Incidentally Detected Agenesis of Dorsal Pancreas on PET/CT: Case Report and Review of Literature

Rasika Kabnurkar, M L Rokade[1], Kalashree Bandekar[1], Nikhil Kamat[1]

Department of Nuclear Medicine, [1]Department of Radiology, Jupiter Hospital, Thane, India

Abstract

Agenesis of Dorsal Pancreas (ADP) is a rare congenital anomaly characterized by the absence of body and tail of pancreas. We report a case of incidentally detected ADP on Contrast Enhanced Computed Tomography (CECT) component of Fluorodeoxyglucose Positron Emission Tomography (FDG PET/CT) in a treated case of carcinoma (Ca) tongue with suspected local recurrence. Dependent Intestine Sign, hallmark of ADP on CECT imaging was noted in our patient.

Key words: *Agenesis dorsal pancreas, dependent intestine sign, FDG PET/CT*

Introduction

The Agenesis of Dorsal Pancreas, also known as Congenital Short Pancreas, is a rare congenital anomaly characterized by the absence of body and tail of pancreas, resulting from failure of development of the dorsal pancreatic bud. ADP was first described in 1911 in an autopsy study and was associated with diabetes mellitus.[1] Till now less than 100 cases of ADP have been reported in literature.[2] We report a case of incidentally detected ADP on Contrast Enhanced Computed Tomography (CECT) component of FDG PET/CT in a non-diabetic patient.

Case Report

A 49 year old male, non-diabetic, case of carcinoma of tongue was treated with radiation therapy and concurrent chemotherapy till Dec 2015. After 4 months, the patient presented with suspicious lesion involving left lateral border of tongue and was referred for FDG PET/CT scan for further evaluation. PET/CT revealed increased FDG uptake in heterogeneously enhancing lesion involving the mid-third of the left lateral border of the tongue and few left cervical nodes. On examination of CECT, the uncinate process and head of pancreas were seen, while the body and tail of pancreas were not visualized anterior to the splenic vein. The potential space in the pancreatic bed was occupied with bowel loops. The patient did not give any history of pancreatitis and abdominal pain. On correlative T2 weighted MRI images of the abdomen, the body and tail of pancreas were not visualized, confirming the diagnosis of congenital agenesis of dorsal pancreas.

Discussion

The embryological development of pancreas begins at the fourth week of gestation from ventral and dorsal buds, which form as outpouchings from the endodermal lining at the junction of foregut and midgut. The ventral pancreatic bud rotates clockwise and fuses with the dorsal bud at the seventh week of gestation. The dorsal pancreatic bud forms the upper part of head, body, and tail of the pancreas, while the ventral bud forms the inferior part of head and uncinate process. The duct of ventral bud fuses with the distal part of duct of dorsal bud to form the main pancreatic duct (duct of Wirsung), while if the proximal part of the duct of the ventral bud persists, it forms the accessory pancreatic duct (duct of Santorini).[3]

The most common pancreatic congenital anomaly is pancreas divisum, which results due to failure of fusion of dorsal and ventral buds. Other pancreatic anomalies include annular pancreas and agenesis

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Kabnurkar R, Rokade ML, Bandekar K, Kamat N. Incidentally detected agenesis of dorsal pancreas on PET/CT: Case report and review of literature. Indian J Nucl Med 2017;32:33-5.

Address for Correspondence:
Rasika Kabnurkar, Department of Nuclear Medicine, Jupiter Hospital, Eastern Express Highway, Thane, India, E-mail id: mailrasika@yahoo.com

Access this article online

Website: www.ijnm.in

DOI: 10.4103/0972-3919.198471

Quick Response Code:

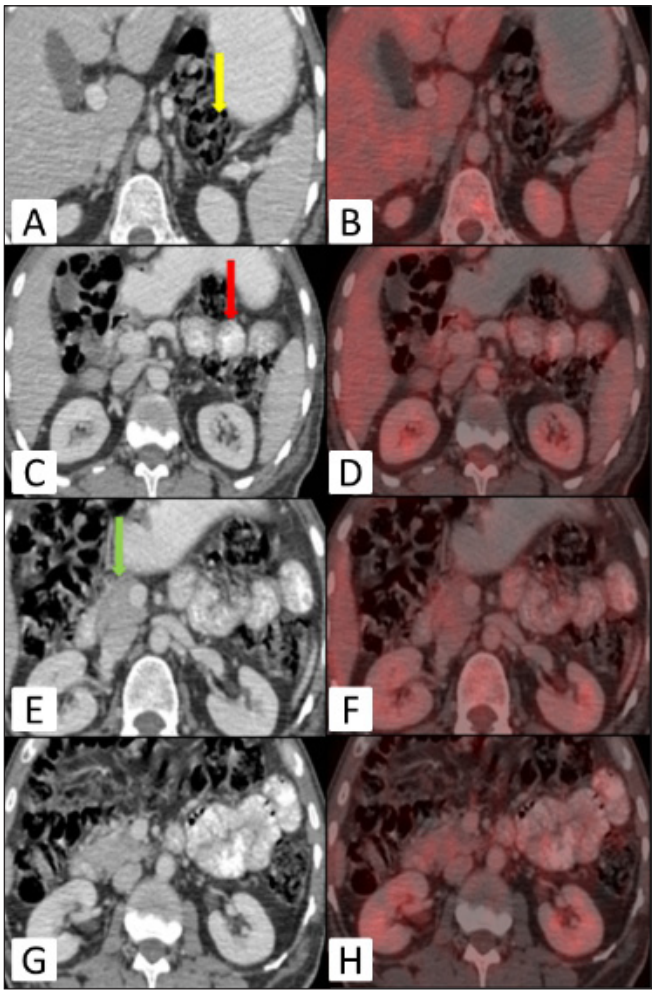

Figure 1: Serial axial CECT and corresponding fused FDG PET/CT images demonstrating presence of pancreatic head and uncinate process (green arrow, image E) with no visualization of pancreatic body and tail. The potential space in the pancreatic bed is filled up with splenic flexure of colon, stomach (yellow arrow , image a) and small bowel loops (red arrow, image C) demonstrating "Dependent stomach sign" and " Dependent Intestine sign" respectively confirming the diagnosis of ADP.

of either the ventral or the dorsal bud.[4] The complete agenesis and ventral agenesis of pancreas are incompatible with life.[5]

The agenesis of dorsal pancreas is a rare anomaly with less than 100 cases reported in literature. The exact mechanism and aetiology of ADP are unknown. A primary dysgenesis of the dorsal pancreatic bud and an ischemic insult to the developing pancreas are possible explanations.[1]

ADP may be partial or complete. In complete agenesis, the pancreatic neck, body, tail, and duct of Santorini are absent. A minor papilla is not present in the duodenum . In partial agenesis or hypoplasia of the dorsal pancreas, pancreatic body, accessory papilla, terminal end of the duct of Santorini are present and only the pancreatic tail is absent.[6]

The patients with ADP may present with non-specific abdominal pain. It is most commonly associated with

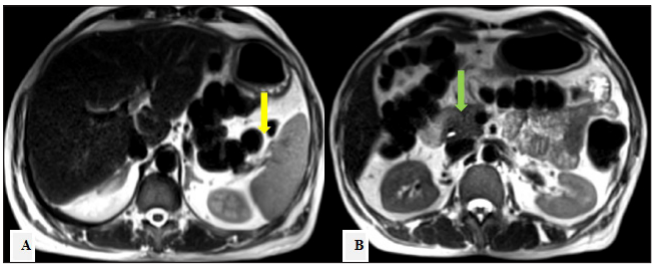

Figure 2: T2 weighted MRI images of abdomen showing absence of pancreatic tail near the splenic hilum . The potential space is filled with intestinal loops correlating with CT images (yellow arrow, image A). The pancreatic head and uncinate process is seen in the image B (white arrow).

diabetes mellitus followed by pancreatitis and sometimes exocrine pancreatic insufficiency.[2] However, with development of new imaging technologies, this congenital anomaly is often incidentally detected in asymptomatic patients undergoing imaging for an unrelated reason, similar to our case. Other associations of ADP reported in literature include polysplenia,[7] bicornuate uterus, [8] adenocarcinoma, etc.[9]

ERCP or MRCP demonstrating absence of the dorsal pancreatic duct is considered as confirmatory for diagnosis of ADP.[10] However, ERCP is an invasive procedure. Abdominal ultrasound has limitations in detecting ADP. This is because air-filled stomach and intestinal loops appear as an echogenic structure in the pancreatic bed, and may be misinterpreted as normal body and tail of pancreas on ultrasound examination.[2]

On CECT, ADP shows presence of pancreatic head with non -visualization of pancreatic body and tail anterior to the splenic vein. This potential space in the pancreatic bed is filled up with stomach and bowel loops called as "Dependent Stomach Sign" and "Dependent Intestine Sign", respectively. These signs are not seen in pancreatic lipomatosis as the pancreatic bed is filled up with fat and there is no space in the pancreatic bed to be occupied. Therefore, demonstration of these signs is considered as hallmark for diagnosis of ADP on CECT imaging, obviating the need for further radiological confirmation.[11] Zhou *et al.* have also reported a case of congenital ADP confirmed by three-dimensional reconstruction CT when no pancreatic tail and splenic artery branches to the pancreas were observed.[12]

In our patient, the pancreatic body and tail were not seen anterior to the splenic vein. There was no evidence of fatty replacement seen in the pancreatic bed. The space in the pancreatic bed was filled up with the splenic flexure of colon and small bowel loops (Dependent Intestine Sign). Hence, the diagnosis of ADP was confirmed in our case. Also, our patient was non-diabetic, which is uncommon in ADP.

As CT has become an integral part of PET studies, Nuclear Medicine Physician should be able to recognize incidental congenital anomalies and report the same. This case aims

to make the Nuclear Medicine Physician aware of this rare congenital anomaly.

Conclusion

This case aims to make the reader aware of recognizing ADP, a rare pancreatic congenital anomaly, which can be incidentally detected on cross-sectional imaging, its common associations, differential diagnosis on imaging and confirmatory methods.

Financial support and sponsorship

Nil

Conflicts of interest

There are no conflicts of interest

References

1. Guimarães AB, Guimarães CA, Manso JE. Agenesis or pseudoagenesis of the dorsal pancreas. Rev Col Bras Cir 2015;42:352-5.
2. Mohapatra M, Mishra S, Dalai PC, Acharya SD, Nahak B, Ibrarullah M. *et al.* Imaging findings in agenesis of the dorsal pancreas. Report of three cases. JOP 2012;13:108-14.
3. Fukuoka K, Ajiki T, Yamamoto M, Fujiwara H, Onoyama H, Fujita T. *et al.* Complete agenesis of the dorsal pancreas. J Hepatobiliary Pancreat Surg 1999;6:94-7.
4. Mortelé KJ, Rocha TC, Streeter JL, Taylor AJ. Multimodality imaging of pancreatic and biliary congenital anomalies. Radiographics 2006;26:715-31.
5. Voldsgaard P, Kryger-Baggesen N, Lisse I, Agenesis of pancreas. Acta Paediatr 1994;83:791-3.
6. Schnedl WJ, Reisinger EC, Schreiber F, Pieber TR, Lipp RW, Krejs GJ. Complete and partial agenesis of the dorsal pancreas within one family. Gastrointest Endosc 1995;42:485-7.
7. Wainwright H, Nelson M. Polysplenia syndrome and congenital short pancreas. Am J Med Genet 1993; 47:318-20.
8. Lal A, Pavunesan SK, Mahalingam H, Garg M, Sinha SK, Khandelwal N. A triad of complete dorsal pancreatic agenesis, pancake kidney and bicornuate uterus. An association or an incidental finding: first case in literature. JOP 2015;16:189-91.
9. Ulusan S, Yakar T, Koc Z, Kayaselcuk F, Torer N. Adenocarcinoma of the pancreas associated with dorsal agenesis. Pancreas 2006;33:437-9.
10. Pasaoglu L, Vural M, Hatipoglu H G, Terekliogl G, Koparala S. Agenesis of the dorsal pancreas. World J Gastroenterol 2008;14:2915-6.
11. Karcaaltincaba M. CT differentiation of distal pancreas fat replacement and distal pancreas agenesis. Surg Radiol Anat 2006;28:637-41.
12. Zhou Y, Chen M, Liu Y. Agenesis of dorsal pancreas confirmed by three-dimensional reconstruction CT. Int J Clin Exp Med 2014;7:3110-2.