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Dorsal Agenesis of Pancreas Manifesting Clinically as Diabetic Ketoacidosis: A Rare Case Study

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Authors' contributions

This work was carried out in collaboration between all authors. Authors PKD, KAB and TP clinically examined the patient, reviewed and followed up on the clinical history, laboratory investigations and radiological investigations. Authors CRV and SKC wrote the working draft of the manuscript that was submitted, managed the literature searches, reviews, references and performed the comparative analysis with previously reported cases that were studied.

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

Case Study

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ABSTRACT

There have been reports of several developmental anomalies of the pancreas in the past two decades, and among these, one of the rare entities is dorsal agenesis of the pancreas [1]. Our report is on an 18-year-old male patient who presented with total agenesis of the dorsal pancreas. A computed tomography (CT) scan of the abdomen revealed a normal pancreatic head, but there was no observation of the body and tail of the pancreas. A magnetic resonance cholangiopancreatography (MRCP) confirmed the absence of the major pancreatic duct. This case was diagnosed as dorsal agenesis of the pancreas. As this is a very unusual case, this has been reported here as a case study due to its rare presentation.

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1. INTRODUCTION

Agenesis of the dorsal pancreas is an extremely rare anomaly which results from defective pancreas formation. A few case reports have been published in the literature about this anomaly. Agenesis of the dorsal pancreas is mostly asymptomatic but abdominal pain, pancreatitis and diabetes mellitus may be associated [2]. Partial agenesis of dorsal pancreas is observed as a short, rounded pancreatic head adjacent to the duodenum with absence of the pancreatic neck, body, and tail [3]. 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 [4]. In the case of complete agenesis of the dorsal pancreas, all the structures appear absent on imaging [5-8]. We present a case of dorsal pancreatic agenesis with clinical features that presented as diabetic ketoacidosis. While many patients with this anomaly are asymptomatic some patients may present with abdominal pain. Diabetes is unusually seen in dorsal agenesis, and may be attributed to the loss of the endocrine tissue due to the agenesis [9,10,11].

2. CASE REPORT

An 18 year old male patient presented with fever, chills abdominal pain and vomiting along with diabetic ketoacidosis; initial US did not visualize the pancreas, a CT was obtained and revealed an absence of the body & tail of pancreas. (Figs. 2, 3 and 4).The Uncinate process of pancreas was seen (Figs. 1 and 5). MRCP was done, and it showed an absence of pancreatic duct (Fig. 6). The patient was managed with symptomatic treatment, IV fluids, IV potassium supplementation, and insulin. The patient's clinical condition improved and was discharged. The patient is now on treatment with insulin for diabetes and is being followed up at regular intervals.

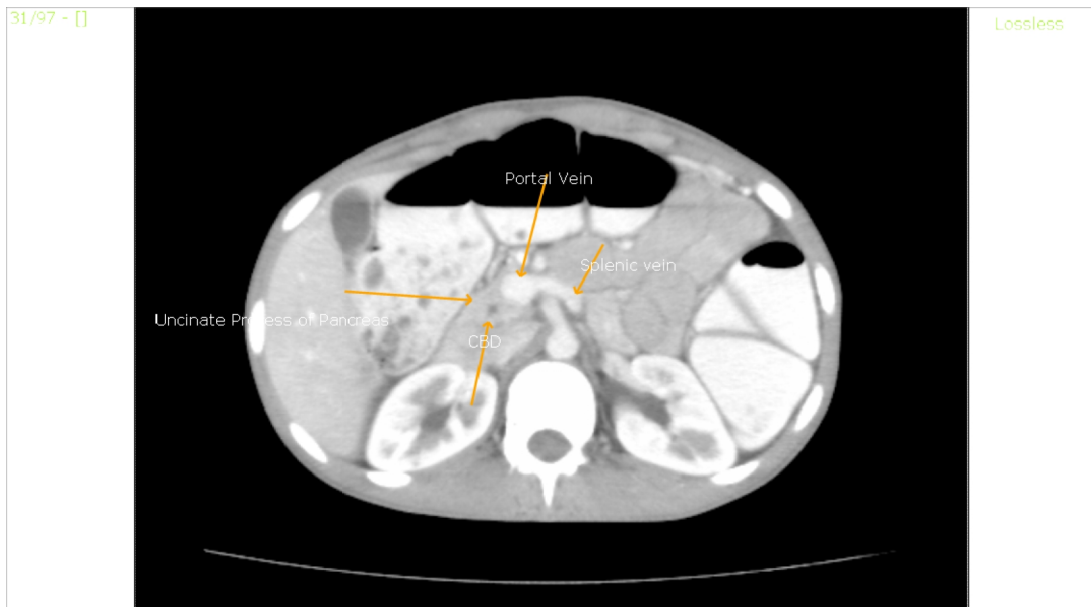


Fig. 1. Axial CT scan image of upper abdomen at level of portal confluence revealed uncinus process of head of pancreas with CBD. Anterior to splenic vein pancreatic body is not seen



Fig. 2. CT scan image at level of splenic vein showed absence of pancreatic body anterior to it



Fig. 3. Axial CT image at splenic hilum revealed absence of pancreatic tail



Fig. 4. Coronal reformatted image reveals absence of pancreatic tissue around the splenic vein

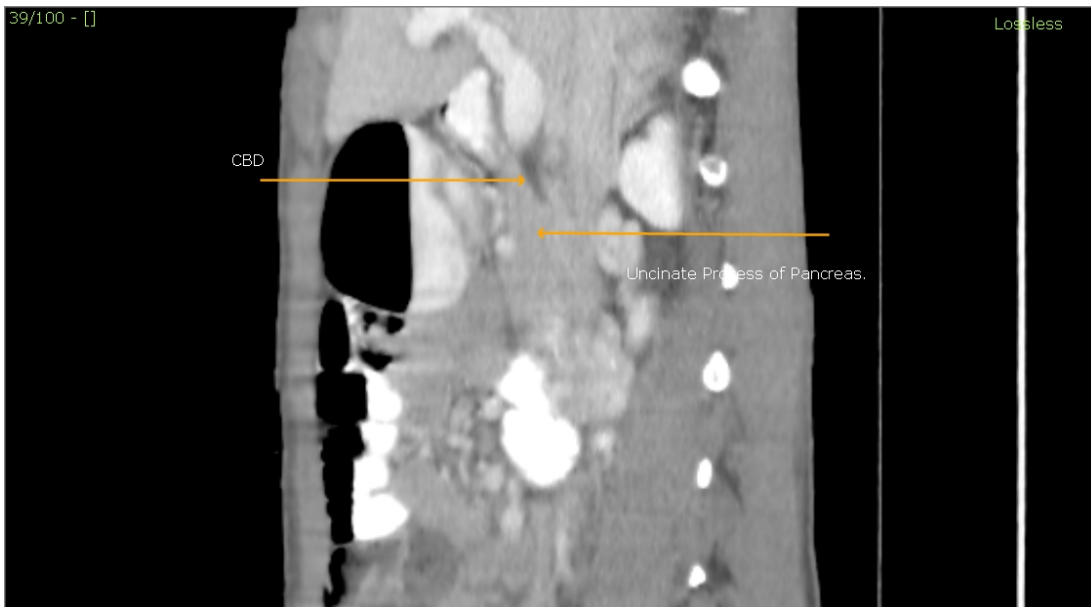


Fig. 5. Sagittal reformatted image revealed uncinete process and CBD

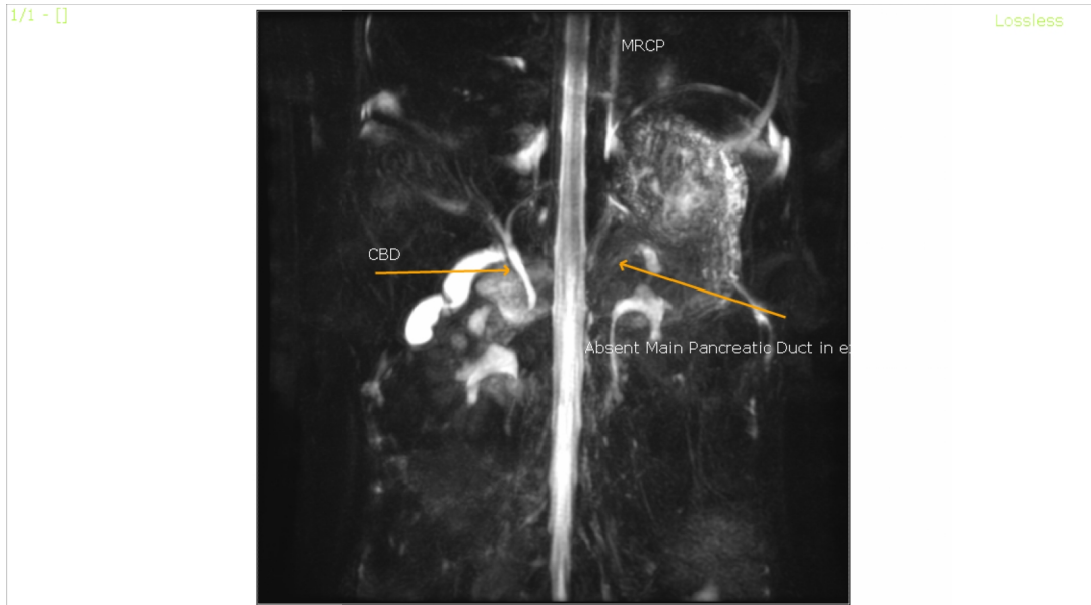


Fig. 6. MRCP image revealed absence of main pancreatic duct

3. DISCUSSION

The pancreas develops by ventral and dorsal endodermal buds. The dorsal bud forms the upper part of the head, body and tail of the pancreas which drains through the Santorini duct. The ventral bud gives rise to the major part of the head and uncinata process which drains through Wirsung duct [2].

Agenesis of the ventral pancreas and complete agenesis of the pancreas are lethal conditions [3].

Dorsal pancreatic agenesis is an extremely rare congenital anomaly. In the literature, about 23 cases of partial agenesis of dorsal pancreas were reported between 1913 and 2007. Pancreas divisum and autodigestion secondary to chronic pancreatitis must be considered in the differential diagnosis of the dorsal pancreatic agenesis. Failure of the ventral and dorsal pancreatic ducts to fuse is called pancreas divisum. In this entity, the ventral duct drains into major papilla, while the dorsal duct drains into separate minor papilla [2]. Atrophy of the body and the tail of the pancreas secondary to chronic pancreatitis and sparing of the pancreatic head is called pseudo-agenesis [4]. This situation may mimic dorsal pancreatic agenesis.

Abdominal CT may not evaluate the pancreatic duct in a detailed fashion. Therefore ERCP or MRCP is necessary for revealing the major and the accessory duct systems. ERCP is an invasive technique. It might challenge the cannulation of the minor papilla in selected cases. There is also radiation risk to the patient. MRCP can help the diagnosis of the dorsal pancreatic agenesis noninvasively with no radiation risk [12,5,6].

In our patient, distal pancreas was absent on CT and MRI images. On MRCP, dorsal duct system was not visualized and a short ventral duct was present which supports the diagnosis of dorsal pancreatic agenesis. In summary, this extremely rare congenital anomaly

must be kept in mind when the corpus and tail of the pancreas are not seen at routine examinations or as in our case an incidental finding at the examinations for different pathologies.

Few cases of agenesis of the dorsal pancreas have been reported in the English literature [7,8]. Most reports describe a single case presenting with diabetes mellitus, weight loss, pancreatitis, jaundice and duodenal obstruction [9,13].

Agenesis of the dorsal pancreas has been most frequently identified from imaging studies during investigation of abdominal pain. The abdominal pain has been assumed to be due to pancreatitis, duodenal obstruction, autonomic neuropathy, or sphincter of Oddi dysfunction [9,10,13,14,15]. Abdominal pain in agenesis of the dorsal pancreas may also be due to hypertrophy of the remnant ventral gland with higher intra-pancreatic duct pressures. Most of them also have diabetes mellitus probably because most of the islet cells are located in the pancreatic body and tail. Absence of dorsal pancreas may co-exist together with polysplenia syndrome and may also cause acute pancreatitis and chronic pancreatitis [1].

Familial occurrence of agenesis of dorsal pancreas in the mother and her sons [11,15] has also been reported in a rare situation. In these reports, the authors suggested that the genetic mode of transmission for this anomaly is most likely autosomal dominant or X-linked dominant.

Agenesis of the dorsal pancreas is usually suggested on abdominal ultrasonogram (US), CT, or MRI when the body and tail of pancreas are not visualized ventral to the splenic vein [16]. When agenesis of the dorsal pancreas is suggested by imaging studies, diagnostic possibilities to exclude fat replacement of the pancreas and atrophy following pancreatitis should be considered. In fat replacement of pancreas, the entire gland is usually involved and the pancreatic duct is present, whereas with atrophy following pancreatitis a relevant clinical history usually exists [17,18,19].

Abdominal CT may not evaluate the pancreatic duct in a detailed fashion. Therefore ERCP or MRCP is necessary for revealing the major and the accessory duct systems. ERCP is an invasive technique. There is also risk of radiation to the patient. MRCP is can help the diagnosis of dorsal pancreatic agenesis noninvasively with no radiation risk. This extremely rare congenital anomaly must be kept in mind when body and tail of pancreas are not seen on routine examinations.

4. CONCLUSION

To conclude, dorsal agenesis must be considered as a differential diagnosis in situations where the corpus and tail of the pancreas are absent either during routine examinations or as in our case as a coincidental finding when following up for diabetes and its complications like ketoacidosis. If agenesis of the dorsal pancreas is suspected, the combined use of CT and ERCP or MRCP is needed for confirmation of the diagnosis.

Recent advances in Computerised Tomography, added with the ability to observe pancreas in the sagittal and coronal sections have added another advantage in detecting congenital lesions such as in our case. Though partially interventional procedures like endoscopic retrograde choledochopancreatography (ERCP) was usually done in the past to investigate diseases of the pancreas, these cases are now detected by noninvasive methods.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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