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CASE REPORT

Isolated Polycystic Disease of the Pancreas

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Abstract

Isolated polycystic disease of the pancreas is an extremely uncommon condition, with very few examples documented in the literature. Pancreatic cystic lesions are usually associated with other genetic disorders that involve other organs as well. However, isolated pancreatic polycystic disease is rare. We reported a symptomatic case of polycystic disease of the pancreas without evidence of cystic lesions in other organs.

Keywords: Pancreatic cysts, Pancreatic insufficiency, polycystic disease, polycystic pancreas

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Introduction

Polycystic disease of the pancreas is also known as dysontogenic cysts of the pancreas [2,3] It usually occurs with cysts in other organs such as the kidneys, liver, spleen, and central nervous system. 10 percent of cases of Polycystic disease of the kidney show cysts in the pancreas [1,3]. This suggests the embryologic connection with different cystic subtypes. Pancreatic cysts can be broadly classified as congenital cysts, developmental cysts, retention cysts duplication cysts, pseudocysts, neoplastic cysts, and parasitic (hydatid) cysts. True cysts of the pancreas occur as a developmental anomaly due to the sequestration of primitive pancreatic ducts [1].

Case Report

A 26-year-old female presented with a complaint of recurrent upper abdominal pain for five months. The pain was vague and located in the epigastric region mainly. Her physical and systemic examination was unremarkable. The bowel sounds were normal. There was tenderness in the epigastric region with no guarding or rigidity. The patient's vitals were normal. Laboratory tests showed a white blood cell count of 10,102 / mm^3, a hemoglobin level of 12.9g/dl, and a platelet count of 255,600 /mm^3.

An ultrasound of the abdomen showed multiple variable-sized cystic lesions involving the head, body, and tail of the pancreas. There was no evidence of cystic lesions in the kidneys and liver as shown in the images. The possibility of Pancreatitis was ruled out as patients had no history of pancreatitis or blunt abdominal trauma. Serum Amylase and Serum Lipase were done and found within normal limits (Serum Amylase – 73 U/L and Serum Lipase – 88 U/L).) (Figures 1 to 3).

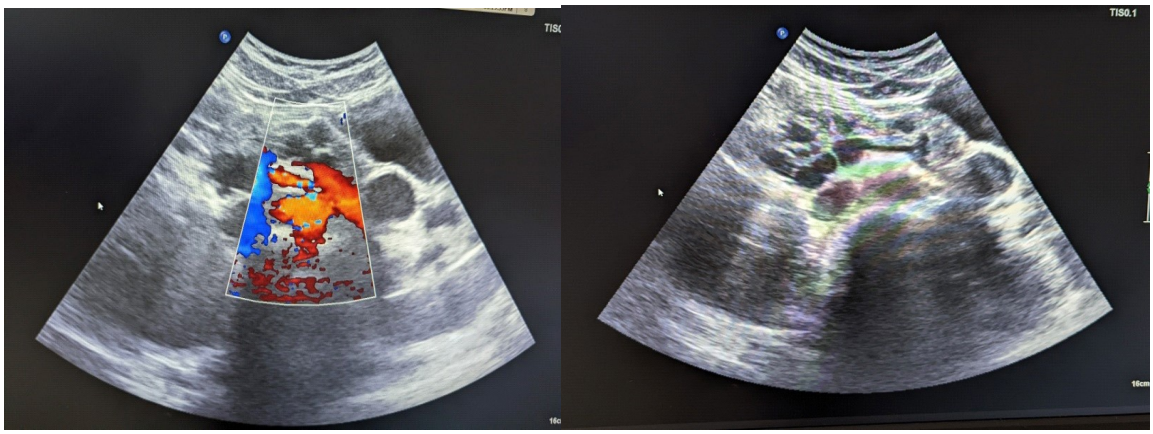

Figure 1. Ultrasound of upper abdomen showing multiple well-defined variable sized anechoic lesions involving almost all of the pancreatic head body and tail. Doppler study reveals absent color flow within the anechoic lesions suggestive of Multiple cystic lesions likely.

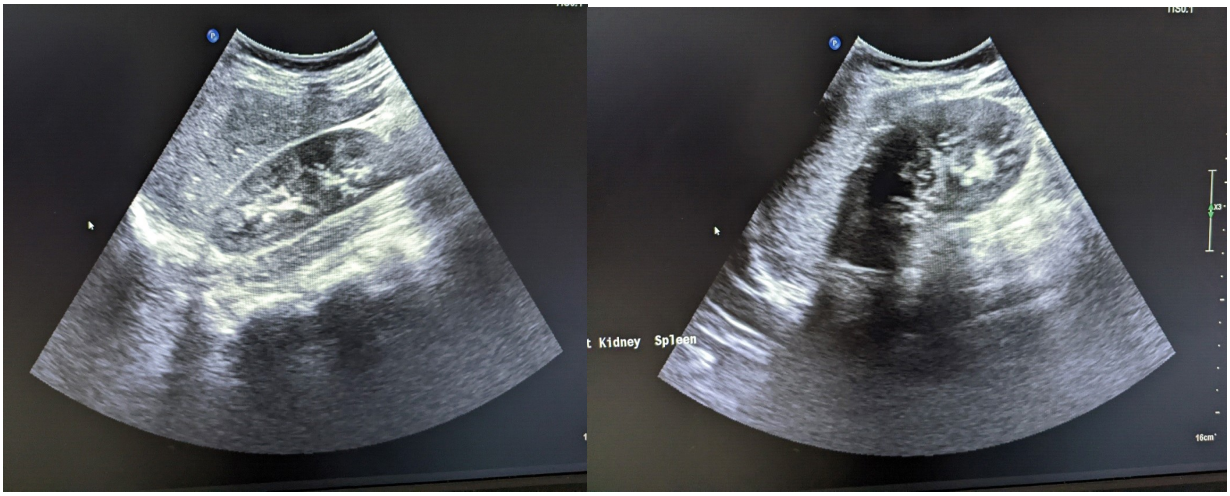

Figure 2. Ultrasound of spleen, right and left kidneys respectively showing normal parenchyma and no evidence of anechoic cystic lesions on either side.

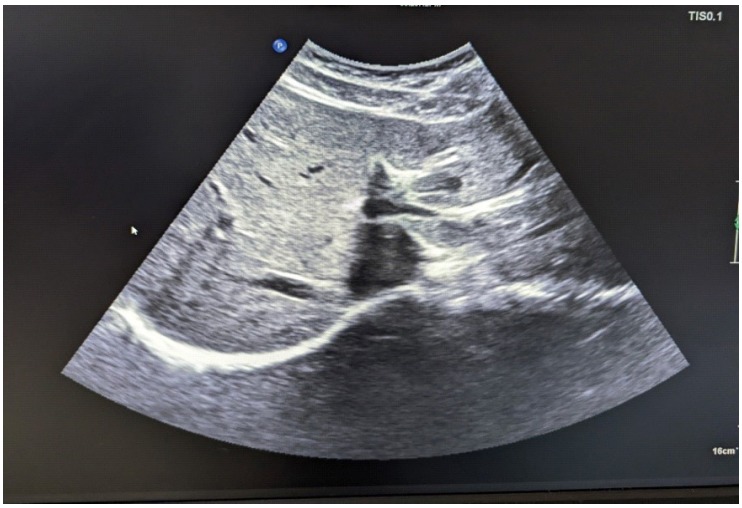

Figure 3. Ultrasound of liver showing normal liver parenchyma with no evidence of focal or diffuse lesion seen.

Contrast enhanced computed tomography of abdomen was performed for further investigation and differential diagnosis. Plain images demonstrated variable sized well well-defined rounded fluid density (10-15 HU) lesions seen in the head, body, and tail region of the pancreas. On arterial and venous phases, the lesions show mild peripheral wall enhancement as shown. The rest of the abdominal study was found unremarkable (Figure 4).

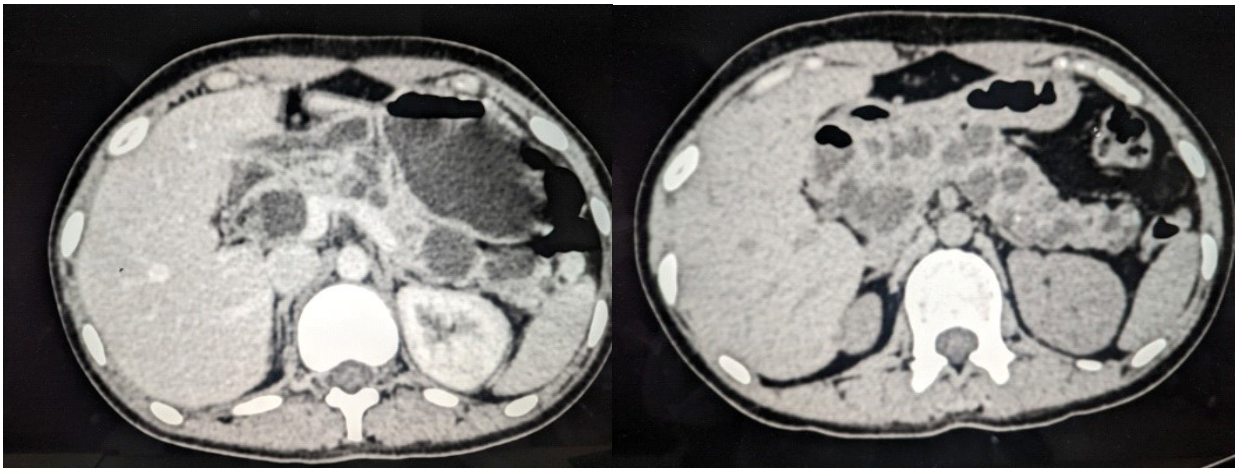

Figure 4. Contrast enhanced CT scan shows mild peripherally enhancing variable sized cystic lesions.

On Magnetic resonance imaging of the abdomen, the lesions appeared well-defined variable sized T2 hyper-intense lesions in the head body, and tail region of the pancreas. The lesion appeared hyper-intense on T2 fat sat images. There was no dilatation of the pancreatic duct and no visible communication of the pancreatic duct with the cysts (Figure 5).

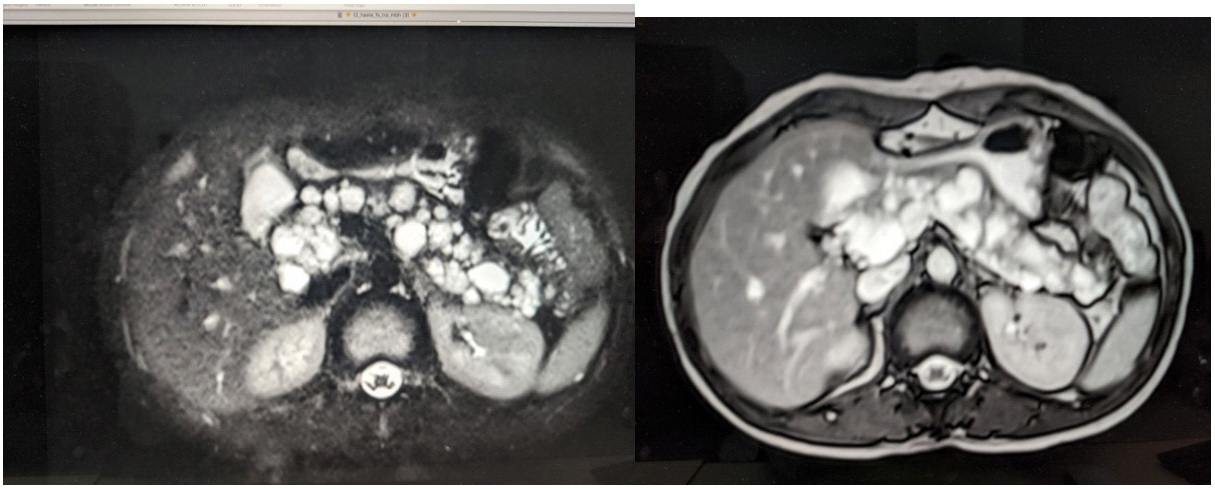

Figure 5. Magnetic resonance imaging of abdomen T2 weighted image and T2 fat sat images showing multiple well defined rounded variable sized thin-walled hyper intense lesions are seen in the head, body, and tail of the pancreas.

MRI brain was normal. Tumor markers (CA – 19-9 and CEA) were normal. A mutation of the VHL tumor suppressor gene on the short arm of chromosome 3 was not found. So, Von Hippel – Lindau disease was ruled out.

Discussion

A vast and intricate network of ducts in the liver and pancreas carries bile and pancreatic secretions to the gut lumen. The epithelial cells that make up the ductal trees originate from the endoderm germ layer lining the primitive gut. Polycystic disease in the liver and pancreas can result from abnormal growth of these duct networks, such as ducts with expanded lumens and multiple cysts. Primitive duct sequestration brought on by developmental abnormalities results in the formation of true cysts [1].

True or congenital pancreatic cysts are distinguished from pseudo cysts by the presence of a true cuboidal or columnar epithelial lining on their inner surface. Thus, histological analysis is the basis for differentiating between actual cysts and other cystic lesions. Pancreatic enzymes are abundant in the cyst fluid, and pseudocysts frequently experience episodes of acute pancreatitis. Single or multiple congenital pancreatic cysts can occur. They could be associated with other systemic diseases or isolated. Multiple cysts are often associated with cystic disease of other organs. Autosomal dominant polycystic kidney disease, Von Hippel-Landau disease, cystic fibrosis, and Beckwith-Wiedemann syndrome are syndromes linked to numerous pancreatic cysts [1].

Although they seldom cause symptoms, congenital cysts can cause vomiting, distension in the abdomen, and vague abdominal pain. Pancreatitis or hepatitis. Particularly useful for differentiating between solid and cystic conditions is ultrasound. Color Doppler imaging can establish if the lesions are vascular because cystic lesions do not exhibit color flow. At some point, characterization and localization of the cysts depend on cross-sectional imaging. The cystic nature of the lesions, as well as the interaction between the cyst and surrounding tissues and the expansions of the cysts, can be established by CT and MRI scans. But MRI does a better job of showing these things than a CT scan does. Simple cysts on MRI appear hyper-intense on T2 weighted images and hypo intense on T1 weighted images. Features of computed tomography include round or oval, homogenous, hypodense appearance, no discernible wall, internal septa, calcification, hemorrhage, or mural nodule, and an attenuation coefficient of 0–20 HU. Patients who have no symptoms can still be treated with follow-up and no interventions. However, a segmental pancreatectomy is required to remove the dominant cyst in symptomatic patients [4].

In conclusion, we reported a rare case of isolated polycystic pancreatic disease on magnetic resonance imaging, contrast-enhanced tomography, and ultrasonography. It is essential to exclude related syndromes before making a final diagnosis [4,5]. When diagnosing isolated polycystic pancreatic disease, MRI is significant.

Conflicts of interest

The authors declares that they do not have conflict of interest.

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