CASE REPORT

Possible congenital dilatation of the pancreatic duct

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Accepted 19 September 2017

SUMMARY

The main pancreatic duct can become dilated in a number of conditions. We describe a patient with gross dilatation of the main pancreatic duct without evidence of causative underlying pathology suggesting congenital dilatation of the pancreatic duct. A 36-year-old man presented with signs of intestinal obstruction and a history of surgery for congenital pyloric stenosis. Incidental findings on CT showed a massively dilated main pancreatic duct. On MRI there was no duct irregularity or solid mural nodule, making a main duct intraductal papillary mucinous neoplasm unlikely. Endoscopic ultrasound findings were in keeping with those on MRI. Fine needle aspiration revealed a non-viscous fluid with a low carcinoembryonic antigen and high amylase concentration, consistent with normal pancreatic fluid levels rather than a mucinous collection. After 1 year, the cyst remains unchanged. This patient will be kept under surveillance with yearly MRI.

BACKGROUND

The pancreatic duct system consists of the main duct and side branches. The main pancreatic duct (MPD) diameter is greatest at the head and neck region (up to 5 mm in older patients) and is slightly narrower towards the body and tail (1–2 mm).^{1,2}

The MPD can become dilated in chronic pancreatitis or pancreatic malignancy. Pancreatic duct dilatation can also occur in the presence of intraductal papillary mucinous neoplasm (IPMN), a precursor to invasive pancreatic malignancy characterised by intraductal tumour growth and mucus secretion.

Here, we present a case of grossly dilated MPD without evidence of causative underlying pathology suggesting congenital dilatation of the pancreatic duct. As far as we know, there has been no documentation in the literature of a patient with congenital, massive enlargement of the pancreatic duct to this extent.

CASE PRESENTATION

A 36-year-old man of Pakistani-Asian descent presented with bilious vomiting, constipation and abdominal distension. Relevant surgical history included laparotomy as a neonate. Although records were not available for this episode, the patient gave a history that was in keeping with surgery for congenital pyloric stenosis.

He had no other comorbidities, was an active smoker and did not consume alcohol. There was no family history of note. He had no symptoms suggesting chronic pancreatitis.

A CT scan of the abdomen demonstrated small bowel obstruction likely secondary to adhesions. A laparotomy and adhesiolysis was performed and the patient made an uneventful recovery.

As an incidental finding, the CT scan demonstrated a massively dilated MPD of approximately 50 mm, with no parenchymal calcification or solid mass (figure 1). Despite atypical imaging, an initial working diagnosis of a main duct-intraductal papillary mucinous neoplasm (MD-IPMN) was made.

INVESTIGATIONS

A subsequent MRI confirmed dilatation of the MPD measuring approximately 50 mm in the head, neck and proximal body of the pancreas (figures 2 and 3). There was no mass at the transition point. The duct itself had a smooth contour in the dilated areas with no side branch dilatation or mural nodules. There was evidence of chronic pancreatitis in the pancreatic tail, with atrophy, reduced T1 signal of the parenchyma indicating fibrosis and side branch dilatation (figures 2 and 3).

Endoscopic ultrasound (EUS) was performed to assess the morphology and potential mural nodules and thickening.³ The EUS findings were in keeping with those on MRI, with no mural thickening or nodules in the very dilated segments of pancreatic duct. Fine needle aspiration of the cyst revealed clear non-viscous fluid with a carcinoembryonic antigen (CEA) concentration of 0.8 ng/mL and amylase over 300 000 IU. Cytology was benign.

The imaging appearances have remained stable over a 12-month period.

DIFFERENTIAL DIAGNOSIS, OUTCOME AND FOLLOW-UP

This patient will be kept under surveillance with yearly MRI pancreas and magnetic resonance cholangiopancreatography (MRCP) since the natural history of such lesions is unknown, and there remains some diagnostic doubt concerning the aetiology of his duct dilatation. After 12 months, the patient's lesion remains unchanged.

DISCUSSION

The causes of MPD dilatation can be categorised into those due to an obstructing mass or stricture and/or those due to a change in the volume or viscosity of pancreatic secretions. MPD dilatation most commonly occurs in the setting of chronic pancreatitis or pancreatic, biliary or ampullary tumour. A combination of CT and MRI plus occasionally EUS with aspiration is usually able to

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To cite: Tavare A,
Gordon-Weeks AN,
Bungay H, et al. *BMJ Case
Rep* Published Online First:
[please include Day Month
Year]. doi:10.1136/bcr-2017-
221191

Figure 1 Axial, portal-phase, contrast-enhanced CT through the pancreas demonstrating marked smooth cystic dilation of the head and neck region of the pancreas with associated duct dilation into the body (arrowheads), without mural nodules or side branch dilatation, and changes of chronic pancreatitis in the tail (arrow), with atrophy and side branch dilatation but no calcification.

differentiate between the main causes of pancreatic duct dilatation (table 1).

In MD-IPMN, the tumour nodule(s) may be very small and not seen on CT or MRI. They produce large amounts of viscid mucin. A combination of overproduction and increased viscosity causes duct dilatation. If a solid nodule enlarges, and especially undergoes malignant transformation, then an element of mass obstruction can also occur. IPMNs are premalignant neoplasms composed of mucin-producing columnar cells. Characteristically, the lesions show papillary proliferation, varying degrees of cellular atypia and cyst formation. IPMNs may involve the MPD, the branch ducts or both. Patients with IPMNs involving the main duct are at high risk of developing malignancy (approximately 70% at 10 years), compared with those identified within branch ducts (approximately 20%).⁴ When the main duct is involved at the ampullary level, there is a typical 'fish mouth' appearance on endoscopy. Fluid aspirated from an IPMN is typically viscous and contains mucin. A positive 'string test' and a high CEA content on biochemical analysis also make a diagnosis of IPMN more likely.^{4 5} Importantly, we identified none of these classical findings, effectively excluding the possibility that this patient has an underlying IPMN as a cause for the duct dilatation.

Figure 3 Axial fat-saturated T1 postintravenous gadolinium in the portal phase demonstrating no evidence of mural nodules or solid tumour in the wall (arrow) of the markedly dilated main pancreatic duct nor at the transition to more normal calibre duct.

The patient does have changes of chronic pancreatitis in the tail of the gland. It is conceivable that the massive dilations of the duct in head, neck and body are also features of chronic pancreatitis, but the absence of clinical features, absence of parenchymal calcification and smooth contour of the very dilated segments without side branch dilatation here make this less likely. We postulate that the chronic pancreatitis changes in the tail are secondary to the primary abnormality of massive duct dilatation in the head, neck and body.

It is therefore hypothesised that this incidental finding may be a congenital pancreatic duct dilation. There have been reports of congenital dilations elsewhere in the biliary tract. Choledochal cyst, for example, is a relatively rare, but well-documented congenital biliary tree dilatation. Most choledochal cysts are diagnosed in childhood; however, up to 20% manifest in adulthood.⁶ Choledochal cysts can affect any part of the extrahepatic or intrahepatic biliary tree and they carry the risk of malignant transformation.^{7 8} It has also been shown that moderately dilated pancreatic ducts and pancreatic cysts are associated with an increased risk of malignancy.^{9 10} However, it is not known whether chronically and massively dilated pancreatic ducts carry malignant risk.

The patient had a history of a probable congenital pyloric stenosis as a neonate. Although there is no clear association

Figure 2 Unenhanced axial fat-saturated T1 MRI demonstrating (A) marked, smooth dilatation of the main pancreatic duct in the head, neck and body (arrowheads) with atrophy and reduced T1 signal (arrow) of the parenchyma in the tail, and (B) preservation of the parenchyma in the head (arrows). (C) Thick heavily T2-weighted magnetic resonance cholangiopancreatography (MRCP) image showing dilated side branches in the tail (arrow) but marked, smooth dilatation in the neck and body (arrowheads).

Table 1 The main causes of pancreatic duct dilatation

Causative category	Examples	Imaging findings
Obstructive mass	PDAC	Infiltrative mass with upstream pancreatic duct dilatation ± parenchymal changes of obstructive pancreatopathy (atrophy and fibrosis)
	Main duct IPMN	Main duct dilatation Open ampulla (fish mouth) Mural nodule Mural thickening
	Other pancreatic tumours	Many types of pancreatic neoplasm may compress the duct.
Intrinsic stricture	Chronic pancreatitis	Calcification Atrophic pancreas Reduced parenchymal T1 MRI signal Pancreatic duct beading
	Autoimmune pancreatitis	Diffusely or focally enlarged pancreas Smoothly narrowed main pancreatic duct, often not visible along the whole length of gland
	Iatrogenic (post-ERCP or pancreatectomy)	Short, isolated duct stricture with proximal dilatation
Increased secretion viscosity	IPMN	See above. In MD-IPMN, the tumour nodule(s) may be very small and not seen on CT or MRI. They produce large amounts of viscid mucin. A combination of overproduction and increased viscosity causes duct dilatation. If a solid nodule enlarges, and especially undergoes malignant transformation, then an element of mass obstruction can also occur.

ERCP, endoscopic retrograde cholangio-pancreatography; IPMN, intraductal papillary mucinous neoplasm; MD-IPMN, main duct-intraductal papillary mucinous neoplasm; PDAC, pancreatic ductal adenocarcinoma.

between congenital pyloric stenosis and biliary tract dilatation, prior reports suggest common developmental malformations of the gastrointestinal tract or its blood supply.^{11–13} Therefore, it is plausible that this patient's pyloric stenosis and pancreatic duct dilatation have an underlying congenital association. There is a small possibility that this patient's massive pancreatic dilatation is associated with the surgery that he underwent as a neonate. However, the incidence of pyloric stenosis is common and the operation is performed frequently. Despite this, there is no documented evidence of associated pancreatic cysts either postoperatively or as an associated abnormality. Therefore, we concluded that the cyst is very unlikely to be due to surgical intervention, but we also cannot definitively exclude it.

As far as we know, there has been no documentation in the literature of a patient with congenital, massive enlargement of the pancreatic duct to this extent. We invite correspondence on

similar cases of massive pancreatic duct dilatation of unknown and possibly congenital aetiology with interest.

Learning points

- Incidental findings on imaging can be a source of clinical dilemma.
- Congenital massive enlargement of the pancreatic duct of this extent has not previously been described in the literature.
- Careful investigation must be performed in order to rule out the other sinister causes of pancreatic duct dilatation.
- This diagnosis prevented unnecessary invasive and life-changing surgery such as pancreatectomy; instead, the patient will be kept under surveillance with yearly MRI.

Contributors AT, ANG-W, HB and MS contributed equally to the writing and editing of the manuscript. MS was the consultant surgeon involved in the patient's care.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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