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Gastrointestinal

Complete dorsal pancreatic agenesis and unilateral renal agenesis

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ABSTRACT

Dorsal pancreatic agenesis is a very rare congenital anomaly. Unilateral renal agenesis, on the other hand, is a relatively common congenital anomaly, although its etiology is not fully understood. Renal and pancreatic embryologic development appears to be nonrelated. We report a case of a 34-year-old man who was referred to our hospital for evaluation of cholestasis and microalbuminuria. Ultrasound and magnetic resonance imaging examinations showed empty right renal fossa and absence of the pancreatic neck, body, and tail. Our case report is the second case of a dorsal pancreatic agenesis and unilateral renal agenesis in a young male patient.

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Introduction

Congenital pancreatic anomalies are not frequently encountered in radiological examinations. In fact, only around 100 cases of dorsal pancreatic agenesis, probably the least common of all, have been reported in the literature.

Unilateral renal agenesis (URA) is the most common congenital anomaly of the urinary system. To the best of our knowledge, there is only one previously published case report of dorsal pancreatic agenesis and URA [1].

Case report

A 34-year-old man was referred for a nephrology and gastroenterology appointment at our hospital because of microalbuminuria and cholestasis. The fasting blood glucose level was normal on repeated examinations.

The patient was asymptomatic, with no relevant past medical history, and physical examination showed no abdominal abnormalities besides a syndromic facies. A sonographic study revealed a solitary left kidney; no right kidney was found in the abdominopelvic cavity and only the pancreatic head was clearly visualized by ultrasound. The patient underwent abdominal and pelvic magnetic resonance imaging, which confirmed an empty right renal fossa. No ectopic kidney was found. The pancreatic neck, body, and tail were missing, corresponding to a medical condition of complete dorsal pancreatic agenesis. The pancreatic bed was occupied by bowel loops. The pancreatic head and uncinate process were normal and no parenchymal calcifications were noted. The ventral pancreatic duct of Wirsung and the common bile duct were not dilated. The rest of the magnetic resonance imaging examination of the abdomen and pelvis was normal (Figs. 1-4).

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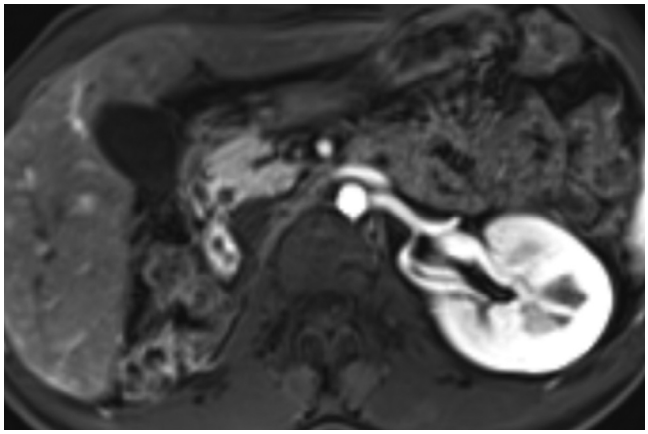

Fig. 1 – Axial T1 weighted MR image shows a normal pancreatic head but no neck, body or tail region, compatible with complete dorsal pancreatic agenesis. The rest of the pancreatic bed is occupied with bowel loops. There is no dilatation of the ventral Wirsung duct. Only the left kidney is present.

Imaging diagnosis was one of complete dorsal pancreatic and right renal agenesis.

Discussion

Dorsal pancreatic agenesis is an extremely rare congenital anomaly; the first case was reported in 1911 [2] and approximately 100 cases have been reported so far [3].

The pancreas is a retroperitoneal organ composed of a head, neck, body, and tail. During embryogenesis, it develops around the fourth week of gestation from the ventral and dorsal buds, arising from the second part of the duodenum.

The pancreatic neck, body, and tail develop from the dorsal bud and drain through the accessory duct of Santorini and the minor papilla. The ventral bud forms the major part of the head and uncinate process. During the seventh week of gestation, the ventral bud rotates dorsally around the duodenum to fuse with the dorsal bud.

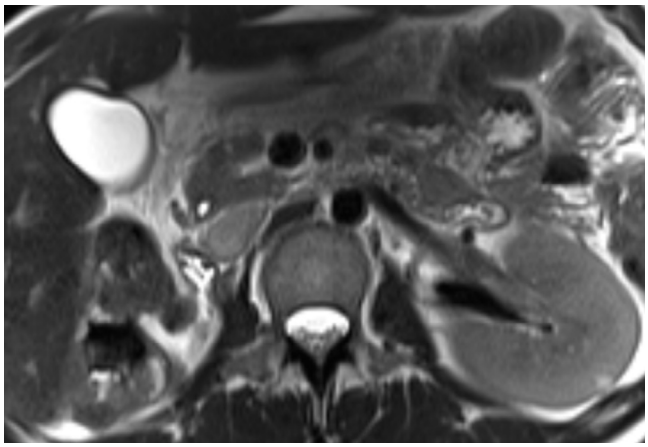

Fig. 2 – Axial T2 weighted MR image shows a normal pancreatic head but no neck, body or tail region. There is no dilatation of the ventral Wirsung duct. Only the left kidney is present.

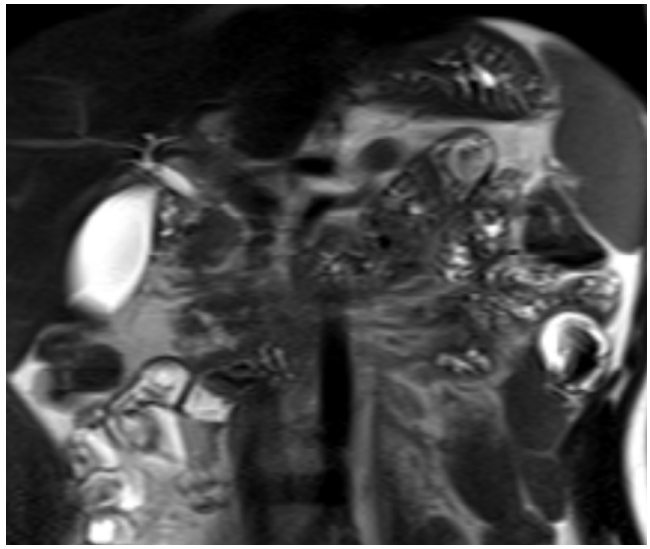

Fig. 3 – Coronal T2 weighted MR image shows that the biliary ducts are not dilated and only the pancreatic head is seen.

Dorsal pancreatic agenesis may be either complete or partial. In complete agenesis, the neck, body, tail, duct of Santorini, and minor duodenal papilla are absent; in partial agenesis, the body, the duct of Santorini, and the minor duodenal papilla are preserved.

Abnormal embryogenesis can lead to a failure in the normal development of the dorsal pancreas, resulting in complete dorsal pancreatic agenesis [4].

Other congenital anomalies have been associated with dorsal pancreatic agenesis, such as coarctation of the aorta, Fallot tetralogy, ventricular septal defects, heterotaxy, polysplenia

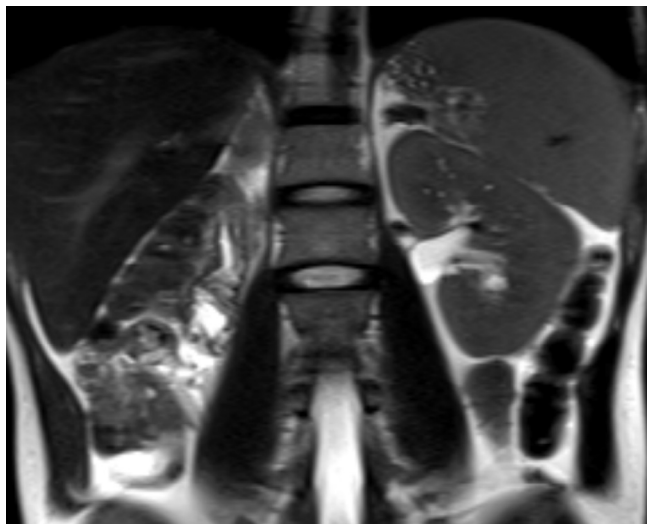

Fig. 4 – Coronal T2 weighted MR image shows the right renal agenesis, with bowel loops in the right renal fossa. The left kidney has a normal appearance.

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