilatation of the dorsal duct near the minor papilla, namely Santorinicele (*arrow*)

duct (duct of Santorini) and ventral pancreatic duct (duct of Wirsung) during embryological development. There are three types or variants of PD. Type I or the complete type, has a total failure of fusion: the dorsal duct drains into the minor papilla; the ventral duct, which is typically short and narrow, drains through the major papilla. The dorsal duct typically shows a larger caliber because of its

dominant function in pancreatic ductal drainage (dominant dorsal duct sign) (Fig. 4). Type II has dorsal duct dominant drainage in the absence of the duct of Wirsung, and type III (incomplete type) has a small communicating branch between the dorsal duct and ventral duct.

The relationship between pancreas divisum and pancreatitis is controversial [8]. The cause of pancreatitis is thought to be incomplete drainage through the minor papilla. Focal dilatation of the distal part of the dorsal pancreatic duct, resulting from relative obstruction at the minor papilla, also known as Santorinicele (Fig. 4), is associated with pancreas divisum.

CT may demonstrate PD but only when the pancreatic duct is visualized. MRCP helps in identifying PD and Santorinicele [9]. Key to diagnosis is the presence of the dorsal duct passing anterior and superior to the CBD on an axial image [10].

Anomalous pancreaticobiliary junction (APBJ)

The junction of the CBD and pancreatic duct can form in a number of ways. Most commonly, in about 80–90 % of cases, the CBD and pancreatic duct join with a short common channel of about 4–5 mm. The prevalence of APBJ varies from 1.5 % to 3.2 % [11]. APBJ refers to the pancreatic duct and CBD uniting outside the ductal wall and forming a long common channel (usually more than 15 mm) (Fig. 5). ERCP allows accurate diagnosis of APBJ, but ERCP is invasive [12]. MRCP has an advantage over ERCP in that it is a noninvasive and accurate imaging method. MRCP can depict the pancreaticobiliary ductal lumen and APBJ, with simultaneous detection of the wall of the bile duct, pancreatic parenchyma and extrapancreatic findings. Because of the long common channel of the CBD and main pancreatic duct, the normal muscle complex of the sphincter of Oddi does not prevent reflux between the pancreatic duct and bile duct, possibly resulting in a choledochal cyst. Conversely, reflux of bile into the pancreatic duct can cause chronic pancreatitis. APBJ is also considered to be a major risk factor for biliary tract malignancies such as cholangiocarcinoma and gallbladder carcinoma [13].

Duplication of pancreatic ductal system

The course of the pancreatic duct varies greatly, and the most common pattern, in approximately half of cases, is a descending course. Other patterns include vertical, loop and sigmoid course [14]. Duplication of the pancreatic duct is not uncommonly seen, especially in the tail (Fig. 6), whereas parenchymal duplication is extremely uncommon.

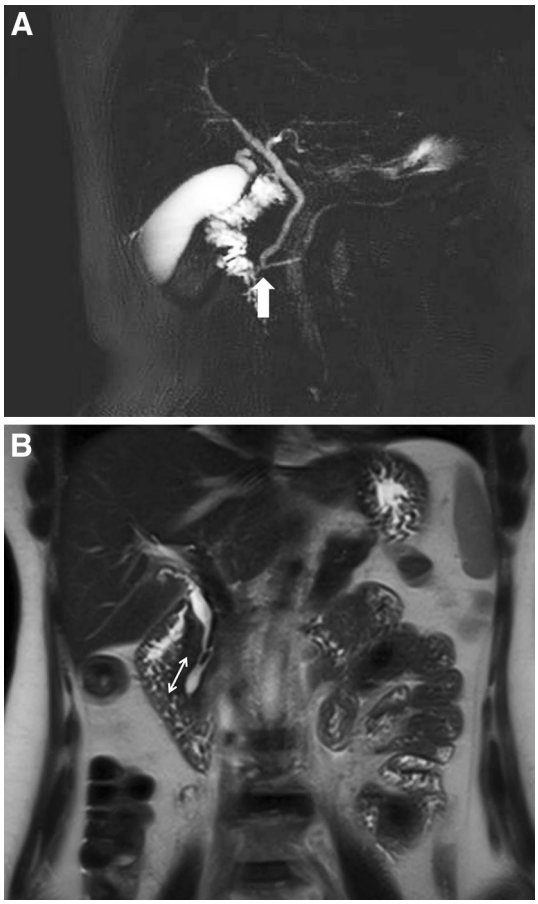

Fig. 5 Normal and anomalous pancreaticobiliary junction. **a** Coronal MRCP in 60-year-old woman with bloating shows normal pancreaticobiliary junction (*arrow*). **b** Heavily T2-weighted MR image in 45-year-old woman with previous pancreatitis shows long common channel of pancreaticobiliary junction approximately 18 mm (*double-headed arrow*), consistent with anomalous pancreaticobiliary junction

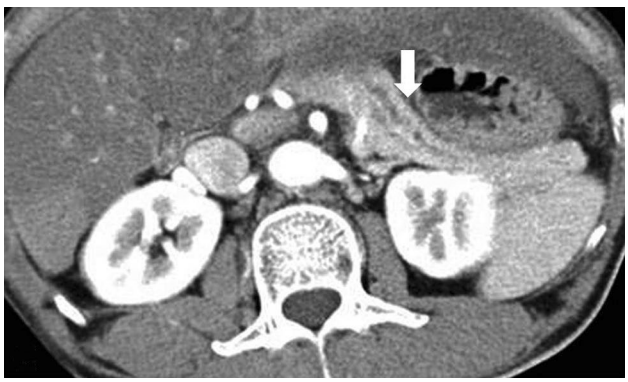

Fig. 6 Axial CT image in 33-year-old man with abdominal discomfort shows duplication of main pancreatic duct at pancreatic body and tail (*arrow*)

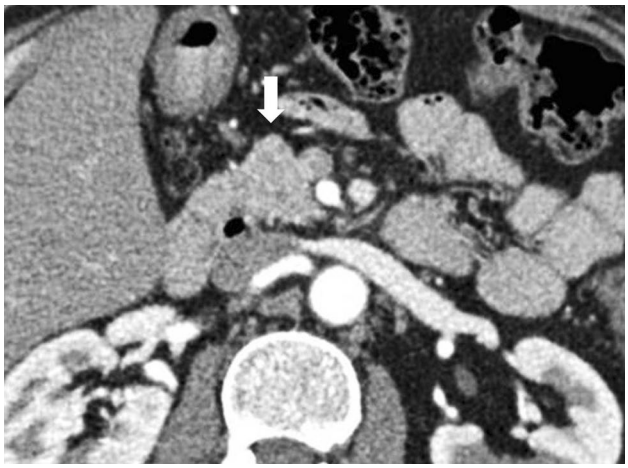

Fig. 7 Axial CT image in 56-year-old man with recurrent abdominal pain and rising CA 19-9 shows well-defined bulging area at anterior surface of pancreas at head-neck junction (*arrow*), which abuts posterior aspect of the lesser omentum consistent with tuber omentale

Pancreatic contour variants

Tuber omentale

Tuber omentale is a type of pancreatic shape variation. It is a well-demarcated prominence of the pancreatic tissue on the anterior surface of the pancreas, usually at the head-neck junction that abuts against the posterior surface of the lesser omentum (Fig. 7). Its right limit is marked by a groove for the gastroduodenal artery. It can simulate a pancreatic tumor or lymph node on imaging, but it can be distinguished from a tumor in that its attenuation, signal intensity and pattern of enhancement is identical to those of the rest of the pancreatic parenchyma.

Pancreatic cleft

The normal pancreas has lobulated contours; however, peripancreatic fat can invaginate between lobulations and produce an appearance that is similar to a cleft (Fig. 8). On imaging, a pancreatic cleft can cause most diagnostic confusion in a trauma patient, as it can simulate pancreatic fracture (Fig. 9). However, normal levels of pancreatic enzyme and lack of peripancreatic fat stranding or abnormality exclude pancreatic injury.

Congenital pancreatic cysts

Congenital pancreatic cysts are very rare and distinguished from post inflammatory pancreatic pseudocysts through the presence of cuboidal or stratified squamous epithelial lining

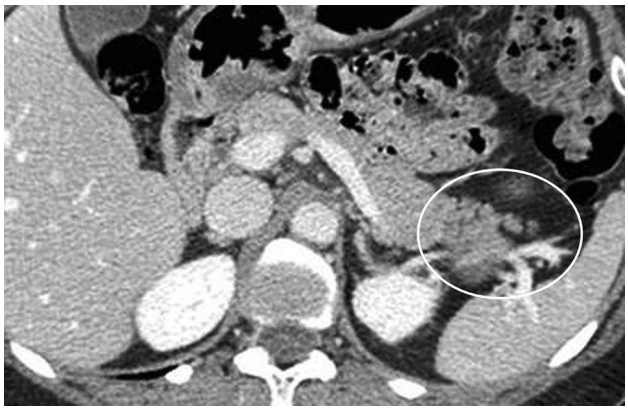

Fig. 8 Axial CT image in 41-year-old man shows small pancreatic cleft which is the invagination of fat into the pancreas (*circle*)

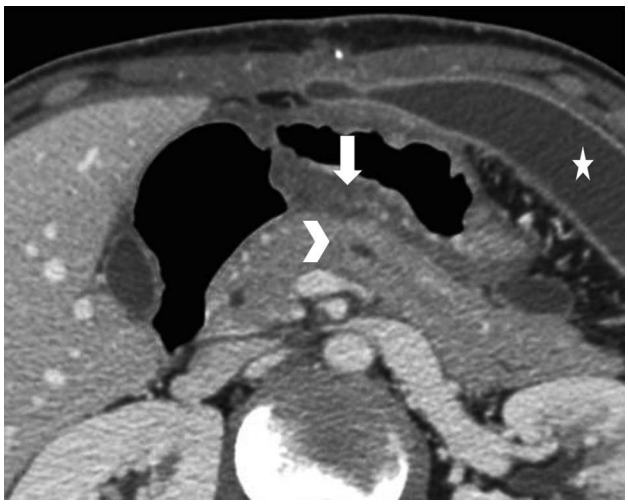

Fig. 9 Axial CT image in 19-year-old man shows linear low density of pancreatic fracture through both surfaces of pancreatic body (*arrowhead*). The peripancreatic fat is misty (*arrow*), with evidence of intra-abdominal collection (*star*)

[15]. Congenital pancreatic cysts are usually multiple and almost all are associated with congenital diseases such as Von Hipple Lindau syndrome (VHL) and autosomal dominant polycystic kidney disease (ADPKD) (Fig. 10). VHL is an autosomal dominant disorder characterized by CNS hemangioblastomas, renal cell carcinomas, retinal angiomatosis, pheochromocytomas and cystic lesions in the kidneys, pancreas and epididymis. Pancreatic cysts are relatively common in about 70 % of patients with VHL. The pancreatic cysts are variable in size, but typically small and multiple, and involvement can range from a solitary cyst to cystic replacement of the gland. Congenital pancreatic cysts must be differentiated from nonneoplastic or neoplastic cystic lesions. On imaging, a unilocular macrocystic lesion is

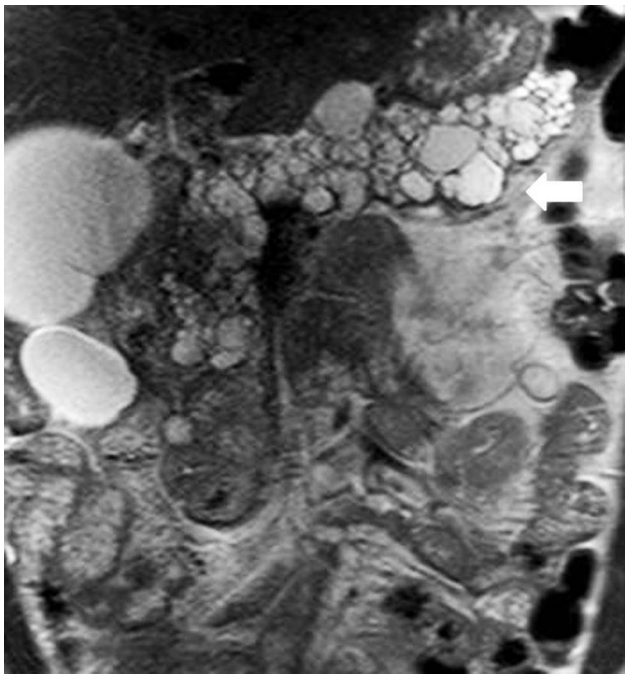

Fig. 10 Coronal T2-weighted MR image through pancreas shows multiple cysts with high signal intensity replacing pancreatic parenchyma (*arrow*). This patient has right adrenal pheochromocytoma, right renal cell carcinoma and CNS hemangioblastoma (not shown). All features are consistent with Von Hippel Lindau syndrome

sufficient to make a diagnosis of pancreatic pseudocyst in a patient with a history of pancreatitis. The presence of internal septations or microcystic lesions is suggestive of a pancreatic cystic tumor, particularly an intraductal papillary mucinous neoplasm, when there is communication between the main pancreatic duct and the cyst. Clustering of multiple cysts makes it difficult to differentiate from serous cystadenoma [16]. However, numerous small cysts that are conjoined in a honeycomb-like formation, a central scar which may be calcified, and no visible communication between the cysts and the pancreatic duct are indicative of serous cystadenoma.

Mimics of pancreatic lesions

Intrapancreatic accessory spleen (IPAS)

IPAS results from failure of fusion of splenic buds in the dorsal mesogastrium during embryonic development. The most common location of accessory spleen is at the splenic hilum, however the splenic rest cells can occur in or around the pancreatic tail to form IPAS. On CT or MRI, it is seen as a well-defined rounded lesion ranging from 4 to 32 mm [17]. The density, signal intensity and dynamic enhancement pattern of IPAS follow the main spleen (Fig. 11). IPAS is often asymptomatic; however, it can be misdiagnosed

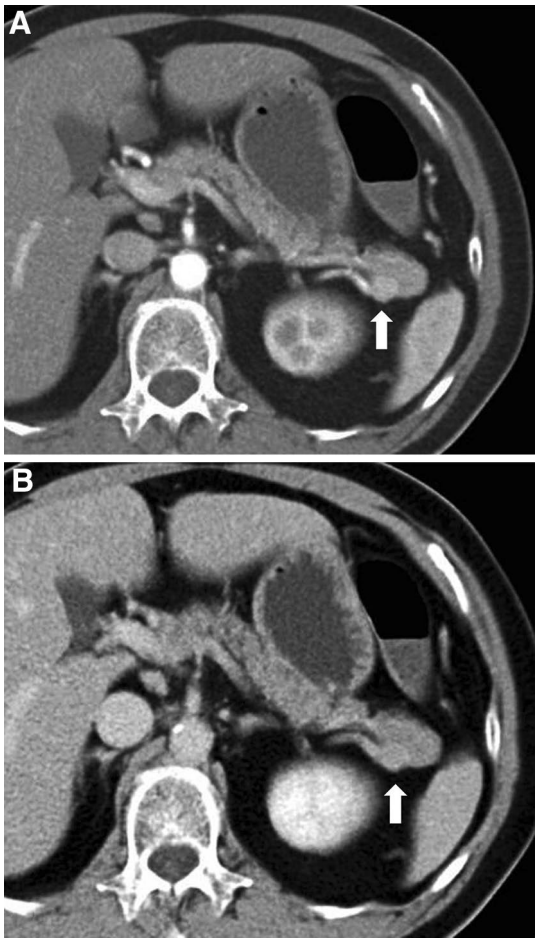

Fig. 11 A 40-year-old man with abdominal pain and vomiting. Axial CT images on arterial (**a**) and portal venous phase (**b**), show well-circumscribed rounded nodule at pancreatic tail (*arrow*), with similar enhancement to the native or anatomical spleen

as a pancreatic neoplasm. The arciform enhancing pattern of spleen during the arterial phase due to flow differences between red and white pulp is also detected in IPAS, which is a helpful imaging criterion for differentiating IPAS from hypervascular neoplasms such as hypervascular metastases or a neuroendocrine tumor. Neuroendocrine tumors have a more homogeneous enhancement during the arterial phase, with relative washout on portal venous phase. In equivocal cases, a definitive diagnosis can be obtained by means of single-photon emission computed tomography (SPECT) with technetium 99m heat-damaged RBCs as the tracer, which are trapped by splenic tissue, and seen as an area of increased uptake at the site of suspected accessory spleen. Superparamagnetic iron oxide enhanced MRI (SPIO enhanced MRI) practically uses the same principle as that of Tc-99m heat-damaged RBCs, as the SPIO-based agent is targeted to the cells of the reticuloendothelial system (RES). Therefore, it can be used to show the function

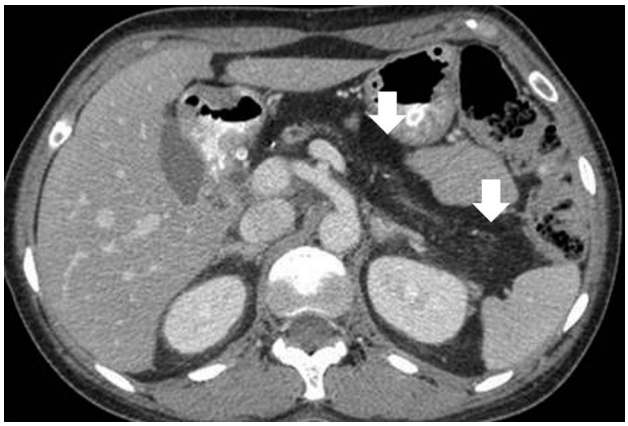

Fig. 12 Axial CT image in 31-year-old man with cystic fibrosis shows diffuse hypoattenuating fat replacing entire pancreas (*arrows*). The fat replacement of the pancreas anterior to the splenic vein prevents the adjacent abdominal organs abutting the splenic vein

of RES within the IPAS as well as that of the main spleen on T2- or T2*-weighted images [18].

Fatty change and replacement of pancreas

Fatty change of the pancreas on imaging is frequently variable. A focal form of fatty change frequently involves the anterior pancreatic head. Uneven fatty change can cause confusion with a pancreatic mass; however, measurement of negative Hounsfield units with CT and detection of intravoxel fat with chemical shift MRI can help confirm the diagnosis. The diffuse form (fatty replacement or lipomatosis of the pancreas) has been associated with diseases such as Shwachman–Diamond syndrome, diabetes mellitus, and cystic fibrosis (Fig. 12). Helpful findings include preservation of normal pancreatic contours, no CBD or pancreatic duct dilatation, and lack of displacement or invasion of adjacent vasculatures.

Conclusion

Key imaging features of congenital pancreatic lesions in this article are helpful for establishing a correct diagnosis and avoiding misdiagnosis.

Conflict of Interest All authors declare that they have no conflict of interest.

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