

Abdominal pain, diabetes mellitus and an absent piece of pancreas

Abstract

Pancreatic ductal anomalies secondary to developmental failure of ventral or dorsal pancreatic buds have been reported. Dorsal pancreatic agenesis is an uncommon congenital pancreatic ductal anomaly leading to recurrent acute pancreatitis. We report a middle aged diabetic gentleman, with recurrent acute pancreatitis, due to an absent piece of pancreas. The diagnosis of dorsal pancreatic agenesis was made on magnetic resonance imaging during the aetiological work up.

Keywords: pancreas, ductal anomalies, recurrent pancreatitis, congenital anomalies, HPB surgery, pancreatic agenesis

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Vidhyachandra Gandhi,¹ Pratik Gautam,² Nitin Pai,³ Sujai Hegde⁴

¹Consultant, GI & HPB Surgery, India

²Resident, General Surgery, India

³Consultant, Medical Gastroenterology, India

⁴Consultant, Surgical oncology, India

Correspondence: Vidhyachandra Gandhi, Department of GI & HPB Surgery, Ruby Hall Clinic, India, Tel +91 99878 84252, Email drgandhiv@gmail.com

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Abbreviations: U/L, Units per Litre; IgG₄, Immunoglobulin G₄; MRCP, Magnetic Resonance Cholangiopancreatography; CT, Computed Tomography

Introduction

Agenesis of dorsal pancreas is an uncommon congenital anomaly due to defective development. Since its description in 1911, till date approximately 100 cases have been reported in literature.¹ We share our experience with a gentleman who had abdominal pain, diabetes and was found to have an absent piece of pancreas on imaging.

Case Report

A 55 years non-alcoholic gentleman was admitted through emergency medical services with sudden severe upper abdominal pain in the epigastrium with radiation to back. The pain in the upper abdomen was associated with recurrent bouts of bilious vomiting. He denied history for any other gastrointestinal symptoms. He is a known diabetic since last 3 years requiring insulin and oral hypoglycaemic agents. He has had similar episode of pain six months ago which was managed conservatively elsewhere. He was clinically stable with tenderness in the epigastrium.

All his laboratory parameters were normal except for raised amylase (650U/L) and lipase (1200U/L). Ultrasound abdomen revealed a bulky oedematous pancreas with minimal free fluid. There were no gall stones and common bile duct was normal. A working diagnosis of mild acute pancreatitis was made as there was no evidence of organ failure. He was managed in the intensive care unit for 48hrs with IV fluids, enteral nutrition and later shifted into ward. His lipid profile, serum calcium, serum parathyroid hormone levels and serum IgG₄ levels were normal. He was advised magnetic resonance cholangiopancreatography (MRCP) for etiological work-up which he denied, only to report again after three months with similar symptoms.

Computed tomography (CT) scan of abdomen showed oedematous pancreatic head (Figure 1A), with absence of pancreatic body and tail

(Figure 1B). MRCP was done which showed pancreatic duct only in the head region with its non visualisation in the body and tail (Figure 2). These features were suggestive of dorsal pancreatic agenesis.

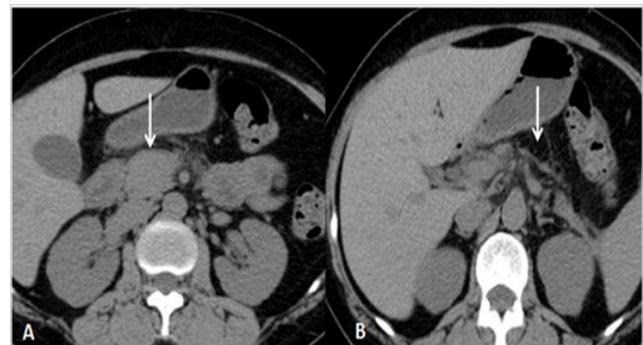


Figure 1 CT scan abdomen image showing oedematous pancreatic head (Figure 1A), with absence of pancreatic body and tail (Figure 1B).



Figure 2 MRCP image showing short ventral duct in the pancreatic head region (short arrow) and absence of duct in the body/tail region (long arrow).

Treatment

The patient was placed on dietary modifications.

Outcome and follow-up

His sugars are in control with insulin and oral hypoglycaemic agents.

Discussion

Congenital ductal anomalies of pancreas are being reported more frequently due to better availability of imaging modalities. Although complete and ventral pancreatic agenesis are very rare and incompatible with life,² few cases of dorsal pancreatic agenesis have been reported.

During embryogenesis the pancreas develops from ventral and dorsal buds on opposite side of foregut. Ventral bud is responsible for pancreatic head/uncinate development, while dorsal bud makes the body and tail. The head and uncinata process drains through the duct of Wirsung, while the body/tail through the duct of Santorini. Failure of development or regression of the dorsal bud is responsible for dorsal pancreatic agenesis.³

Approximately fifty percent of these patients are asymptomatic or diagnosed incidentally. In symptomatic patients the commonest presentation is recurrent abdominal pain secondary to recurrent acute pancreatitis. There are several reasons claimed for the occurrence of recurrent acute pancreatitis. Dysfunction of Sphincter of Oddi, hypertrophy of ventral pancreas and raised pancreatic intraductal pressure are believed to be responsible for acute pancreatitis.⁴ Diabetes occurs due to lack of islet cells which are dominant in body and tail of pancreas. Pancreatic exocrine insufficiency has also been reported in patients with dorsal pancreatic agenesis.⁵

The reported anomalies with agenesis of dorsal pancreas are heterotaxy, polysplenia syndrome, ectopic spleen, malrotation of gut, coarctation of the aorta, tetralogy of Fallot, atrioventricular valvular abnormalities, and total anomalous pulmonary venous connection.⁶ Pancreatic cancer,⁷ neuroendocrine tumour,⁸ serous cystadenoma⁹ and solid pseudopapillary tumour¹⁰ have been reported in patients with dorsal ductal agenesis.

This condition needs to be differentiated from pancreatic divisum, pancreatic lipomatosis and pseudoagenesis. Chronic pancreatitis can lead to complete atrophy of body and tail (pseudoagenesis) mimicking dorsal agenesis.¹¹ Individuals with chronic pancreatitis often are chronic alcoholics/smokers with antecedent history of recurrent abdominal pain with typical findings on imaging. Although our patient had recurrent acute pancreatitis he denied history for alcohol or smoking and also CT failed show any features of chronic pancreatitis. Distal pancreatic lipomatosis is characterised by significant amount of fat in the body/tail region which appears hypodense on the CT scan mimicking dorsal agenesis. Absence of dependant stomach and small bowel sign would help to differentiate this condition from dorsal agenesis.¹²

MRCP and CT scan are useful tools in diagnosing agenesis of dorsal pancreas. Ultrasound has limitations due to its operator dependence and overlying bowel gas. CT scan shows absence of pancreatic body and tail. The pancreatic space is occupied by the stomach or small bowel termed as dependant stomach or small bowel

sign.¹² MRCP is a non invasive accurate diagnostic test revealing absence of ductal system in the body/tail region. A complete absence of dorsal pancreatic duct with presence of short ventral pancreatic duct is essential for diagnosis of dorsal duct agenesis, as was seen in our patient. In our patient CT scan and MRCP helped to clinch the diagnosis. EUS is also described as a modality to diagnose dorsal agenesis,¹³ but is expensive, invasive and operator dependant.

Treatment of this condition is managing the episodes of pain with medications. The pancreatitis is usually mild in nature and there are no reports of these patients requiring necrosectomy for remnant pancreas. Our patient did require repeated admissions with symptoms being controlled with medications. Dietary modifications with low fat diet have decreased his episodes of pain.

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Conflict of interest

There is no conflict of interest.

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