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Complete Agenesis of the Dorsal Pancreas: A Clinical Case

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Abstract: **Case Report**

Complete agenesis of the dorsal pancreas (ADP) is an exceptionally rare congenital malformation, resulting from the failure of the dorsal pancreatic bud to develop during embryogenesis. This anomaly leads to the absence of both the pancreatic body and tail. We describe the case of a patient presenting with atypical manifestations, including long-standing abdominal pain and recurrent episodes of acute pancreatitis beginning in childhood. Diagnosis was established through abdominal computed tomography (CT) imaging, which confirmed the complete absence of the dorsal pancreas. This observation adds to the limited number of cases—slightly more than one hundred—reported worldwide in the medical literature.

Keywords: Dorsal pancreatic agenesis, Abdominal pain, Acute pancreatitis.

I. INTRODUCTION

The pancreas originates from the embryological fusion of two distinct buds: the ventral bud, which forms the lower head and uncinate process, and the dorsal bud, which gives rise to the upper head, body, and tail of the gland. Complete Dorsal Pancreatic Agenesis (DPA) is a rare congenital anomaly defined by the failure of the dorsal bud to develop. Although frequently clinically silent, it can present with abdominal pain and recurrent bouts of acute or chronic pancreatitis. We report an additional case of DPA, diagnosed in a 41-year-old woman with a protracted history of abdominal pain and repeated acute pancreatitis episodes.

II. OBSERVATION

We report the case of a 41-year-old female patient with a history of chronic abdominal pain that had been unresponsive to treatment since childhood. Her medical history includes a cholecystectomy five years prior and recurrent episodes of acute pancreatitis. The patient presented with epigastric pain of a pancreatic nature. Laboratory tests showed hyperlipasaemia (12 times the normal level), with normal lipid and calcium levels. The abdominal ultrasound was unremarkable. An abdominal computed tomography (CT) scan visualised a normal-looking pancreatic head but confirmed the complete absence of the body and tail of the pancreas (Fig 1).

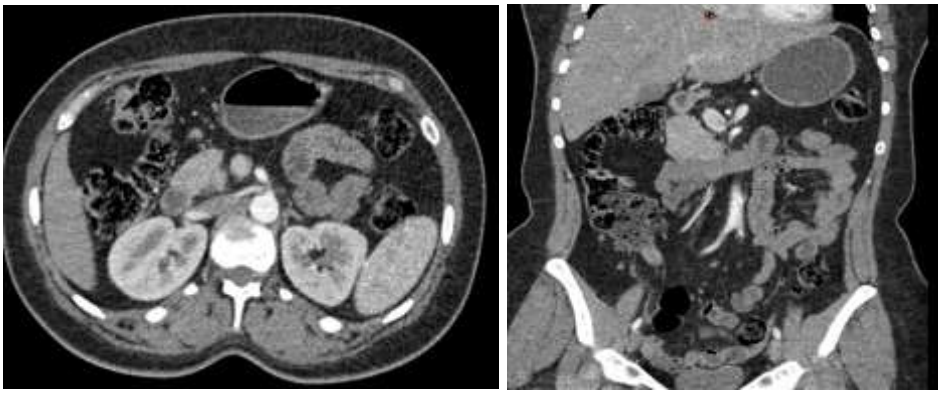

Fig 1: **Abdominal computed tomography (CT) scan reveals a normal-appearing pancreatic head and the complete absence of the body and tail**

III. DISCUSSION

Dorsal Pancreatic Agenesis (DPA) is an exceptionally rare congenital malformation, with little more than a hundred cases documented globally since its first report in 1911. Its noted incidence is increasing, a trend attributed primarily to the greater use of sophisticated cross-sectional imaging techniques. DPA is classified as either partial (absence of the tail) or complete (absence of the body and tail), with the latter being the focus of most studies. Notably, DPA is compatible with life, unlike other complete pancreatic agenesis variants. While the precise aetiopathogenesis remains unclear, a mutation in the HNF1B gene has been implicated in some cases, often linking DPA to broader genetic syndromes. Patients with DPA rarely present with pathognomonic symptoms; the diagnosis is frequently made incidentally during imaging investigations for non-specific abdominal pain. However, DPA can be clinically significant, often complicated by acute or chronic pancreatitis. Proposed mechanisms for this complication include sphincter of Oddi dysfunction, compensatory hypertrophy of the remaining ventral pancreatic head, or ductal hypertension. Accurate diagnosis is crucial, as DPA must be differentiated from pseudo-agenesis, which is secondary pancreatic atrophy caused by chronic pancreatitis that can mimic the congenital malformation. The list of differential diagnoses is extensive and also includes pancreatic head carcinoma, pancreas divisum, pancreatic pseudolipodystrophy, and distal pancreatic lipomatosis. It is therefore paramount to secure a thorough medical history and employ a multi-modality imaging approach, including ultrasonography, Computed Tomography (CT), and high-resolution techniques such as Magnetic Resonance Pancreatography (MRCP) and Endoscopic Ultrasound (EUS), to definitively exclude these differential conditions and confirm the characteristic absence of the dorsal pancreatic duct.

IV. CONCLUSION

Dorsal Pancreatic Agenesis (DPA) is a rare congenital anomaly resulting from a defect in embryonic organogenesis, presenting in either a partial or complete form. The observed incidence is increasing, particularly owing to enhanced imaging capabilities. Although often an incidental finding, DPA is clinically significant due to its strong association with endocrine and exocrine complications, notably hyperglycaemia/diabetes mellitus (affecting up to 50% of cases) and recurrent pancreatitis. Magnetic Resonance Cholangiopancreatography (MRCP) has emerged as the gold-standard imaging modality for definitively confirming the characteristic absence of the dorsal ductal system, complementing the CT scan, which remains the preferred tool for the initial visualisation of parenchymal absence. Early recognition of DPA is essential for the proactive management of associated long-term risks.

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