CASE REPORT

A Rare Congenital Anomaly of the Pancreas: A Cadaveric Case Report

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ABSTRACT

Context The pancreas is formed by ventral and dorsal pancreatic buds which arise from the endodermal lining of the gut. When the duodenum rotates to the right, the ventral pancreatic bud migrates dorsally and finally come and lie below the dorsal pancreatic bud. The developmental errors in the rotation of these components lead to annular pancreas. **Case report** We report a rare type of the incomplete annular pancreas around the contents of right free margin of lesser omentum. There was an extra pancreatic mass situated horizontally immediately above the superior border of the pancreas, situated behind the lesser omentum. The right end of this mass extended into the epiploic foramen and incompletely encircled the portal vein, bile duct and hepatic artery proper from behind. The left end of the extra pancreatic mass was extended towards the lesser curvature of the stomach. Further, this mass completely surrounded the origin of three branches of the celiac trunk. Its right end was found to be continuous with the head of the pancreas, close to the pylorus. Histology of the extra pancreatic mass revealed the presence of normal pancreatic tissue. **Conclusion** preoperative diagnosis of this rare anomaly is of clinical importance during surgeries involving the contents of right free margin of lesser omentum and epiploic foramen.

INTRODUCTION

The pancreas is formed by ventral and dorsal pancreatic buds originating from the endodermal lining, at the junction of foregut and midgut during fourth week of gestation. The dorsal pancreatic bud is situated in the dorsal mesentery, whereas ventral pancreatic bud is closely related to the developing bile duct. Eventually when duodenum rotates to the right, the ventral pancreatic bud migrates dorsally and finally comes to lie immediately below and behind the dorsal pancreatic bud. Later, the parenchyma and duct system of ventral pancreatic bud fuses completely with the dorsal pancreatic bud. The ventral pancreatic bud forms the inferior and posterior part of the head and the dorsal pancreatic bud forms the remainder of the head, and the whole of the body and tail of the gland [1].

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Owing to its complex process of development, the congenital anomalies of the pancreas occur more frequently. Choledochal cysts, anomalous pancreatico-biliary junction, annular pancreas, heterotopic pancreas and pancreas divisum are the reported anomalies of the pancreas in the past [2, 3, 4, 5, 6]. Amongst, the annular pancreas is a rare type of congenital anomaly in which a mass of pancreatic tissue is found surrounding the second part of the duodenum like a ring. Its incidence is estimated to occur in 1 out of 12,000-15,000 newborns [7]. In most of the cases, this anomaly is asymptomatic [8]. Some cases it results in clinical complications like obstruction of the duodenum, pancreatitis, obstructive jaundice, peptic ulcer, and perforation of the duodenum or peritonitis.

Herein, we report a rare type of incomplete annular pancreas in a formalin embalmed male cadaver.

CASE REPORT

During the regular dissection classes for medical undergraduates, we came across a rare type of congenital anomaly of the annular pancreas. It was observed in an approximately 55-year-old male cadaver of South Indian origin. An apparently normal sized pancreas was situated across the posterior abdominal wall. It had presented classical

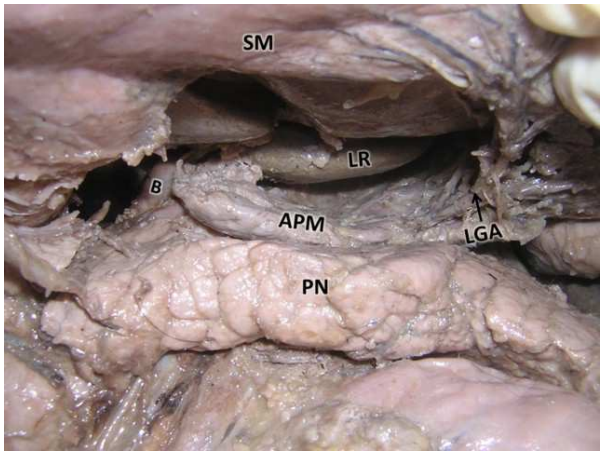

Figure 1. Dissection of supracolic compartment of abdomen showing the accessory pancreatic mass (APM), above the superior border of body of pancreas (PN). Note the accessory pancreatic mass extending horizontally from the right free margin of lesser omentum to the lesser curvature of the stomach (SM). Left gastric artery (LGA) piercing the accessory pancreatic mass is also seen.
B: bile duct; LR: liver

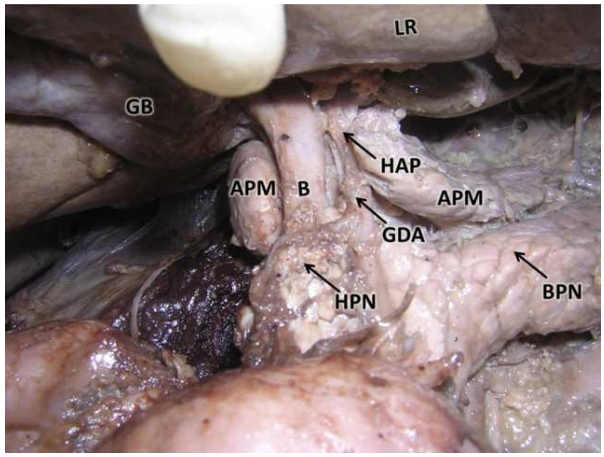

Figure 3. Closer view of dissection of supracolic compartment of abdomen, showing the accessory pancreatic mass (APM) above the pancreas. Note the accessory pancreatic mass is incompletely encircling the contents of right free margin of the lesser omentum.
B: bile duct; BPN: body of the pancreas; GB: gallbladder; GDA: gastroduodenal artery; HAP: hepatic artery proper; HPN: head of the pancreas; LR: liver

parts; head, body and tail. Head was situated within the concavity of the duodenum and uncinate process, arising from its inferior border which was smaller than the normal. There was an extra pancreatic mass situated horizontally immediately above the superior border of the pancreas, behind the lesser omentum (Figure 1). The right end of this mass extended towards the epiploic foramen and incompletely encircled the portal vein, bile duct and hepatic artery proper from behind. At the level of epiploic foramen, the mass was found to be continuous with the head of the pancreas (Figures 2 and 3). The left end of the extra pancreatic mass extended towards the lesser curvature of the stomach (Figure 1). This mass completely surrounded the origin of three branches of the celiac trunk. Further, the left gastric artery coursed through the substance of this mass on its way to the cardiac end of the stomach (Figure 1). The initial segment of the splenic artery was found in the gap between the extra pancreatic mass and superior border of body of the normal pancreas. No other associated anomalies were observed in the supracolic compartment of the abdomen. Histology of the extra pancreatic mass revealed the presence of classical pancreatic lobules with serous acini, similar to the native pancreas (Figure 4).

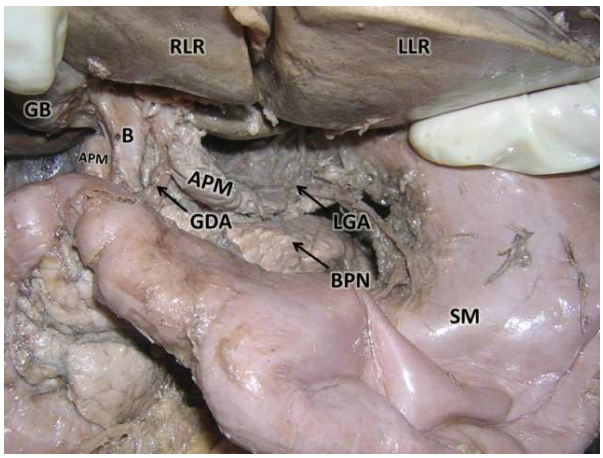

Figure 2. Dissection of supracolic compartment of abdomen, showing the accessory pancreatic mass (APM) above the pancreas. Note the accessory pancreatic mass is incompletely encircling the contents of right free margin of the lesser omentum.
B: bile duct; BPN: body of the pancreas; GB: gallbladder; GDA: gastroduodenal artery; LGA: left gastric artery; LLR: left lobe of the liver; RLR: right lobe of the liver; SM: stomach

DISCUSSION

Annular pancreas is a rare congenital anomaly where a band of pancreatic tissue surrounds the second part of the duodenum either completely or incompletely [7]. Usually, this band is in continuity with the head of the pancreas. This anomaly results

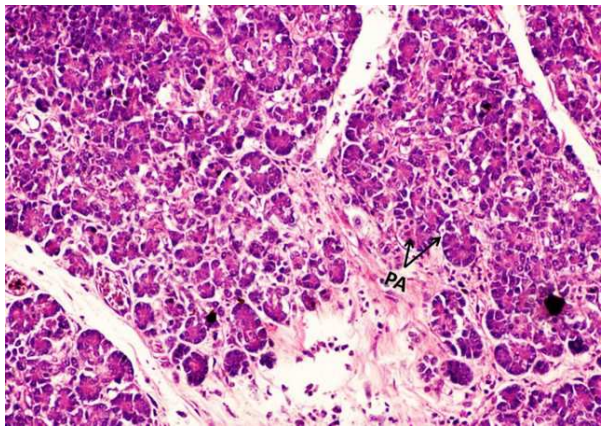

Figure 4. Histology of extra pancreatic mass showing the pancreatic tissue with classical lobular arrangement. Note the pancreatic serous acini (PA) in the lobule. (H&E staining, 10x magnification).

due to the embryonic developmental failure. Under normal conditions, the two components of ventral pancreatic bud (right and left) fuse and rotate clockwise along with the duodenum in such a way that they come to lie below the dorsal pancreatic bud [1]. The pathogenesis of annular pancreas has been demonstrated by two main hypotheses. According to Lecco, the tip of the right ventral pancreatic bud adheres to the duodenal wall and stretches to form a ring during normal rotation [9]. The other hypothesis by Baldwin: the persisted portion of left ventral bud develops into a complete circle of pancreatic tissue around the duodenum [10]. As the annular pancreas rich in PP-rich islets which is said to be derived from the right ventral pancreatic bud, some pathologists support Lecco's hypothesis [11, 12]. Both Lecco's and Baldwin theories could convincingly explain all the proposed classifications of the annular pancreas [13, 14, 15, 16, 17]. The detailed study on developmental stages of pancreas, in the lower species gave a convincing explanation on the etiology of the annular pancreas in humans [18, 19, 20]. In pigs this anomaly is resulted by the two ventral and one dorsal pancreatic buds [18]. In chickens and frogs, the normal pancreas is formed by the fusion between two ventral and one dorsal pancreas [19, 20]. By considering the developmental stages in other species, the annular pancreas might be attributed to the persistence of the left ventral bud in the humans.

Annular pancreas is estimated to occur in one of every 12,000-15,000 live births [7]. The formed pancreatic ring around the duodenum may be of complete (25%) or partial (75%) [16, 21]. In autopsy cases, the incidence of annular pancreas has been reported to be 0.005-0.015% [22]. This anomaly occurs commonly in males. The congenital abnormalities such as esophageal atresia, imperforate anus, congenital heart disease, malrotation of the midgut, and Down syndrome are usually associated with annular pancreas. Usually, this anomaly is asymptomatic throughout life, and it may go unnoticed. But, occasionally it may cause duodenal constriction and obstruction at any point of age. It can also leads to inflammation or malignancies of the annulus, pancreatitis, obstructive jaundice, peptic ulcer, and perforation of the duodenum or peritonitis [23]. An extensive review on patients with annular pancreas by the Jimenez *et al*. has observed that majority of the cases (75%) were presented during the first week of life [24]. Siew *et al.* have reported a case of annular pancreas causing upper gastrointestinal bleeding in a child [23]. The uncinate process may surround the portal vein and superior mesenteric vein like a ring, and it may result in a rare anatomical anomaly which is called as portal annular pancreas. Earlier, incidences of such cases have been reported [25, 26]. Prior knowledge of these rare types of cases is of clinical importance and should be considered carefully during pancreatic resections. Preoperative diagnosis of the annular pancreas can be made using MRI with cholangiopancreatography [27]. A study by Maker *et al*. has revealed that though despite having various diagnostic tools including endoscopic retrograde cholangiopancreatography preoperative diagnosis is successfully reached in only 60% of patients. Enteroenterostomy is believed to be the best intervention choice as it provides wide array of surgical options when additional factors need to be addressed [28].

The extra pancreatic tissue which is situated outside the usual anatomic location of the pancreas without any anatomic or vascular continuity with the native pancreas is defined as heterotopic pancreas [29]. Usually, it occurs at various sites in the gastrointestinal tract especially in the wall of the stomach and small intestine. It is also found to occur in the common bile duct, esophagus, mesentery, spleen and liver [30]. Genetically and physiologically heterotopic pancreas is similar to the native pancreas [31]. Majority cases of heterotopic pancreas are asymptomatic [32] and are usually observed incidentally during laparotomy or autopsy [33]. In the past, the reported incidence of heterotopic pancreas during laparotomy is 0.5% and at autopsy is 1.7% [29]. The embryological basis for the occurrence of heterotopic pancreas has been explained by several possible theories. Amongst one of the theory explains that as the development of the normal pancreas follows several evaginations, and originates from the wall of the primitive duodenum, one or more evaginations may remain in the intestinal wall or unusual shifting of this embryonic remnant along with the development of the gastrointestinal tract results in ectopic pancreatic tissue [34]. On other hand, another theory demonstrates that during embryogenesis pancreatic metaplasia of the endodermal tissues localized in the gastric submucosa may occur [34]. The extra pancreatic mass observed in the present case may not be the heterotopic pancreas as it is clearly said that heterotopic pancreas should not have any anatomical or vascular communication with the main pancreas, but in our case the extrapancreatic mass was connected to the head of the pancreas behind the bile duct.

In the present case, the observed extra pancreatic mass can be a rare type of incomplete annular pancreas and is named so because it is connected to the head of the main pancreas and partially encircled the portal vein, bile duct and the hepatic artery. This mass of pancreatic tissue might compress the portal vein or bile duct at any stage of life resulting in obstructive jaundice or portal

hypertension. The anomaly observed here is likely to be prenatal in development as celiac trunk branches were budding through the observed extra pancreatic mass. The histology of pancreatic mass and the normal sized coeliac, pancreaticosplenic and hepatic nodes ruled out the possibility of a tumour of pancreas. It is different from the usual annular pancreas where the pancreatic tissue forms a ring around the third part of the duodenum or rarely ring of pancreatic tissue is also observed around the portal vein forming the portal annular pancreas. The developmental cause of this rare anomaly needs further investigation. Since the accessory pancreatic mass was obstructing the epiploic foramen, it may give wrong impression during the preoperative internal hernia diagnosis at this foramen. Prior knowledge of this anomaly also is of clinical importance for surgeons operating on contents of right free margin of lesser omentum and celiac trunk branches.

Conflicts of interest The authors have no potential conflicts of interest

References

1. Langman J. Medical embryology. 6th ed. Baltimore, Md: Williams & Wilkins; 1989: pp. 245-247.

2. Tadokoro H, Suyama M, Kubokawa Y,Sai JK. Persistence of the left part of the ventral pancreas may cause congenital biliary dilatation. Pancreas 2003; 27(1):47– 51.

3. The Japanese Study Group on Pancreaticobiliary Maljunction. Diagnostic criteria of pancreaticobiliary maljunction. Journal of Hepato-Biliary-Pancreatic Surgery 1994; 1(3):219– 221.

4. Tadokoro H, Takase M, Nobukawa B. Unusual fusion between ventral and dorsal primordia causes anomalous pancreaticobiliary junction. Pathology International 2008; 58(8):498–502.

5. Kiernan PD, ReMine SG, Kiernan PC, ReMine WH. Annular pancreas: may clinic experience from 1957 to 1976 with review of the literature," Archives of Surgery 1980; 115(1):46–50.

6. Neupert G, Appel P, Braun S, Tonus C. Heterotopic pancreas in the gallbladder. Diagnosis, therapy, and course of a rare developmental anomaly of the pancreas. Chirurg 2007; 78:261-264.

7. Lainakis N, Antypas S, Panagidis A et al. Annular pancreas in two consecutive siblings: an extremely rare case. European Journal of Pediatric Surgery. 2005; 15(5): 364–368.

8. Ravitch, MM. The pancreas in infants and children. Surg Clin North Am. 1975; 55(2):377–385

9. Lecco TM. Zur morphologie des pankreas annulare. Sitzungsberichte der Heidelberger Akademie der Wissenschaften 1910; 119: 391–406.

10. Baldwin WA. A specimen of annular pancreas. The Anatomical Record 1910; 4(8):299–304.

11. Sessa F, Fiocca R, Tenti P et al. Pancreatic polypeptide rich tissue in the annular pancreas. A distinctive feature of ventral primordium derivatives. Virchows Archiv a Pathological Anatomy and Histology 1983; 399(2):227–232.

12. Suda K. Immunohistochemical and gross dissection studies of annular pancreas. Acta Pathologica Japonica 1990; 40(7):505–508.

13. Agha FP, Williams KD. Pancreas divisum: incidence, detection, and clinical significance. American Journal of Gastroenterology 1987; 82(4):315–320.

14. Lehman LA, Sherman S. Diagnosis and therapy of pancreas divisum. Gastrointestinal Endoscopy Clinics of North America 1998; 8(1):55–77.

15. Morgan DE, Logan K, Baron TH, Koehler RE, Smith JK. Pancreas divisum: implications for diagnostic and therapeutic pancreatography. American Journal of Roentgenology 1999; 173(1): 193–198.

16. Sandrasegaran K, Patel A, Fogel EL, Zyromski NJ, Pitt HA. Annular pancreas in adults. American Journal of Roentgenology 2009; 193(2):455–460.

17. Soto JA, Lucey BC, Stuhlfaut JW. Pancreas divisum: depiction withmulti-detector row CT. Radiology 2005; 235(2): 503–508.

18. Lewis FT. The bi-lobed form of the ventral pancreas in mammals. American Journal of Anatomy 1911; 12(3):389–400.

19. Kim SK, Hebrok M, Melton DA. Pancreas development in the chick embryo. Cold Spring Harbor Symposia on Quantitative Biology 1997; 62:377–383.

20. Slack JM. Developmental biology of the pancreas. Development 1995; 121(6):1569–1580.

21. Gress F, Yiengpruksawan A, Sherman S et al. Diagnosis of annular pancreas by endoscopic ultrasound. Gastrointestinal Endoscopy 1996; 44(4):485–489.

22. Ravitch MM, Woods AC Jr. Annular pancreas. Annals of Surgery 1950; 132(6):1116–1127.

23. Siew JX, Yap TL, Phua KB et al. Annular Pancreas: A Rare Cause of Upper Gastrointestinal Bleeding in a Child. J Pediatr Gastroenterol Nutr. 2012 Aug 3.

24. Jimenez JC, Emil S, Podnos Y, Nguyen N. Annular pancreas in children: a recent decade's experience. J Pediatr Surg 2004; 39(11):1654-7.

25. Kobayashi S, Honda G, Kurata M, Okuda Y et al. Pancreaticoduodenectomy in portal annular pancreas: report of a case. Surg Today 2012 Aug 2. [Epub ahead of print]

26. Jang JY, Chung YE, Kang CM, Choi SH et al. Two cases of portal annular pancreas. Korean J Gastroenterol 2012; 60(1):52-55.

27. Cunha JE, de Lima MS, Jukemura J, Penteado S, et al. Unusual clinical presentation of annular pancreas in the adult. Pancreatology 2005; 5(1):81-85.

28. Maker V, Gerzenshtein J, Lerner T. Annular pancreas in the adult: two case reports and review of more than a century of literature. Am Surg 2003; 69(5):404-410.

29. Mulholland KC, Wallace WD, Epanomeritakis E, Hall SR. Pseudocyst formation in gastric ectopic pancreas. JOP 2004; 5: 498-501.

30. Ormarsson OT, Gudmundsdottir I, Marvik R. Diagnosis and treatment of gastric heterotopic pancreas. World J Surg 2006;30:1682-1689.

31. Zhang L, Sanderson SO, Lloyd RV, Smyrk TC. Pancreatic intraepithelial neoplasia in heterotopic pancreas: evidence for the progression model of pancreatic ductal adenocarcinoma. Am J Surg Pathol 2007; 31: 1191-1195.

32. Christodoulidis G, Zacharoulis D, Barbanis S, et al. Heterotopic pancreas in the stomach: a case report and literature review. World J Gastroenterol 2007;13:6098-6100.

33. Barbosa J, Dockerty MB, Waugh JM. Pancreatic heterotopia review of the literature and report of 41 authenticated surgical cases. Surg Gynecol Obstet 1946; 82;527-542.

34. Chandan VS, Wang W. Pancreatic heterotopia in the gastric antrum. Arch Pathol Lab Med 2004; 128: 111-112.