The "sausage pancreas" on magnetic resonance imaging

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This image illustration refers to the case of a 49-year-old female patient suffering from gradually increasing jaundice and recurrent dull aching upper abdominal pain for many months. The patient was also suffering from diabetes mellitus which was diagnosed during the course of this presentation. Physical examination revealed mild tender hepatomegaly and mild icterus. Rest of the physical examination was unremarkable. Initial evaluation with laboratory tests revealed hyperglycemia (fasting blood sugar up to 320 mg/dL), hyperbilirubinemia (total: 2.4 mg/dL, conjugated 1.3 mg/dL), high serum alkaline phosphatase (1302 U/L), elevated serum glutamic oxaloacetic transaminase/serum glutamate pyruvate transaminase (145 U/L, 122 U/L, respectively), and marginally elevated serum lipase (48.4 U/L).

Ultrasound revealed mild dilatation of bilobar intrahepatic biliary ducts, small calculi in the gall bladder (GB), and a mildly bulky pancreas with no peripancreatic collection. Magnetic resonance imaging (MRI) with magnetic resonance cholangiopancreatography (MRCP) was done for further evaluation. It revealed a diffusely bulky pancreas with effacement of surface lobulations causing smoothened featureless surface outline, mild T2 hyperintensity of pancreatic parenchyma, and diffusion restriction. There was also T2-hypointense smooth capsule-like rim along pancreatic margins (Figs. 1 and 2). There was no peripancreatic collection or any significant soft tissue stranding. The main pancreatic duct (MPD) was partially effaced and partially visualized with a maximum diameter of up to 2.2 mm in the proximal body region. There was no significant dilatation of MPD or side branches (Fig. 3a). MRCP also revealed multifocal narrowing and dilatation of extrahepatic as well as intrahepatic biliary ducts, along with tiny calculi in the GB and mild thickening of duct walls (Figs. 1 and 3). Mild hepatomegaly was noted. Rest of the liver was unremarkable. Considering these findings, the possibility of Type-1 autoimmune pancreatitis (AIP) with cholangiopathy was considered. Further, work-up revealed elevated serum immunoglobulin (Ig)G4 levels and IgG4 positivity in a biopsy sample taken from the pancreatic head using endoscopic ultrasound. The patient was managed on an outpatient/daycare basis.

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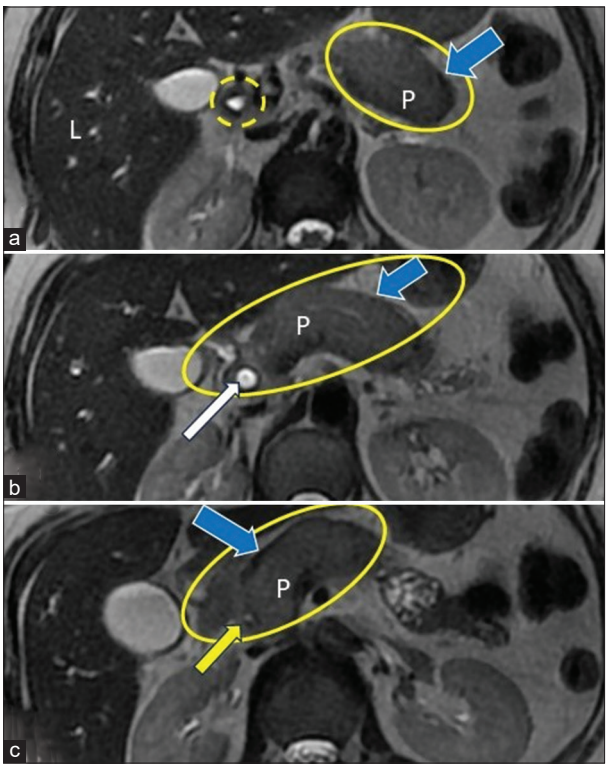

Figure 1: (a-c) T2-weighted axial images upper abdomen in craniocaudal direction showing mildly bulky pancreas (P, encircled) with diffuse hyperintense signal in parenchyma, loss of surface lobulations giving smoothened flat appearance to margins with T2-hypointense capsule-like rim (blue arrows) referred as "featureless" or "sausage pancreas". There is no significant peripancreatic fat stranding. Furthermore, noted areas of narrowing and dilatation in CBD (yellow and white arrows, respectively). Wall thickening in CBD (dotted circle). CBD: Common bile duct

AIP is an immune-mediated inflammatory disease of the pancreas. There are two types of this condition: Type-1 characterized by pancreatic as well as biliary involvement, elevation of serum IgG4 level, and deposition of IgG4 in affected tissues. It may be part of multisystem IgG4-related disease. In Type-2 AIP, there is isolated involvement of the pancreas. Pancreatic involvement in AIP may be diffuse, focal, or multifocal [1,2]. The diffuse form is classically characterized by "sausage-shaped" enlargement of the pancreas, an appearance resulting from diffuse enlargement of the pancreas with loss of

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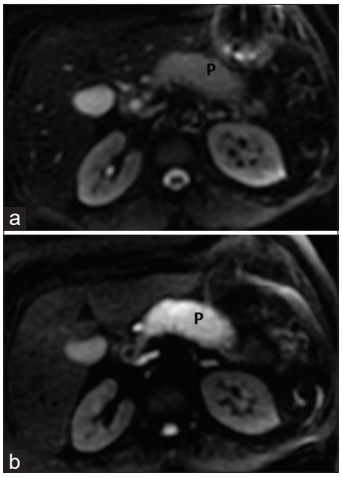

Figure 2: (a and b) Axial diffusion-weighted images at b-values of b0 (a) and b1000 (b) showing brighter hyperintense signal on high b-value (b) suggestive of diffusion restriction. Smoothened pancreatic margins with clear peripancreatic fat can be appreciated. P: Pancreas

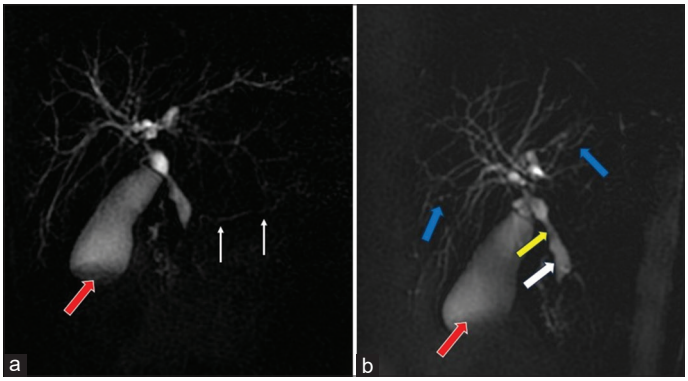

Figure 3: (a and b) Thick slab MRCP image showing alternating areas of dilatation (white arrow) and narrowing (yellow arrow) in CBD with beaded appearance of intrahepatic biliary ducts (blue arrows). Furthermore, there were multiple tiny calculi in GB lumen (red arrow). Thin white arrows indicate MPD. MRCP: Magnetic resonance cholangiopancreatography; CBD: Common bile duct; MPD: Main pancreatic duct, GB: Gall bladder

its normal surface lobulations/clefts [1,3,4]. This appearance can be seen in both computed tomography and MRI. MRI shows a diffusely bulky pancreas with T2 hyperintensity of parenchyma, loss of surface lobulations (featureless pancreatic surface), thin smooth capsule-like rim along the pancreatic surface which may be T2-hypointense (Fig. 1), diffusion restriction in the parenchyma, and minimal to no peripancreatic inflammatory changes (Fig. 2) [1,3,5-7]. In addition, Type-1 AIP may be also accompanied by cholangiopathy with multifocal bile duct narrowing and dilatation (Fig. 3) [1,4,5-7].

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