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Case Report

Radiology Section

Complete Dorsal Pancreatic Agenesis and Unilateral Renal Agenesis: First Case Report and Review of Literature

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ABSTRACT

Dorsal pancreatic agenesis is very rare congenital anomaly of pancreas. Patients with this anomaly are usually asymptomatic or may present with abdominal pain or acute or chronic pancreatitis. We report the case complete dorsal pancreatic agenesis and absent right kidney in a young patient not described previously in literature.

Keywords: Embryogenesis, Endoderm, Mesoderm, Metanephrons

CASE REPORT

A 24 years old male patient presented with non specific and non localized pain in abdomen. Patient's symptoms were not related to meal and past history was unremarkable. No evidence of tenderness or rigidity noted on clinical examination. Routine blood analysis including the fasting blood sugar was normal. Patient was referred for the sonographic examination of abdomen. Sonography study revealed absence of right kidney in renal fossa or elsewhere in abdomino-pelvic region. Rests of the examination was normal. Patient underwent MRI (Plain) examination of abdomen and pelvis after proper consent to rule out ectopic or atrophic right kidney.

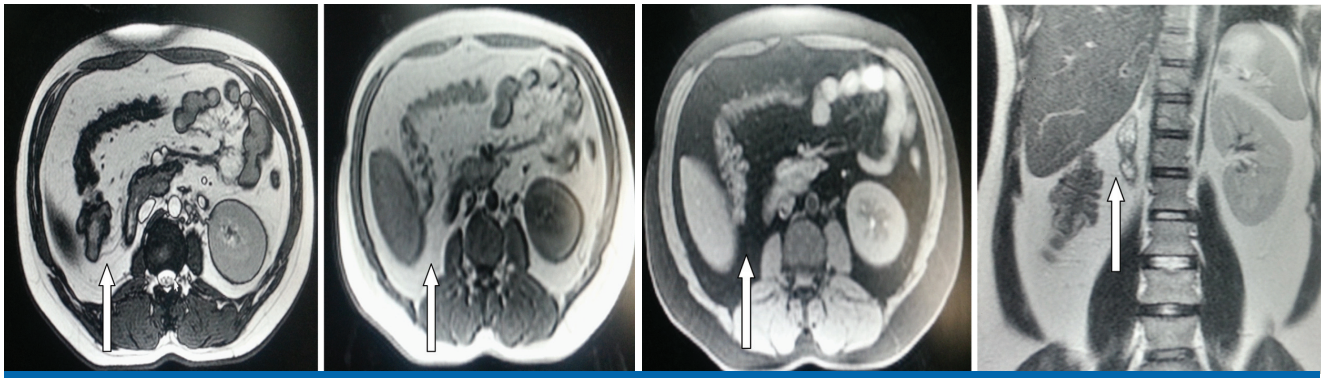
MRI study revealed empty right renal fossa [Table/Fig-1-4]. No evidence of ectopic or atrophic kidney noted in the whole abdomen. Detailed MRI examination showed absence of pancreatic neck, body and tail region [Table/Fig-5-8]. Pancreatic bed was occupied by the bowel loops. These findings were compatible with complete dorsal pancreatic agenesis. Rests of the MRI examination of abdomen and pelvis was normal.

Imaging findings were compatible with the complete dorsal pancreatic and right renal agenesis. We could not identify cause of non specific abdominal pain in patient. Dietary modification like low fat-diet was adviced to the patient. Patient had a symptomatic relief on follow-up.

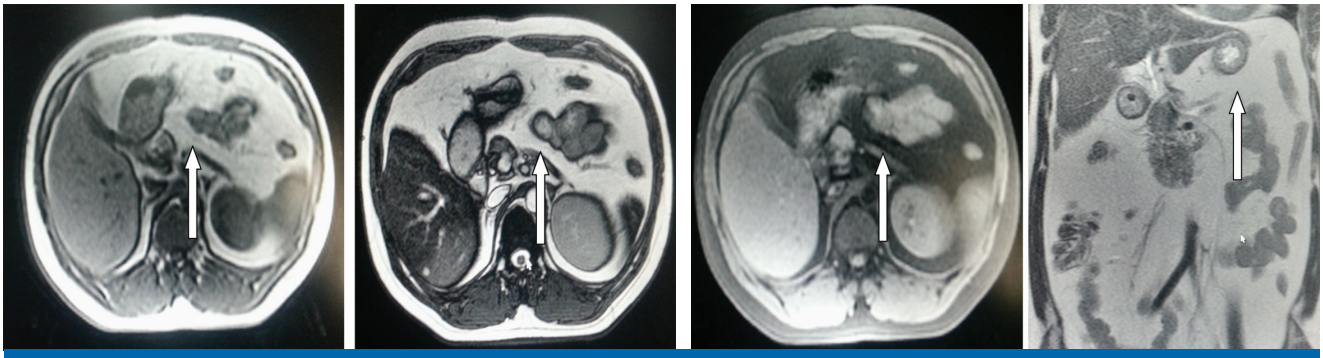
Embryological development of pancreas and urinary system is different, pancreas is endodermal whereas kidney is mesodermal in origin. Co-existence of dorsal pancreatic and unilateral renal agenesis is not described previously in the literature to the best of our knowledge.

DISCUSSION

Pancreas consist of head, neck, body and tail parts and located in retroperitoneal space extending between second part of duodenum and splenic hilum. During embryogenesis it develops from the two focal outpouchings namely ventral



[Table/Fig-1 to 4]: Axial T2, Axial T1, Axial fat sat and Coronal T2 - Images with empty right renal fossa.



[Table/Fig-5 to 8]: Axial T1, Axial T2, Aixal fat sat and Coronal T2 images -Absent pancreatic neck, body & tail with bowel loops into pancreatic bed.

and dorsal buds arising from the second part of duodenum at 4 weeks of gestation. Pancreatic neck, body, and tail develops from dorsal bud and drain through the accessory duct of Santorini and minor papilla. Major part of the head and uncinate process are developed from ventral bud. During the 7th week of gestation, the ventral bud rotates dorsally around the duodenum to fuse with the dorsal bud and form the mature gland [1].

In complete agenesis of the dorsal pancreas, the neck, body, and tail of the pancreas, the duct of Santorini, and the minor duodenal papilla are all absent [2]. With partial agenesis of the dorsal pancreas, the size of the body of the pancreas varies, there is a remnant of the duct of Santorini, and the minor duodenal papilla is present [3]. Dorsal pancreatic agenesis reported to occur with very rare conditions including heterotaxy, polysplenia syndrome and congenital heart diseases [4]. Exact etiogenesis of this condition is unknown however Autosomal dominant, X-linked inheritance and sporadic occurrence have been described. Dorsal pancreatic agenesis is usually asymptomatic condition often diagnose incidentally on imaging. It can presents clinically with abdominal pain which can be due to pancreatitis. Hyperglycaemia is seen in approximately 50% of the affected individuals [5].

On USG, dorsal pancreatic agenesis is diagnosed when the part of head and uncinate process of the pancreas are seen while the body and tail of the pancreas are not visualized, and the pancreatic bed anterior to the splenic vein is replaced by the stomach or bowel loops [6]. CT-scan demonstrate empty pancreatic fossa with deficient pancreatic body and tail and stomach or gut loops occupying the pancreatic fossa anterior to the splenic vein, also know as the dependent stomach or dependent intestine signs [7]. MRI can evaluate the pancreatic ductal and parenchymal anatomy clearly and helps in diagnostic confirmation [8]. There is an apparent increased risk of developing pancreatic cancer in these patients [9]. The definitive treatment in such cases is total pancreatectomy. Diabetes mellitus with insulin therapy is the complication of pancreatectomy.

Differential diagnosis of dorsal pancreatic agenesis is lipomatosis and pseudoagenesis. Chronic pancreatitis may result into atrophy of the pancreatic body and tail with sparing of the head and uncinate process. Absence of short dorsal duct of Santorini in dorsal pancreatic agenesis helps in differentiating it from pseudo-agenesis and lipomatosis [6].

The genitourinary system develops from the intermediate plate mesoderm. The kidneys develop from the metanephros which appears in the 5th week of embryonic life. Renal agenesis refers to a congenital absence of one or both kidneys [10]. Renal agenesis results from the embryologicalnon development of the metanephros with complete absence of a kidney [10]. Renal agenesis occurs usually occurs at (6-7th) weeks in early gestation with unknown etiology [11]. Unilateral renal agenesis is non fatal and common compared to fatal bilateral renal agenesis [12]. Renal agenesis can be associated with a number of chromosomal abnormalities including trisomy 13, trisomy 18, trisomy 21, and VACTERL association [12]. Occasionally patients with unilateral renal agenesis, develop secondary hypertension [13]. Sonography can easily detect renal agenesis with empty renal fossa. CT and MRI imaging are helpful to differentiate ectopic or atrophic renal disease from agenesis.

The embryological development of the pancreas and kidney appears completely different with the pancreas being endodermal origin and the genitourinary system mesodermal in origin. Anupam Lal et al., [14] reported the case of dorsal pancreatic agenesis with associated pancake kidney and bicornuate uterus. Our case is unique because complete dorsal pancreatic agenesis with associated unilateral renal agenesis is not previously described in the literature to the best of our knowledge.

CONCLUSION

Dorsal pancreatic agenesis is very rare condition and association of unilateral renal agenesis is not reported

previously in the literature. In our case dorsal pancreatic agenesis was the incidental imaging finding. Our case is unique not only because of its rarity but also because of its clinical implication for proper patient management.

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