Case Report

A missing piece of pancreas: A rare case report of dorsal pancreatic agenesis presenting as acute pancreatitis

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ABSTRACT

There are various congenital anomalies of the pancreas, among which dorsal pancreatic agenesis is a very rare entity <120 cases reported till now. It is due to absent dorsal bud derivatives. It can be asymptomatic or can present as abdominal pain, pancreatitis, and hyperglycemia. A 42-year-old gentleman presented with sudden onset abdominal pain and, on evaluation, was diagnosed to have acute pancreatitis. Incidentally noted to have dorsal pancreatic agenesis, a congenital anomaly which presented as acute pancreatitis. He was managed with hydration, multimodal analgesia, and nutrition. Complications associated with it must be kept in mind, such as pancreatitis and diabetes/hyperglycemia secondary to insulin deficiency. Early diagnosis and prompt treatment of complications of dorsal pancreatic agenesis is the key.

Key words: Acute pancreatitis, Congenital anomaly, Dorsal pancreatic agenesis

Dorsal pancreatic agenesis is a rare congenital anomaly due to the absence of dorsal bud derivatives. It is an incidental finding in many of the cases during general evaluation or during autopsy. But in symptomatic cases where common presentations are abdominal pain, pancreatitis, and hyperglycemia, it is identified by various imaging modalities done as a part of evaluation. It can be managed symptomatically based on the presentation, but few cases have been reported where it is associated with other organ anomalies and pancreatic adenocarcinomas [1]. The rationale behind reporting this case is due to its rarity and only <120 cases have been reported till now.

CASE

A 42-year-old gentleman, athlete by profession, with a history of Lichen planus (Fig. 1) on the legs for 5 years on treatment, presented to us with sudden onset abdominal pain radiating to the lower chest and upper back for the past few hours. On arrival to the emergency room (ER), he was saturating 98% on room air, hemodynamically stable (Blood pressure: 142/88 mmHg) with sinus bradycardia (50–55 beats/min) but had severe abdominal pain scoring 8/10 on Visual Analog Score. Pain was managed by multiple intravenous (IV) medications (IV Paracetamol 1 g, IV Tramadol 50 mg, IV Hyoscine 20 mg, IV Diclofenac 75 mg, and IV Fentanyl 25 mcg) in the ER. He underwent contrast-enhanced computed tomography (CECT) of the abdomen and pelvis and shifted to the medical intensive care unit (MICU) for pain management.

In MICU, detailed evaluation for pain abdomen was done, which revealed elevated serum amylase (1163 U/L) and serum lipase (5595 U/L) enzymes, CECT abdomen and pelvis showed bulky head and uncinate process of pancreas with minimal peripancreatic fat stranding and deficiency of distal body and tail of pancreas, possibly an anatomical defect (Fig. 2). Other laboratory investigations showed hematocrit 41.9%, total white blood cell count 10,940/µL, serum creatinine 1.21 mg/dL, C-reactive protein 2.5 mg/L, and normal liver function. He was diagnosed to have mild acute pancreatitis. Other causes of pancreatitis, including autoimmune cause in view of lichen planus, were thoroughly evaluated, and all results were negative (Table 1).

He was managed with Intravenous and oral hydration, multimodal analgesia (Paracetamol 4 g/day, IV Fentanyl infusion followed by transdermal patch), and nutrition. He was symptomatically better and was shifted out from intensive care unit after 48 h. Subsequently, he was discharged from the hospital in the next 3 days.

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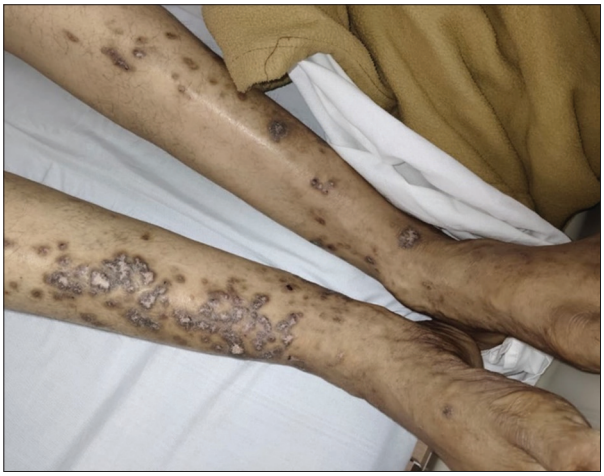

Figure 1: Lichen planus on lower limbs

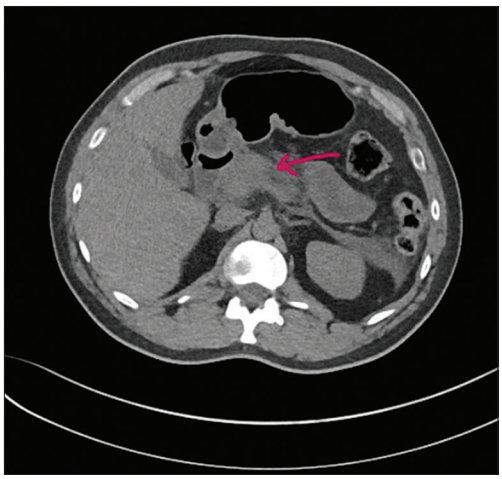

Figure 2: Computed tomography scan showing pancreas: Bulky head and uncinate process (red arrow), absent distal body and tail (dorsal pancreatic agenesis)

Table 1: Detailed work-up for acute pancreatitis in our patient

Possible etiology	Result	Correlation
History of ethanol consumption	NO	NO
Gall stones	CECT: No gall stones Ultrasound: No gall stones	NO
Liver function test	Normal	NO
Triglycerides	73 mg/dL (normal)	NO
Serum Calcium	8.5 mg/dL (normal)	NO
Autoimmune	IgG4 levels: 0.692 g/L (normal) ANA-IF: Negative	NO NO
Anatomical anomaly	High probability (deficiency of the distal body and tail of the pancreas)	

CECT: Contrast-enhanced computed tomography, ANA-IF: Antinuclear antibody immunofluorescence assay, IgG4: Immunoglobulin G 4

DISCUSSION

The pancreas develops from the large ventral bud and small dorsal bud during the fifth embryonic week. The complete pancreas is formed by the fusion of both the buds during foregut rotation. Any deviation from this process will lead to various congenital anomalies of the pancreas. An extremely rare anomaly is dorsal pancreatic agenesis with absent dorsal bud derivatives (head, body and tail) [2]. The first case of dorsal pancreatic agenesis was reported in 1911 during an autopsy [3]. There are only <120 cases of dorsal pancreatic agenesis reported till now.

Studies conducted in mice showed that a mutation in retinaldehyde dehydrogenase 2 (Raldh2) and gene H1xb9 or a deficiency of retinoic acid caused dorsal pancreatic agenesis [4], but the association of any genetic factor in humans is still unknown. It is associated with other organ anomalies as well, such as biliary atresia, polycystic kidney disease, and splenic deformities. There are a few reports of association with adenocarcinoma of the pancreas, but the exact pathophysiology is unknown [1].

It can have various clinical presentations, including abdominal pain, pancreatitis, hyperglycemia, and insulin-dependent diabetes [5,6]. In some cases, identification of the cause of the above presentations is challenging unless dorsal pancreatic agenesis is identified with various imaging modalities. Imaging modalities include ultrasound, computed tomography (CT), magnetic resonance cholangiopancreatography (MRCP) and endoscopic retrograde cholangiopancreatography (ERCP) [7]. Differential diagnosis for dorsal pancreatic agenesis is pancreas divisum, pancreatic atrophy, pancreatic tumor, or pancreatic lipomatosis. Each one can be differentiated from each other various imaging modalities mentioned here.

Management is always symptomatic based on the presentation. Hyperglycemia and diabetes mellitus are secondary to insulin deficiency as the majority of insulin-producing beta-cells are in the body and tail of the pancreas. Management includes insulin therapy, pancreatic enzymes supplementation, and diet modification [8].

In our case, the patient presented as acute pancreatitis, and during evaluation, CT showed an incidental finding of dorsal pancreatic agenesis. On literature search, we could not find any direct association between Lichen planus and pancreatitis due to dorsal agenesis. Our patient has a risk of developing insulin-dependent diabetes in his future. He was educated about the requirement of further evaluation and possible complications in the future. Many cases of dorsal pancreatic agenesis go unnoticed because not all cases are symptomatic.

CONCLUSION

Dorsal pancreatic agenesis is a rare congenital anomaly of the pancreas with <120 cases reported till now. It can present as various forms. Many cases are asymptomatic and the anomaly is incidentally noted. Some cases present as abdominal pain, pancreatitis, or hyperglycemia, where various imaging modalities, including ultrasound, CT, MRCP, and ERCP helps to identify this anomaly. The genetic factor associated with dorsal pancreatic agenesis

is still a mystery in human beings and further studies are necessary to ascertain the exact pathogenesis.

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