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Case Report

Portal annular pancreas: the pancreatic duct ring sign on MRCP

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ARTICLE INFO

Article history:

Received 14 July 2015

Accepted 23 August 2015

Available online 21 October 2015

Keywords:

Portal annular pancreas

Pancreatic duct anomalies

MRCP

ABSTRACT

Portal annular pancreas is a rare pancreatic variant in which the uncinate process of the pancreas extends and fuses to the dorsal surface of the body of the pancreas by surrounding the portal vein. It is asymptomatic, but it can be mistaken for a pancreatic head mass on imaging and could also have serious consequences during pancreatic surgery, if unrecognized. We report this case of a 53-year-old female patient who was diagnosed to have portal annular pancreas on the basis of an unusual course (ring appearance) of the main pancreatic duct on magnetic resonance cholangiopancreatography, not described earlier in the radiology literature.

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Introduction

Because of its complex embryologic development, the pancreas has a broad spectrum of anatomic variants like pancreas divisum, pancreas annulare, or portal annular pancreas (PAP) [1,2]. PAP is the rarest of these anomalies and is usually asymptomatic [1]. In PAP, the uncinate process wraps around the portal vein and fuses with the body of pancreas posteriorly [3]. This anomaly of pancreatic parenchymal development is usually not associated with pancreatic ductal abnormality except for pancreas divisum. We report a unique case where PAP is associated with an unusual ringlike configuration of the main pancreatic duct encircling the portal

vein. The case highlights an important sign of PAP, which we call the pancreatic-duct ring sign.

Case report

A 53-year-old woman presented to her primary physician with 1 week of painless jaundice associated with 25 lbs weight loss and diarrhea over the last year. There was no history of alcohol use or any previous surgeries. Physical examination revealed a mildly cachectic woman with icterus and without any hepatosplenomegaly. Ultrasound evaluation revealed intrahepatic ductal dilatation and a heterogenous appearing

Case originated at University of Arkansas Medical Sciences, 4301 West Markham Street, Little Rock, AR 72205.

Disclosures: The authors have nothing to disclose.

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<http://dx.doi.org/10.1016/j.radcr.2015.08.005>

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liver. Computed tomography of the abdomen showed a low attenuation lesion measuring approximately 4.1×2.2 cm at the confluence of the right and left hepatic ducts with diffuse intrahepatic ductal dilatation suspicious for a Klatskin tumor (Fig. 1). No pancreatic mass was identified. Endoscopic retrograde cholangiopancreatography demonstrated a severe stricture at the confluence of the hepatic ducts. The patient underwent a percutaneous internal and/or external biliary drain and brush biopsy, which demonstrated an adenocarcinoma. Subsequent imaging (magnetic resonance imaging and magnetic resonance cholangiopancreatography [MRCP]) for staging of tumor and pretreatment evaluation for liver transplant confirmed abrupt narrowing of both hepatic ducts at the confluence corresponding to a cholangiocarcinoma. On review of the MRCP images, attention was drawn to an unusual ringlike configuration of the pancreatic duct (Fig. 2). On further examination, the portal vein flow void was seen through this segment of the pancreatic duct (Fig. 3). These findings prompted a careful review of the pancreatic anatomy on magnetic resonance images that demonstrated pancreatic head parenchyma that was seen circumferentially surrounding the portal vein and demonstrated fusion with the rest of the body of pancreas representing a PAP (Fig. 4).

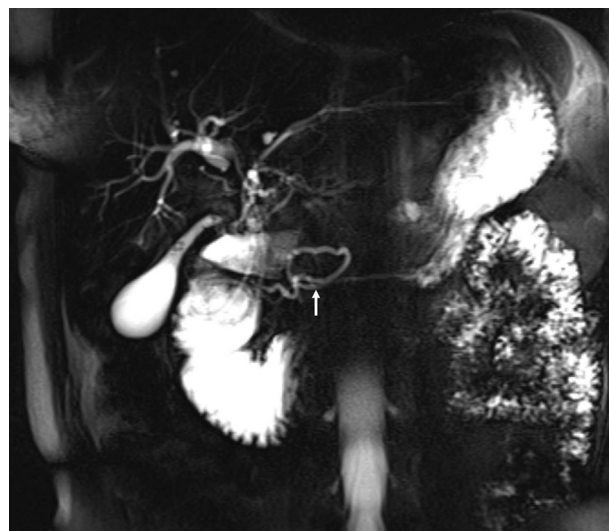


Fig. 2 – Single coronal image from the MRCP sequence shows an unusual ringlike configuration of the main pancreatic duct (arrow). Also note the intrahepatic biliary ductal dilatation due to a central obstructing lesion shown on the computed tomography.

Discussion

PAP is a pancreatic malformation in which the pancreatic tissue, usually the uncinate process, wraps annularly around the portal vein and fuses posteriorly to the body of pancreas. This entity is 7 times more common in females [3]. There are only a few reported cases in literature, with the first case reported in 1987 [3,4]. This entity is commonly seen in the swine pancreas. It has a prevalence of 1.14% in humans, observed by Karasaki et al [3].

The mechanism of development of PAP is uncertain; however, it is hypothesized that during fetal development, the ventral pancreas rotates initially to the right and then to the left about the axis of the primitive gut to fuse posteriorly with



Fig. 1 – Axial contrast-enhanced computed tomography through the liver shows intrahepatic biliary ductal dilatation (arrows) due to an infiltrative mass in the liver (M) that was confirmed to be a cholangiocarcinoma.

the dorsal pancreas. In rare instances, this fusion occurs to the left of the mesenteric and/or portal vein, thus encircling the portal vein [1,5]. This condition is asymptomatic and is usually an incidental finding while imaging the abdomen for some other indication [6].

A classification proposed by Joseph et al describes 3 subtypes of PAP. PAP may be classified as type I when ventral bud of pancreas fuses with body and ductal system of pancreas posterior to the portal vein, type II when type I is associated with pancreas divisum, and type III when the uncinate process is involved in the process in the encasement of the vessels and fusion. Each may be further divided in to a, b, and c (suprasplenic, infrasplenic, and mixed) depending on its relationship to the splenic vein [1].

Another classification describes this entity as suprasplenic (commonest), infrasplenic, and mixed type based on the fusion of uncinate process with the body posteriorly above or below the level of the splenoportal confluence. It may be associated with the abnormal course of pancreatic duct (retroportal pancreatic duct) or pancreatic divisum [1,6]. The mean length of fusion is approximately 9.4 mm [3].

Our case shares some similarities with the suprasplenic subtype of PAP but is unique in that the pancreatic duct forms a near complete ring around the portal vein. This anatomic variation of the pancreatic duct forming a ring around the portal vein in association with a PAP has never been reported in the literature and does not fit with any of the classifications described in the literature.

A computed tomography scan is usually adequate for diagnosing this anomaly, which can be demonstrated by continuity of the extension of the uncinate process into the body of the pancreas in more than 2 slices [1]; however, the presence of this anomaly can also be suggested by demonstrating the abnormal course of the pancreatic duct when present on an

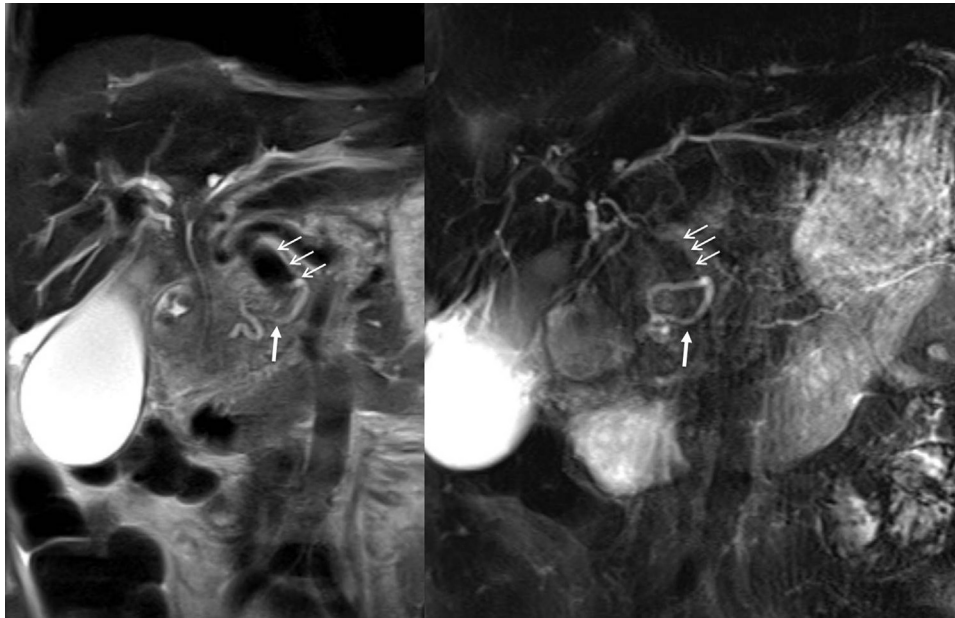


Fig. 3 – Coronal half Fourier acquisition single shot turbo spin echo (left) and MRCP (right) images show a ringlike configuration of the pancreatic duct in the head of the pancreas (thick arrows). Notice the portal vein flow void coursing through the loop of the pancreatic duct consistent with portal annular pancreas (fine arrows).

MRCP (as in our case). We propose the pancreatic-duct ring sign as a diagnostic sign for PAP when seen on MRCP or endoscopic retrograde cholangiopancreatography.

Clinical issues related to PAP emerge mainly when operative intervention is required. During pancreatic surgery in patients with PAP, the pancreas needs to be dissected to

separate the lingual projection from the dorsal surface of the body of the pancreas [3]. The knowledge of extent of the abnormal tissue and the course of the duct is important for the surgeon, as inadvertent excision of the tissue or the duct could potentially lead to fistula formation [6]. From an imager's perspective, familiarity with this entity is important,

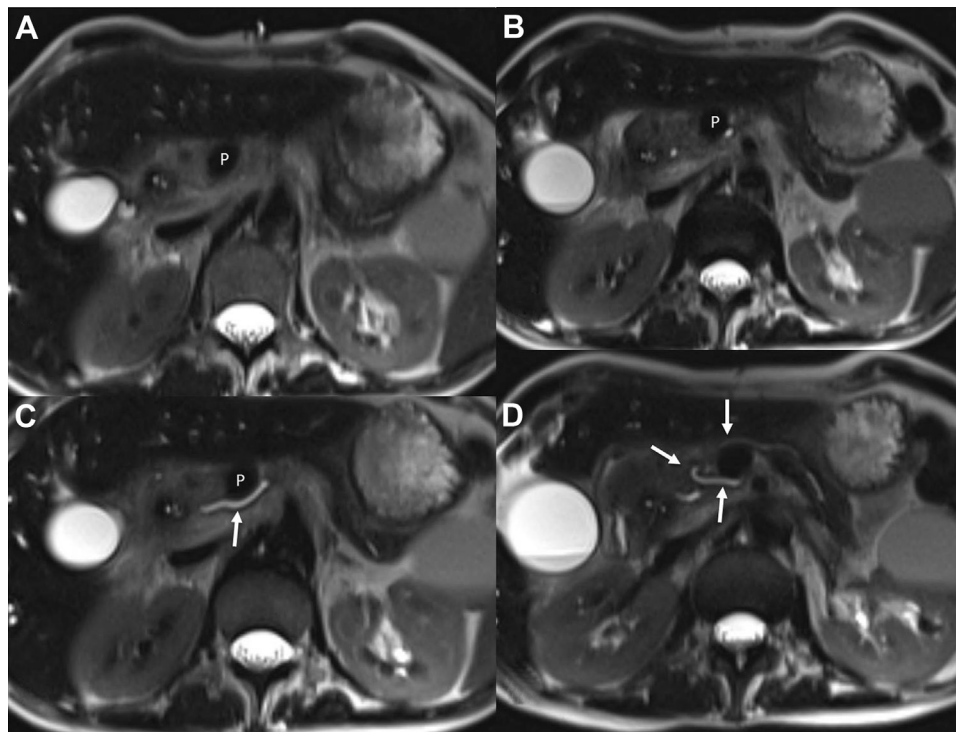


Fig. 4 – Sequential axial half Fourier acquisition single shot turbo spin echo images through the course of the portal vein (P) starting at the suprapancreatic level (A) and through the pancreatic neck (B, C) and head (D) show the circumferential pancreatic parenchyma around the portal vein with looping of the pancreatic duct (arrows).

not only to alert the surgeon to its presence but also to prevent confusion with a pancreatic mass [7].

Conclusion

PAP is a rare anomaly in humans. It can potentially be mistaken for a pancreatic head mass and may have important implications for pancreatic surgery in affected patients. We emphasize that the ring appearance of the pancreatic duct around the portal vein should prompt the diagnosis of this rare, asymptomatic, surgically important developmental variant of the pancreas.

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