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Case Report

An Unusual Presentation of Pancreatic Dorsal Agenesis

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Abstract

Dorsal pancreatic agenesis is a rare congenital anomaly. Besides, polysplenia syndrome concomitant with situs inversus totalis and distal pancreatic hypoplasia are rare congenital anomalies. We aimed to present such a rare congenital variation with computed tomography, magnetic resonance, and magnetic resonance cholangiopancreatography findings.

Keywords: Polysplenia syndrome, situs inversus totalis, pancreas, pancreatic hypoplasia

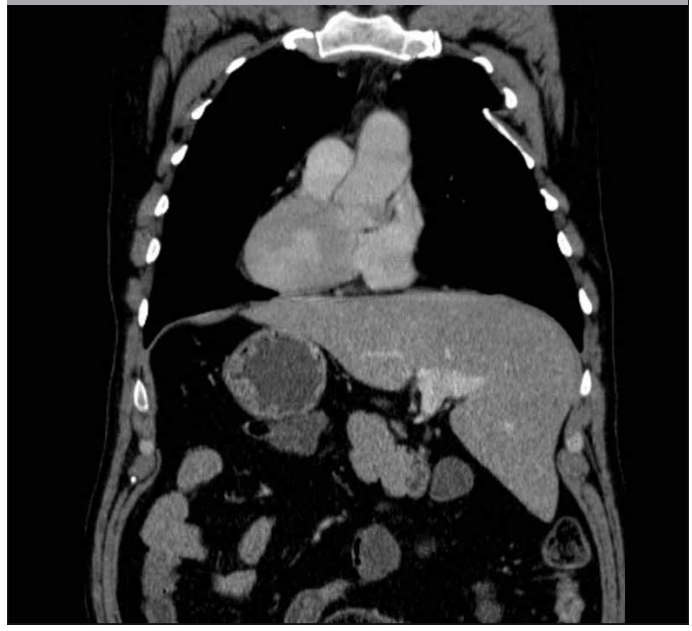
INTRODUCTION

Polysplenia syndrome (PS) is a rare syndrome of congenital abnormalities involving the cardiovascular system and visceral organs in the abdomen that are diagnosed incidentally in adults. Cardiac anomalies are less common in PS than are in asplenia syndrome (1). PS concomitant with situs inversus totalis (SIT) and pancreatic hypoplasia is very rare in the general population. Short pancreas and pancreas hypoplasia may be observed in PS due to dorsal pancreas agenesis (2). Annular pancreas and semiannular pancreas may also be observed in PS (3, 4). Isolated dorsal pancreas agenesis is very rare in the general population.

CASE PRESENTATION

A 51-year-old male presented to the emergency department with a 6-month history of epigastric abdominal pain. He also reported nausea and vomiting. He denied fever, weight loss, heart disease, previous surgery, and any history of trauma. A physical examination revealed mild tenderness to palpation in the right upper quadrant. Chest radiography showed dextrocardia. Laboratory test results and electrocardiographic findings were unremarkable. Contrast-enhanced computed tomography (CT) and magnetic resonance cholangiopancreatography (MRCP) were performed (Figures 1-4). His pain reduced with proton pump inhibitors.

Figure 1. Contrast-enhanced thorax and upper abdomen CT scan with coronal reformatted image obtained with standard soft tissue settings (window width, 350 Hounsfield Unit (HU), window center, 50 HU) shows dextrocardia, a left-sided liver, a right-sided stomach, and left-sided anterior costal fusion



Contrast-enhanced abdominal CT revealed SIT in the abdominal solid organs and colon and small bowel (Figure 1 and 2a). The liver and gall bladder were located in the left upper quadrant. The stomach was radiologically normal and was located in the right upper quadrant. The inferior vena cava (IVC) was left-sided. The cecum and appendix were located in the left lower quadrant, and all small bowel segments were seen in the right upper and right lower quadrants (Figure 2b). The Treitz ligament was seen at the duodenojejunal junction, as normal. In the right upper quadrant, numerous spherical nodular spleens were seen (Figure 5). In addition, the head of the pancreas was observed in the left paramedian region, to the anterior of the portal vein. The body and tail of the pancreas could not be visualized in CT images (Figure 6). The third

Figure 2. a, b. Contrast-enhanced abdomen coronal-reformatted CT image reveals SIT in the abdominal solid organs, colon, and small bowels (a). Coronal CT images show cecum and appendix (red arrows) located in the left lower quadrant (b)

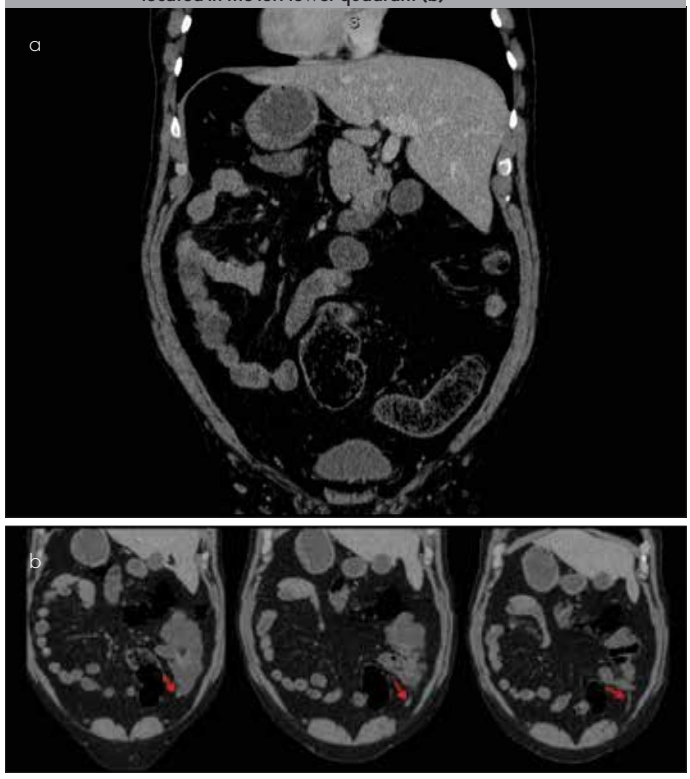


Figure 3. Coronal T2-weighted fast spin-echo MRI (repetition time ms/echo time ms, 698/80) shows dorsal pancreas hypoplasia with normal parenchymal signal intensity and a normal main pancreatic duct (arrows)

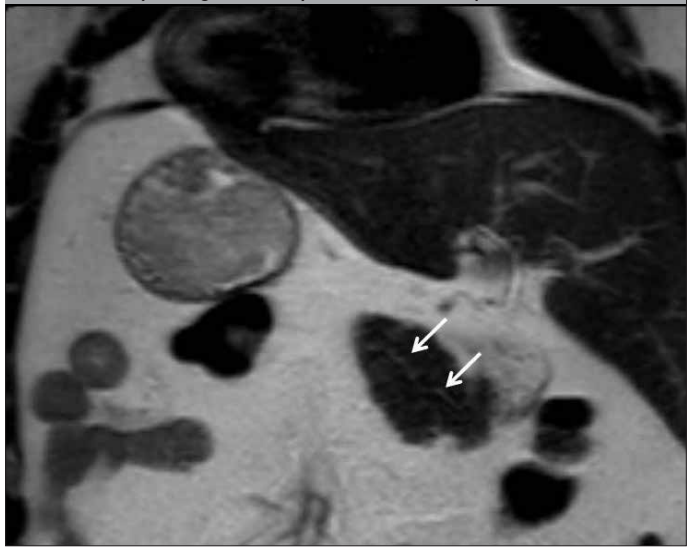


Figure 4. Single-slab fat-saturated HASTE image in the coronal oblique plan (repetition time ms/echo time ms, 8000/900) indicates a normal main pancreatic duct (arrows), and there was no finding of any other accompanying pancreatic anomaly

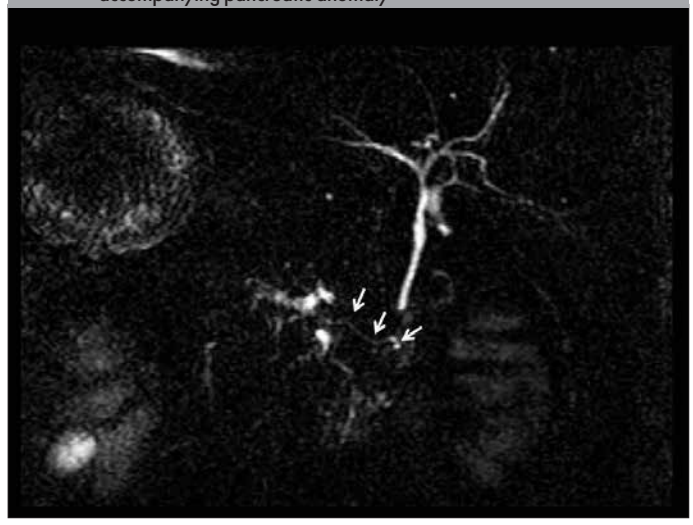
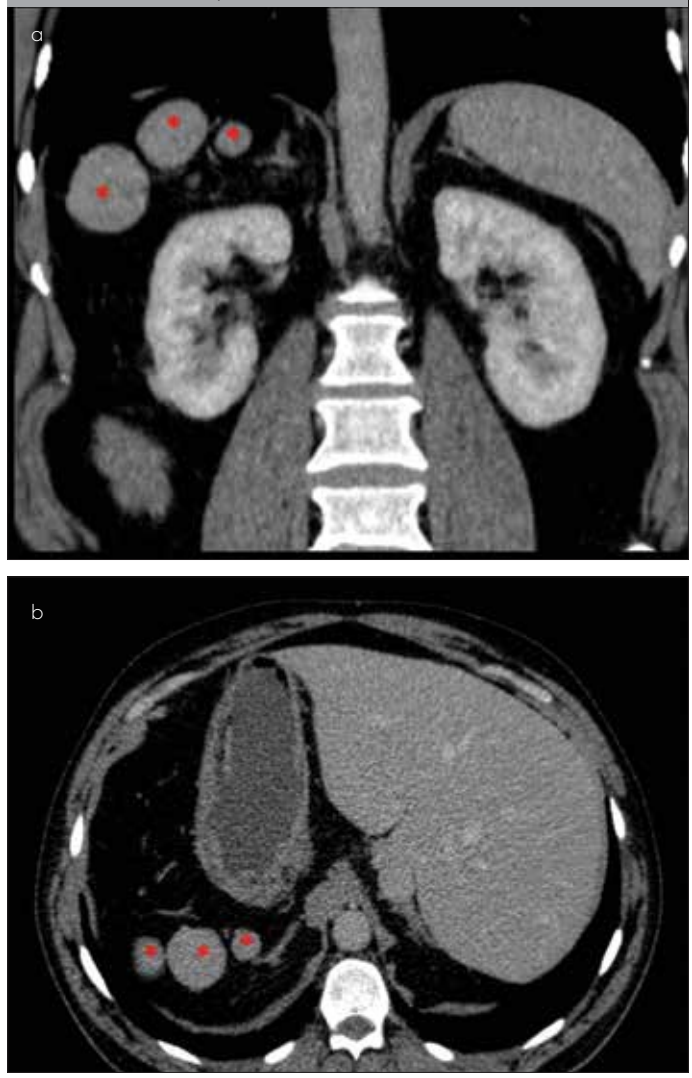
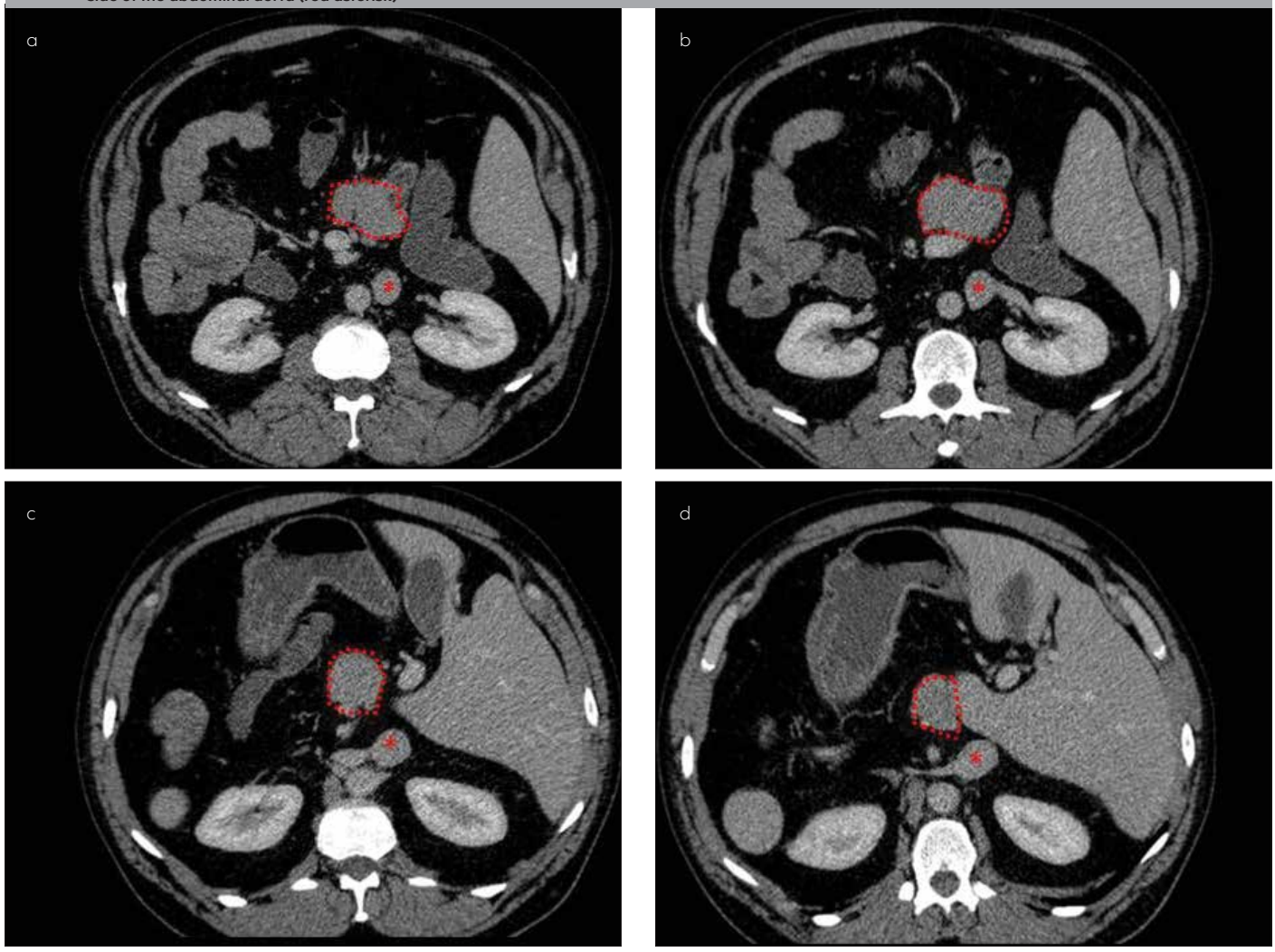


Figure 5. a, b. Coronal and (b) axial CT scans reveal nodular soft tissue density masses according to polysplenia in the left upper quadrant (red asterisks)



and fourth anterior costal fusion was also observed on the left side (Figure 1). MRCP was performed to further evaluate the pancreatic duct and biliary system. MRCP showed the complete absence of the body and tail of the pancreas. The main pancreatic duct was short in MRCP images, whereas no abnormal finding was observed in the pancreatic duct or biliary system (Figures 3 and 4).

Figure 6. Pancreatic hypoplasia was seen in contrast-enhanced axial CT images at the level of the pancreas (stippled red curves). The IVC was observed on the left side of the abdominal aorta (red asterisk)



Both magnetic resonance imaging (MRI) and CT demonstrated polysplenia, SIT, and pancreatic hypoplasia as incidental findings. Other findings were within normal limits and there was no evidence of congenital heart disease (CHD). The patient underwent endoscopy for epigastric pain and had mild gastritis. He was treated with proton pomp inhibitors.

DISCUSSION

Situs anomalies are rare congenital disorders that can lead to confusing differential diagnoses. Three main types of situs variation have been described previously (5). "Situs solitus" indicates that the heart, abdominal vascular structures, intestinal segments, and visceral organs are in the normal locations. The incidence of CHD is less than 1% in this condition. "Situs inversus" represents a left-to-right reversal of the viscera and gastrointestinal system, relative to the normal position. It is inherited in an autosomal-recessive pattern. SI has two different presentations: SI with dextrocardia and SI with levocardia. SI with dextrocardia is more common than the other type, and is also known as "situs inversus totalis." CHD is observed in 3%–5% of patients with SI with dextrocardia and all patients affected with SI with levocardia. "Situs ambiguous," also known as heterotaxia, is described as abnormal positioning of visceral abdominal organs, major vessels, and intestinal segments. The small bowels are located in the right upper and right lower quadrants, and the cecum, ascending colon, and transverse colon are displaced medially. The descending colon and recto-sigmoid segment are situated normally. It is seen usually with dextrocardia. Two major types of situs ambiguous (SA) have been described: SA with polysplenia (also known as PS, left isomerism, or bilateral left-sidedness) and SA with asplenia (also known as right isomerism or bilateral right-sidedness). The incidence of CHD is very high in both SA types. SA with polysplenia (left isomerism) has multiple spleens, located in the left upper quadrant and a midline-located liver. The stomach is usually in the normal left upper quadrant location. SA with asplenia (right isomerism) is characterized by mirror-image locations of the abdominal visceral organs and stomach. In this type of SA, the spleen is congenitally absent.

Polysplenia syndrome is a rare disorder that belongs to the situs anomalies. It is usually diagnosed when the patient is a child due

to CHD. Most patients with PS die before 5 years of age, secondary to severe cardiovascular disorders. The incidence of PS is one case per 100,000 live births (6). The most common radiological manifestations of PS include polysplenia multiple nodular spleens in the course of the greater curvature of the stomach in the left upper quadrant, left-sided IVC, midline liver, preduodenal portal vein, short pancreas, malrotation of the intestines, and dextrocardia (1, 2, 7). Classically, PS is usually seen with left isomerism. The presence of SIT and SA in the same patient is extremely rare. In our case, costal fusion was other concomitant radiological finding. These findings are not specific to PS or SIT.

In conclusion, PS and SIT are rare congenital abnormalities that can be seen with congenital pancreatic anomalies. The association of SIT and PS is extremely rare. These abnormalities may be detected incidentally in adults. Abdominal CT typically provides correct diagnoses in patients with SIT and PS.

Informed Consent: Written informed consent was obtained from patient who participated in this case.

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