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## Research Article

### DORSAL PANCREAS AGENESIS ASSOCIATED WITH UNFUSED CROSSED RENAL ECTOPIA- A RAREST OF THE RARE ANOMALIES

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#### ABSTRACT

Dorsal pancreatic agenesis is an extremely rare developmental anomaly of gastrointestinal system. It is mostly an incidental finding and detected in adulthood. Pancreatic anomaly associated with renal positional defect is very rare association. We have reported a case of complete dorsal pancreas agenesis associated with crossed renal ectopia in a 30year-old gentleman who was investigated for gastric malignancy. Contrast enhanced computed tomography (CECT) abdomen is an important imaging to see the anomalous pancreas, abnormal renal position and evaluation of gastric malignancy for its further management. Upper abdominal magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP) is a non-invasive imaging to see the pancreatic duct.

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#### INTRODUCTION

Dorsal pancreatic agenesis is an extremely rare developmental anomaly of gastrointestinal system. It is mostly an incidental finding and detected in adulthood. Pancreatic anomaly associated with renal positional defect is very rare association. Only a few case reports of this anomaly have been published in the literature. The anomaly is asymptomatic in most of the patient; however it can be present with nonspecific symptoms such as abdominal pain and acute or chronic pancreatitis. Sometimes it can present with features of diabetes mellitus due to absence of islet cell in pancreas tail agenesis (Mortele et al, 2006). Renal anomaly can be positional or fusion anomalies. Horseshoe kidney is the most common urogenital anomalies followed by crossed renal ectopia. In crossed renal ectopia, both kidneys are located on the same side and can be with fusion (85%) or without fusion (<10%) (Lal et al, 2015). We have reported a case of complete dorsal pancreas agenesis associated with crossed renal ectopia in a 30year-old gentleman who was investigated for gastric malignancy.

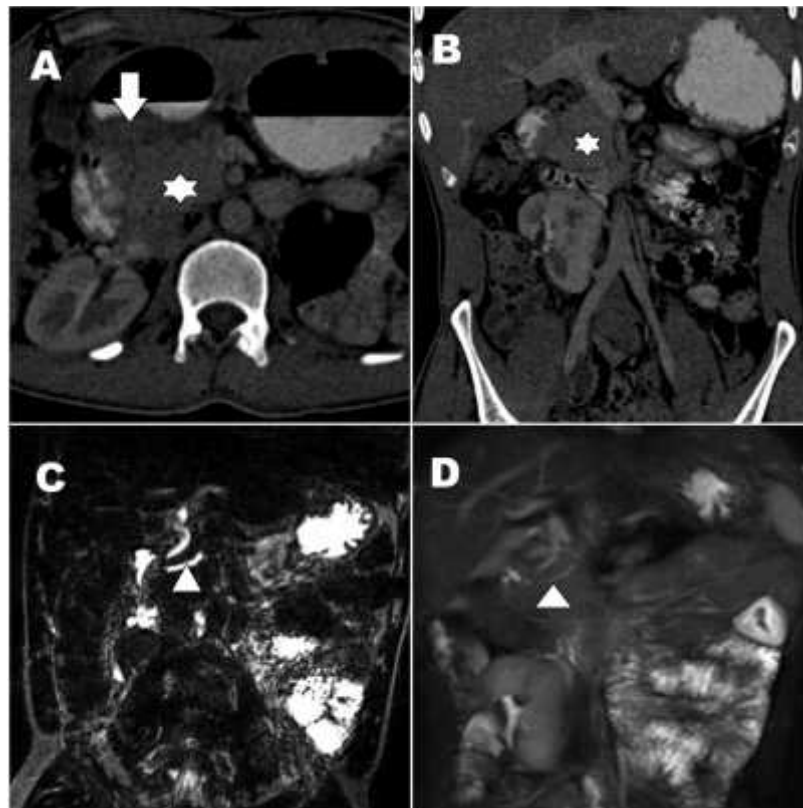
#### CASE REPORT

A 30-year-old male patient was referred to the department of radiology & imaging for evaluation of a biopsy proven gastric adenocarcinoma. On general examination, patient was conscious and cooperative. Vitals are stable. There was mild

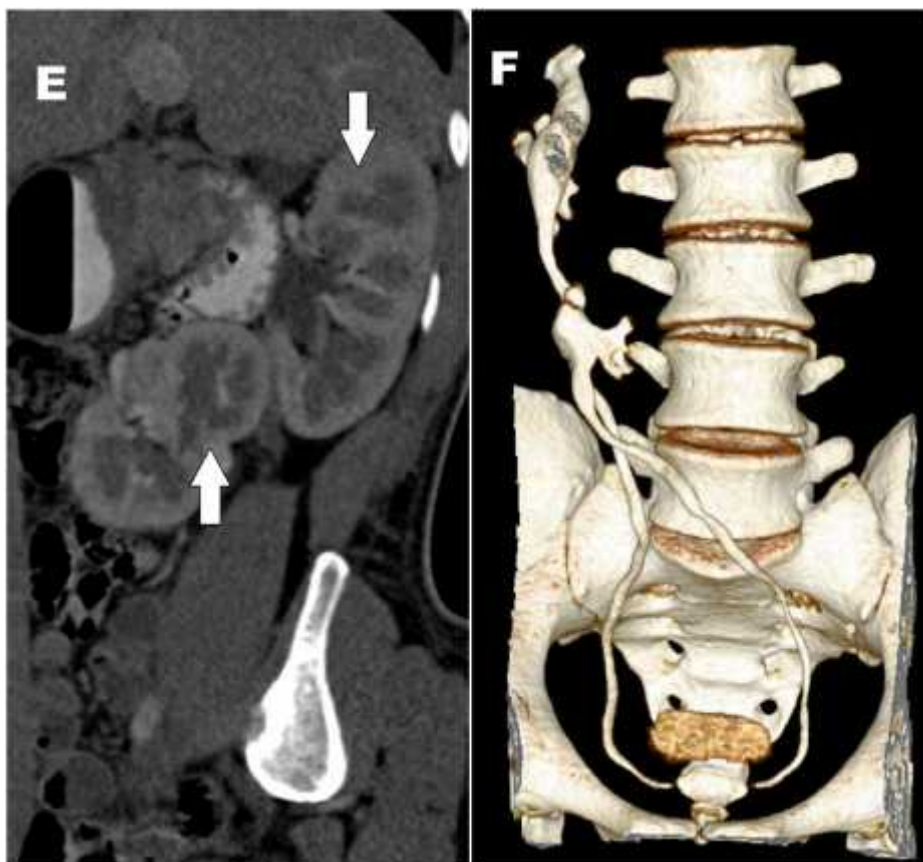
pallor with absence of icterus, pedal oedema and lymphadenopathy. Laboratory investigation reveals normal serum amylase, serum lipase and plasma glucose levels. Contrast enhanced computed tomography (CECT) abdomen was done in 128multidetector CT scan, Definition AS+ Excel (Siemen, Germany). CECT abdomen revealed irregular thickening of antro-pyloric part of stomach with partial gastric outlet obstruction and invasion of pancreatic head [Figure 1 A]. Few round perigastric lymph nodes ranging between 5 mm and 9 mm were demonstrated. The pancreatic head and neck were mildly bulky in size with non-visualized body and tail [Figure 1 A, B]. The left kidney was ectopic in location and found below the right kidney on right side of retro peritoneum [Figure 2 E, F]. No fusion was demonstrable between the kidneys and the collection system was draining normally in the trigone of urinary bladder [Figure 2 E, F]. Abdominal MRI and MRCP were carried by 1.5Tesla MR Scanner (Avanto, Siemens AG, Germany) which revealed similar findings along with non-visualised dorsal pancreatic duct [Figure 1 C, D]. The final diagnosis of dorsal pancreatic agenesis associated with crossed renal ectopia was made.

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**Figure 1** 30 year male patient evaluated for gastric adenocarcinoma. **A.** CECT Abdomen axial image demonstrated thickening of antropyloric part of stomach (Arrow) and bulky pancreatic head (Star).  
**B.** CECT Abdomen coronal image demonstrated bulky pancreatic head (Star) with absent body and tail.  
**C & D.** MRI Abdomen with MRCP coronal images revealed absent dorsal duct system and a short ventral duct (Triangle).



**Figure 2** 30 year male patient evaluated for gastric adenocarcinoma. **E.** CECT Abdomen sagittal image revealed crossed unfused kidneys (Arrows).  
**F.** VRT image demonstrated unfused pelvic/ureteral systems with normally draining ureters into urinary bladder.

## DISCUSSION

The pancreas is a retroperitoneal organ which develops as focal outpouchings from the foregut in 4<sup>th</sup> weeks of gestational life. The body and uncinata process develops from ventral pancreatic bud. The body and tail develops from dorsal bud (Mortele *et al*, 2006). At 7th week of gestational life, the ventral bud rotates around the duodenum to attain posterior relation to the dorsal bud. However there is constant relationship between dorsal bud and the bile duct. The ventral bud develops the posterior part of head and uncinata process of pancreas which drains by duct of Wirsung in to the duodenum at major papilla. The anterior pancreatic head, neck, body and tail are derived from embryological dorsal endodermal bud. All these parts drain through the duct of Santorini or minor duct into the duodenum by minor papilla (Mortele *et al*, 2006). The embryological developmental failure of dorsal foregut bud results variable degree of dorsal pancreatic agenesis which can be partial or complete. The complete dorsal agenesis lacks pancreatic neck, body and tail, the duct of Santorini and minor duodenal papilla. In partial subtype, there is presence of variable amount of pancreatic body, remnant of accessory duct and minor papilla (Lal *et al*, 2015).

Total pancreas agenesis is a lethal condition and extremely rare. However partial pancreatic agenesis has compatible life and results from variable amount of developmental failure in the ventral or dorsal pancreatic buds. (Turkvatan *et al*, 2013). Dorsal pancreatic agenesis is much more common than that of the ventral bud.

Dorsal pancreatic agenesis may be seen alone or as a component of heterotaxy syndromes. It is asymptomatic in most of the patient and seen incidentally in adulthood during radiological imaging done for other causes. There may be present with nonspecific abdominal pain or features of pancreatitis. There is increased risk of diabetes mellitus due to absence of islet cells.

Sometimes, pancreatic divisum and autodigestion secondary to chronic pancreatitis can mimics dorsal pancreatic agenesis and must be kept in mind in the differential diagnosis. In this case, the secondary atrophy of pancreatic parenchyma due to chronic pancreatitis is called pseudo-agenesis (Gold *et al*, 1993). The laboratory investigation reveals increased serum pancreatic enzymes. There may be history of recurrent abdominal pain and vomiting. CECT abdomen and MRI reveals variable amount of pancreatic parenchyma along with signs of chronic pancreatitis. However in dorsal bud agenesis, MRCP reveals complete absence of the dorsal duct and short ventral duct. In our reported case, there were no radiological signs of chronic pancreatitis. The body and tail of the pancreas was completely absent on CT and MR Imaging. MRCP revealed absent dorsal duct system and a short ventral duct (Mortele *et al*, 2006; Lal *et al*, 2015).

The upper urinary tract anomalies can be anomalies of ascent or the anomalies of form and fusion. The anomalies of ascent are due to failure of ascent resulting positional anomaly such as simple renal ectopia and crossed renal ectopia. The fusion anomalies can involves normally positioned kidneys e.g. horseshoe kidney or ectopic kidneys e.g. crossed renal ectopia. The kidneys are located in same side of retroperitoneum in crossed renal ectopia and may occur with fusion (85%) or without fusion (<10%) (Gutierrez *et al*, 2013). The fusion involves variable amount of renal tissue between kidneys, mostly in lower pole. Since all these anomalies are mostly asymptomatic, it is detected incidentally on routine imaging studies done for other causes. Sometimes, recurrent UTI or abdominal symptoms are associated secondary to urinary tract obstruction. The left-right ectopia is three times more common than right-left ectopia. Crossed renal ectopia is frequently seen in male (2:1) as urogenital developmental anomaly. In our report, the left-right renal ectopia without fusion was noted which is the most uncommon variety among the upper urinary tract anomalies (Gutierrez *et al*, 2013).

## CONCLUSION

The dorsal pancreatic agenesis in association with crossed unfused renal ectopia is an extremely rare congenital anomaly. The diagnosis of dorsal pancreatic agenesis must be kept in mind in absence of pancreatic body and tail. Crossed fused renal ectopia is diagnosed when both kidneys are noted on same side of retro peritoneum. Both the conditions are usually demonstrated at routine examinations or an incidental finding in the evaluation for different pathologies as in our case. CECT and MR imaging of abdomen along with high resolution MRCP sequences are extremely important to see the pancreatic morphology and ductal anatomy.

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