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Ritish Gupta  
PGIMS, dritishgupta@gmail.com

Ashima Mittal  
Department of Radiodiagnosis, Star Imaging and Diagnostic Centre, Delhi, India

Bhavna Arora  
Department of Radiology, PGIMS, Rohtak, Haryana

Riya Aggarwal  
Department of Radiology, PGIMS, Rohtak, Haryana

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CASE REPORT

Complete dorsal pancreatic agenesis: A case report

Ritish Gupta a,*, Ashima Mittal c, Bhavna Arora b, Riya Aggarwal a

a Department of Radiodiagnosis, PGIMS, Rohtak, Haryana, India
b Department of Radiology, PGIMS, Rohtak, Haryana, India
c Department of Radiodiagnosis, Star Imaging and Diagnostic Centre, Delhi, India

Abstract

Complete agenesis of the dorsal pancreas (ADP), an extremely rare congenital anomaly, is caused as ‘the dorsal pancreatic bud fails in developing the body and tail of the pancreas at the time of embryological development.’ There are nearly 100 cases of dorsal pancreatic agenesis till date. Most of the individuals with ADP are without symptoms; however, few may present with clinical manifestations like pancreatitis, epigastric pain, hyperglycemia, and diabetes mellitus. This is a case report of a 19-year-old female with ADP, who was diagnosed incidentally while undergoing radiological assessment for abdominal pain in left hypochondrium. As complete ADP is very rarely found, clinical knowledge related to the ADP would be helpful in the differential diagnosis and taking decision regarding the management.

Keywords: Agenesis, Computed tomography, Dorsal pancreas, Endoscopic retrograde cholangiopancreatography

1. Introduction

Dorsal pancreatic agenesis is a congenital embryological development defect [1–3]. It was first noticed in 1911 and since then a total of 100 cases have been reported [4]. It can present in two forms—partial or complete agenesis of dorsal pancreas (ADP). Both partial and complete forms can have other accompanying malformations [5–7].

The etiopathogenesis is linked to familial nature with certain gene-signaling pathways (hedhehog and retinoic acid) [8]. The patients with ADP are generally asymptomatic. But, in some symptomatic cases, the chief complaint is the epigastric pain, which is localized to the epigastric region and aggravates after meals [9].

The disease can be screened radiologically by ultrasonography (USG) and computed tomography (CT). Then the patients may be confirmed by MRI or MR cholangiopancreatography (MRCP) [3].

We present here a rare case of complete ADP that was detected incidentally during a radiological screening for nonspecific abdominal pain in a 19-year-old female. This case shall elaborate on the literature about ADP and its management.

2. Case report

A 19-year-old female presented with a complaint of abdominal pain in the left hypochondrium for the past 5 days. Her height was 4 feet 11 inches. She had no history of trauma, nausea or vomiting, intake of drug or alcohol. She had no other complaints. There was past history of appendicectomy 8 years back due to a similar complaint. There was no family history of recurrent infection, stillbirths, or early infant mortality. On physical examination, the patient had tenderness above the right iliac fossa.

Laboratory investigations were performed, which demonstrated random blood sugar in normal range (81 mg/dl, reference value 80–140 mg/dl), serum amylase in normal range (83 U/l, reference value: 0–140 U/l), and normal levels of serum pancreatic lipase (42 U/l, reference value 0–160 U/l), with the findings being not in accordance with diabetes mellitus or pancreatitis.

On USG, the pancreatic head was normal in size, contour, and echotexture; there were no presence of dilatation of duct or parenchymal calcification. Moreover, there was no visualization of pancreas’s body and tail, anterior to the splenic vein and portal confluence.
The abdominal CT and MRI were done, which showed that pancreas was partially visualized. There was visualization of the head and the uncinate process of the pancreas, with the margins being ill-defined. However, the distal neck, body, and tail of the pancreas were not visible.

The MRCP was performed for confirming the diagnosis. We could not use thin slab techniques on the patients because she could not hold the breath for long duration. Instead, thick-slab heavily T2-weighted sequences were used.

The MRCP revealed that ‘the dorsal pancreatic duct of Santorini’ and ‘the minor duodenal papilla’ were not visualized. There was normal and clear visualization of the common bile duct as well as ‘the ventral pancreatic duct of Wirsung,’ which was consistent with characteristics of complete ADP and thus confirmed the diagnosis (Figs. 1–4). Thus, there was no requirement for performing the ERCP. The diet with a low-fat diet was advised to the patients, along with frequent monitoring of blood sugar because the patient was asymptomatic.

3. Discussion

The significance of the present case stems from the rarity of it. A rare disease with a common presentation of epigastric pain necessitates reporting of such cases to enmass our data on the diagnosis and management of such patients.

The present case of a female patient presented at the age of 19 years although it is a congenital condition. Among other case reports, patients of higher age were found. Robert et al. [3] reported that a 34-year-old male had ADP. Yang et al. [10] reported that a 30-year-old man had ADP. Moreira et al. [11] also found ADP in a 34-year-old man. Mei et al. [12] reported that ADP was found in two women: one was 65 years old and the other was 61 years old. Jain et al. [13] conducted a case study on a 37-year-old woman with ADP.

The clinical presentation in our study was of abdominal pain in the left hypochondrium which was not ascribed to any cause. Among other cases, Moreira et al. [11] reported that the patient was asymptomatic and referred for nephrology and gastroenterology appointment due to microalbuminuria and cholestasis. Mei et al. [12] reported that one woman had abdominal pain with nausea, bloating, and acid reflux, and another woman had abdominal pain with fever, vomiting, and bloating. In a case report by Jain et al. [13], woman presented with complaints of recurrent upper abdominal pain, dizziness, fatigue, and pain in lower extremities for past few months.

Fig. 1. Ultrasound image showing the pancreatic head (marked with calipers), which was enlarged in size with normal echotexture. However, the pancreatic body and tail were not visualized anterior to the splenic vein and portal confluence.

Fig. 2. Axial abdominal computed tomography image in portal venous phase showing visualization of the head and uncinate process of the pancreas (white arrow). However, distal neck, body, and tail of the pancreas were not visible.

Fig. 3. Axial abdominal T2W MR image showing visualization of the head and uncinate process of the pancreas (white arrow) with non-visualization of rest of the pancreas.
While monitoring such cases for general presentations, certain associations must be evaluated. In ~50% of the affected cases, diabetes mellitus may be present. In cases of partial agenesis, abdominal pain is generally found, whereas, diabetes mellitus is frequently present among patients with complete agenesis. The development of recurrent or intermittent pancreatitis occurs in most of the patients with ADP in the long duration. The steatorrhea as well as signs of pancreatic exocrine failure is also reported in some cases [14]. In our case, sugar levels were normal and there were no signs of pancreatitis.

In similarity with the report by Robert et al. [3], the patient had normal glucose and normal serum amylase levels but slightly increased serum pancreatic lipase levels, which was consistent with ADP. The correct diagnosis of ADP among all these differentials has become possible because of the recent advancements in modern imaging modalities. The main imaging modality is USG, but due to the organ screen and the overlying bowel gas shadows, USG findings in some individuals appear to be suspicious. As the blood flow to the viscera can be viewed, three-dimensional (3D) reconstruction CT is a better approach for the diagnosis of ADP. When the dorsal pancreas is absent, stomach or intestine fills the distal pancreatic bed, this abuts the splenic vein [3].

Because MRCP is a noninvasive technique, it is considered as an imaging procedure for confirming the diagnosis of ADP. Improvement in diagnostic accuracy of MRCP can be done by 3D reconstruction or dynamically with secretin injection. ERCP has superior spatial resolution and thus helpful for a detailed description of pancreas. Moreover, it is taken into account as the gold standard investigation. It is, however, an invasive technique, and exposes to radiation. It is associated with the risk of morbidity because of pancreatitis caused due to catheterization of the minor duodenal papilla. The S-MRCP (with secretin injection) improves diagnostic accuracy, without complications, and eliminates the risks associated with the ERCP [10].

Another comparatively novel minimally invasive imaging technique is EUS that helps in directly visualizing the total pancreatic parenchyma as well as the pancreatic ductal system. In addition, it helps in performing fine needle aspiration cytology, and is as good a technique as ERCP. In the present case study, diagnosis of the ADP was done at the time of assessment of the patient for abdominal pain in the left hypochondrium. The patient was diagnosed with complete dorsal pancreatic agenesis. The management was done conservatively; the analgesics and antibiotics were prescribed. She was without symptoms in 15 days.

Management of patients with ADP depends on the symptoms of patients and radiological findings. Yang et al. [10] reported that as the patient presented with abdominal pain and diabetic ketoacidosis, he was prescribed combination therapy including insulin, pancreatic enzyme supplements, and mosapride citrate and gradually, there was resolution of pain. In the case report by Robert et al. [3], the patient was asymptomatic and thus for savoring the pancreas, a low-fat diet and regular blood sugar monitoring was advised to him. In another case report, Mei et al. [12] reported that in woman with gastrointestinal symptoms and no clear positive signs during the physical examination, pancreatin enteric-coated capsules were advised, which subsequently improved symptoms. In a case report by Jain et al. [13], the patient was advised dietary modification and treatment of diabetes mellitus. Symptoms of patient improved after treatment.
One of the limitations was that genetic analysis was not performed in the presented case. Tests for the diagnosis of pancreatic exocrine insufficiency were not conducted, thus, data was incomplete in this regard.

3.1. Conclusion

In summary, a case of ADP presents with nonspecific abdominal symptoms. Imaging is helpful for the diagnosis. Simple ADP does not require special treatment and can be managed conservatively; however, complicated cases may need insulin therapy and surgery. As complete ADP is a very rare congenital anomaly, this case report adds to the already existing literature and act as a stepping stone for understanding the disease process and management of the patient.

Conflicts of interest

There are no conflicts of interest.

References