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CASE REPORT

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Isolated polycystic pancreatic disease

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ABSTRACT

Congenital pancreatic cysts, also called true cysts, refer to non-neoplastic and non-inflammatory cystic lesions of the pancreas. Single true cysts of the pancreas are rare and they are mostly seen in newborns. Multiple true cysts or polycystic pancreatic disease is usually associated with other genetic disorders that involve cystic disease of the other organs. However, isolated polycystic pancreatic disease is very rare and there are very limited cases in the literature. A 22-year-old female presented with vague epigastric pain. Abdominal magnetic resonance imaging demonstrated multiple cysts at the pancreas with no other organ involvement. The patient was diagnosed to have isolated polycystic pancreatic disease.

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Pancreas; polycystic pancreas disease; magnetic resonance imaging

Introduction

Prevalence of pancreatic cysts is estimated as 2.4%–24% in imaging and autopsy series, and pseudocysts are the most common cystic lesions of the pancreas [1]. Congenital pancreatic cysts refer to non-neoplastic, non-inflammatory cysts of the pancreas arising from abnormal segmentation of primitive pancreatic ducts and they are very rare conditions. Solitary congenital cysts may be seen isolated which means there is no other organs/systems involvement, but multiple congenital cysts are usually associated with cystic diseases of the other organs [1]. Polycystic pancreatic disease without other organ/system involvement is extremely rare [2]. Herein, we report a patient with an isolated polycystic pancreatic disease.

Case Presentation

A 22-year-old woman was admitted to our hospital with the complaints of vague epigastric pain for the last 1 month. Her physical examination revealed no abnormality. Patient's vital signs were all within normal limits. Laboratory tests revealed a white blood cell count of 8,600/mm^3, a hemoglobin level of 7.3 g/dl, and a platelet count of 524,000/mm^3. An infectious etiology was not considered. Ultrasound (US) examination demonstrated multiple cystic lesions in the pancreas and there was no abnormality of the other abdominal organs. Subsequent to this finding, suspicion of pancreatitis had raised and the blood level of pancreatic amylase and lipase evaluated. They were also within the normal limits. Abdominal magnetic resonance imaging (MRI) was performed for further investigation and differential diagnosis. MRI and MR cholangiopancreatography demonstrated multiple different sized cysts replacing almost all of the pancreas. Cysts were homogeneously hyperintense on T2-weighted (T2W) axial images and hypointense on T1-weighted (T1W) axial images with no evidence of intracystic hemorrhage (Fig. 1a and b). Contrast-enhanced images obtained after the administration of gadolinium-based contrast agent revealed no enhancement or solid component of the cysts suggesting neoplasm. No diffusion restriction was seen on diffusion-weighted imaging.

Abdominal MRI also demonstrated that there was no other abdominal organ involvement of the cysts. Coronal MR cholangiopancreatography image (Fig. 1c) demonstrated multiple hyperintense, rounded, well-defined cysts with very thin regular walls. There was no dilatation of the main

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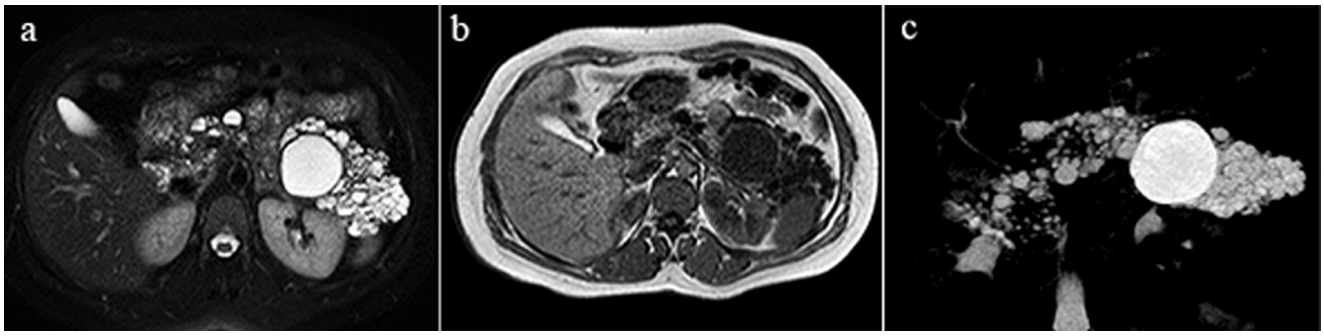


Figure 1. MRI examination demonstrates multiple cystic lesions of the pancreas that are hyperintense on axial fat-suppressed T2-weighted images (a) and hypointense on axial T1-weighted images (b). MR cholangiopancreatography image (c) demonstrates multiple hyperintense cystic lesions in the pancreas.

pancreatic duct and no identifiable communication of the pancreatic duct with the cysts. There was no abnormal finding in brain MRI. So, we diagnosed the patient as isolated polycystic pancreatic disease with no other organ/system involvement. Six and twelve months later, US revealed a stable appearance of the cysts and the patient was relieved from her symptoms after analgesic drugs. We diagnosed the patient as having isolated polycystic pancreatic disease and decided to follow-up.

Discussion

Cystic masses of the pancreas may be classified as congenital, inflammatory, or neoplastic [3]. Pseudocysts are the most common cystic lesions of the pancreas and they constitute more than 75% of all cystic lesions. Congenital pancreatic cysts of the pancreas, also called true pancreatic cysts, have an epithelial lining in their inner surface which is absent in pseudocysts. Therefore, differential diagnosis of a true cyst from other cystic lesions is in fact based on histopathological examination.

Congenital pancreatic cysts are usually solitary but they may be solitary or multiple. They may be isolated or may be associated with other systemic diseases. Solitary cysts are usually seen isolated whereas multiple pancreatic cysts are usually associated with cystic diseases of other organs, including autosomal dominant polycystic kidney disease, Von Hippel–Lindau disease, cystic fibrosis, and Beckwith–Wiedemann Syndrome [1]. Isolated form of the polycystic pancreatic disease is extremely rare and only a few cases were reported in the medical literature [2,4].

Imaging modalities show cystic nature of the lesions. MRI demonstrates simple cysts that are hypointense on T1W images and hyperintense on T2W images. Computed tomography features are round/oval homogeneous hypodense appearance with an attenuation coefficient of 0–20 HU with no perceptible wall, internal septa, calcification, hemorrhage, or mural nodule [2]. Treatment option for dominant symptomatic cyst is segmental pancreatectomy with cystectomy [2]. Asymptomatic diffuse pancreatic involvement may be followed-up and therapeutic intervention may not be needed [2].

In conclusion, we reported a rare case of the polycystic pancreatic disease with MRI findings. It is essential to exclude associated syndromes for definitive diagnosis. MRI plays an important role in the diagnosis of isolated polycystic pancreatic disease. Both clinicians and radiologists should be familiar with its imaging features.

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