A Rare Annular Pancreatic Anomaly

Shanubhoganahalli Puttamallappa Vinutha[1], Mathada Vamadevaiah Ravishankar[2]

[1]Department of Anatomy, JSS Medical College, JSS Academy of Higher Education and Research, Mysore, Karnataka, India. [2]Department of Anatomy, JSS Medical College, JSS Academy of Higher Education and Research, Mysore, Karnataka, India.

PRESENTATION OF CASE

During routine abdominal dissection of an adult male cadaver aged about 60 years, an anatomical variation was found in the pancreaticoduodenal area. The dissection was performed in the Department of Anatomy, JSS Medical College, Mysuru. It was observed that the pancreatic tissue completely encircled second part of duodenum. It consisted of a 360-degree pancreatic ring, it measured around 2 cm in width on its lateral aspect and 5 cm width on its posterior aspect. It measured 4 cm width on its anterior aspect. The part of the duodenum, which lies proximal and distal to the annular pancreas was found distended. A piece of annular pancreas was collected, later was subjected to the histopathological examination under H & E stain. The microscopic structure showed the normal architecture of the pancreatic tissue.

Understanding the pancreas development is essential to understand its congenital defects. It develops from ventral and dorsal pancreatic buds; it lies near the developing primitive duodenum. Due to the axial rotation of the second part of the duodenum, the ventral pancreatic bud winds around the posterior surface of the duodenum to meet with the dorsal pancreatic bud. This normal event may tend to show some derangement leading to variable conditions of pancreatic anomalies like the annular pancreas, pancreas divisum, pancreatic duct defects, etc. Though the congenital pancreatic anomalies are relatively uncommon, they may remain asymptomatic for a long duration, often they may find incidentally in some investigations or during surgeries. During our routine dissections, we have come across one such anomaly of the annular pancreas. Meeting such anomalies or variations during gross anatomy dissections will certainly create an overwhelming interest among the students to understand and correlate its embryological basis, and its clinical implications.

Annular pancreas (AP) was first described by Tiedemann (1818), the name "Annular Pancreas" coined by Ecker (1862). It is a rare congenital anomaly formed by a thin band of normal pancreatic tissue that completely or partially encircles the second part of duodenum. The reported incidence in adults varies from 0.005 to 0.015 %.[1]

Dual gland pancreas starts developing in the 5th week of the gestation period. It is formed by two buds, dorsal and ventral, originating from the endodermal lining of the duodenum. Dorsal pancreatic bud is in the dorsal mesentery, whereas the ventral pancreatic bud lies close to the bile duct. When the duodenum rotates to the right, it becomes C shaped; the ventral pancreatic bud moves dorsally like the shifting of the entrance of the bile duct. Finally, the ventral bud comes to lie immediately below and behind the dorsal bud. Later the parenchyma and the duct systems of the dorsal and ventral pancreatic buds fuse.

The ventral bud forms the uncinate process and the inferior part of the head of the pancreas. The remaining part of the pancreas is derived from the dorsal bud. The ventral pancreatic bud consists of two components that normally fuse and rotate around the duodenum so that they come to lie below the dorsal pancreatic bud. Occasionally, right portion of the ventral bud migrates along its normal route, but left migrates in the opposite direction. In this manner, the duodenum is surrounded by pancreatic tissue, and an annular pancreas is formed. During the third month of fetal life pancreatic islets of Langerhans develop from the pancreatic parenchyma, which is scattered throughout the pancreas dominating in its tail end; Insulin hormone secretion begins at around 5th month of fetal life.[2,3]

Corresponding Author:
Dr. Vinutha S P.
Assistant Professor,
Department of Anatomy,
JSS Medical College,
JSS Academy of Higher Education and Research,
Mysuru – 570015, Karnataka, India.
E-mail: vinuthasp@jssuni.edu.in

DOI: 10.14260/jemds/2020/624

How to Cite This Article:
Vinutha SP, Ravishankar MV. A rare annular pancreatic anomaly. J Evolution Med Dent Sci 2020;9(38):2859-2862, DOI: 10.14260/jemds/2020/624

Submission 25-05-2020,
Peer Review 29-07-2020,
Acceptance 04-08-2020,
Published 21-09-2020.

Figure 1. Anterior View of the Annular Pancreas in the Dissected Specimen (Yellow Arrow)

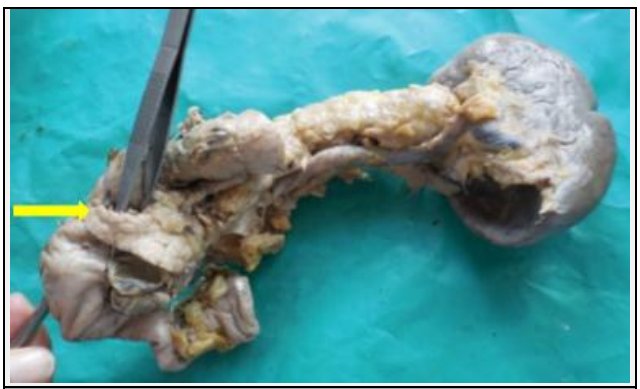

Figure 2. Lateral View of the Annular Pancreas in the Dissected Specimen (Yellow Arrow)

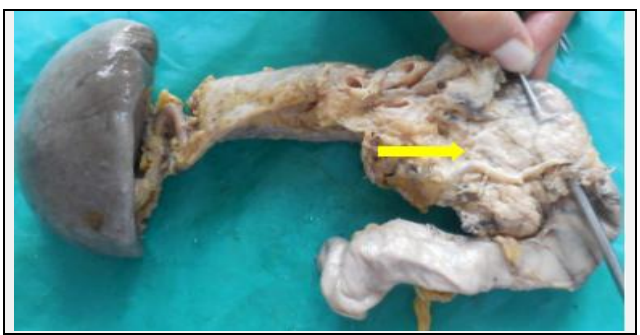

Figure 3. Posterior View of the Annular Pancreas in the Dissected Specimen (Yellow Arrow)

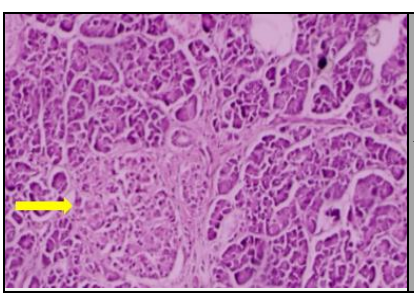

Figure 4. Microscopic Image of Annular Pancreas Showing Normal Pancreatic Architecture and Islets of Langerhans (Yellow Arrow)

DISCUSSION

Several types of congenital pancreatic anomalies have been described; one of them is the annular pancreas. It is an encircling pattern of pancreatic tissue around the second part of the duodenum. Choledochal cyst is an anomaly related to abnormal fusion of pancreatic duct with the bile duct. Sometimes the ventral and dorsal pancreatic buds will remain separated resulting in pancreatic divisum.[4]

The annular pancreas may exist in complete or incomplete form. Through contrast CT it was found that in case of the complete annular pancreas, a ring of pancreatic tissue was encircling the duodenum near the gastric outlet was presented with the abnormal dilation of stomach and duodenum. Under radiological investigations, an incomplete annular pancreas is showing an appearance of "crocodile jaw".[5] A rare case of the double annular pancreas was reported; where the small segment of pancreatic tissue is extended over the junction between the wall of the first and second parts of the duodenum. Another segment of the pancreas is covering the third part of the duodenal wall from the anterior aspect.[6]

The annular pancreas may also coexist with other congenital defects in children; the most common congenital chromosomal anomaly associated with AP is Down's syndrome, low birth weight, cardiovascular anomaly, etc.[7]

An annular pancreas usually requires surgical treatment. Presently through advanced investigative techniques have contributed a lot in understanding the pathology of the disease, which has enabled the surgeon to diagnose and take appropriate surgical measures.[8] Circum portal annular pancreas is another condition where the portal vein is surrounded by aberrant pancreatic tissue. Such patients were presented with unusual clinical signs and symptoms. Preoperative radiological investigations are playing a vital role in the identification of such rare presentations. Hence surgeons can plan accordingly to prevent complications during and after surgical interventions.[9]

There are several cases of the pancreatic anomaly was also found to be associated with the malignant pancreatic changes.[10,11]

Histologically we can also identify developmentally displaced accessory pancreas tissue called ectopic pancreas; it can be found in the wall of some abdominal organs like stomach, duodenum, jejunum, etc. During the embryological developmental process, the small fragments/sluff of pancreatic tissue seeming to be dislodged into some other abdominal organs; mostly they are seen in the upper part of the GI tract. Such ectopic tissue may find to be placed evenly between the layers of the gastrointestinal tract.[12,13,14]

Molecular Basis

There are several experimental genetic studies that have revealed insight into the pancreatic anomaly. By creating suppression of certain genes like Pdx1 (pancreatic duodenal homeobox gene), Hlxb9, Isl1, Hex, etc. has resulted in the arrest of pancreas development at a very early stage. Disruption of transcription factor of the gene has curtailed development of pancreatic endocrine component where the formation of insulin-producing cells is affected. Such a genetic correlation has provided much interesting information about the genetic origin of diabetes mellitus.[15]

Hedgehog (Hh) gene plays an important role during the embryological development and tissue differentiation of the pancreas. Hh gene signals also play an important role in adult life as well. It was found that reduced Hh gene activity can lead to severe congenital birth defects in the pancreas as well as the intestine of humans and rats. Aberrant expressions of Hh gene can lead to defects like the annular pancreas. Interestingly Hh

signal deregulation was also found to correlates with a variety of cancers with metastatic changes.[16,17]

Bone morphogenetic proteins (BMPs) and fibroblast growth factors (FGFs) signalling play a role in the development of ventral and dorsal pancreatic buds. Several cellular gene transcription regulators like IPF1/PDX1, PBX1 and PDF1-P48 are showing their holistic influence on complete pancreas formation.[18]

There is a vast variation in the formation of the duct of Wirsung, it includes three main patterns of its presentations A) migration B) fusion and C) duplication anomaly. Anatomical knowledge of pancreatic duct patterns is also medically important to deal with unusual episodes of pancreatitis and its management. Knowledge of variation in pancreatic duct patterns will help in proper planning in the prevention of recurrent pancreatic infections and ductal injuries during surgical procedures. Hence radiological findings like magnetic resonance cholangio-pancreaticography (MRCP) will play an important role to establish anomalous pancreatic duct profile and management of recurrent pancreatic infections.[19,20,21]

Annular pancreas is a rare congenital anomaly, it may remain silent till late adult age. It can be presented clinically in adults with variable signs and symptoms, which often becomes difficult and challenging for the surgeon to diagnose. The radiological investigations like contrast CT scan can help to illustrate the AP, and ERCP (Endoscopic Retrograde Cholangio Pancreatography) and MRCP (Magnetic Resonance Cholangio Pancreatography) can help to trace the pancreatic anomaly and its duct pattern. In some cases, surgery may be an inevitable option to confirm the diagnosis. Depends on its severity of stenosis, the duodenal bypass surgery can be planned. Surgically it is often difficult to handle the pancreatic tissue because there is a risk of formation pancreatic fistula and adhesions caused by the fibrosis postoperatively. Duodeno-duodenostomy or duodeno gastrostomy is the better surgical options to handle such cases.[22,23] In a study by Satheesha nayak et al, the annulus lies in close relation to the right kidney, liver and renal vessels, it has to be preserved carefully during kidney transplants and liver surgeries. The surgeons may remove annular pancreas along with kidney, because the annulus resembles the fat surrounding the kidney. Its removal may lead to leaking of pancreatic juice, and can cause damage to the surrounding tissue by the active pancreatic enzymes.[24]

CONCLUSIONS

Pancreatic anomalies are relatively rare. They are seen in all age groups. Its vague clinical manifestations are often seen during the late adult life. Advanced diagnostics have helped in early diagnosis of such unusual clinical presentations. It is interesting to teach and correlate the embryological basis of such gross anomalies seen during cadaveric dissections. For the surgeons, it is important and challenging to handle such rare presentations.

Financial or Other Competing Interests: None.

REFERENCES

[1] Rondelli F, Bugiantella W, Stella P, et al. Symptomatic annular pancreas in adult: report of two different presentations and treatments and review of the literature. Int J Surg Case Rep 2016;20S(Suppl)21-4.
[2] Sadler TW. Langman's medical embryology. 11th edn. Lippincott Wiliams and Wilkins 2010:222-3.
[3] Baldwin WA. A specimen of annular pancreas. The Anatomical Record 1910;4(8):299-304.
[4] Tadokoro H, Takase M, Nobukawa B. Development and congenital anomalies of the pancreas. Anatomy Research International 2011;2011:1-7.
[5] Mittal S, Jindal G, Mittal A, et al. Partial annular pancreas. Proc (Bayl Univ Med Cent) 2016;29(4):402-3.
[6] Nayak S, Pamidi N, George BM, et al. A strange case of double annular pancreas. J Pancreas 2013;14(1):96-8.
[7] Yigiter M, Yildiz A, Firinci B, et al. Annular pancreas in children: a decade of experience. Eurasian J Med 2010;42(3):116-9.
[8] Russo A, Armand-Ugon G. Annular pancreas: case report and review of the literature. Eur J Anat 2015;19(2):197-200.
[9] Dhanapal B, Shankar G, Krishnaraj B, et al. Portal annular pancreas: case report of a rare anomaly. Cureus 2018;10(3):e2366.
[10] Bronnimann E, Potthast S, Vlajnic T, et al. Annular pancreas associated with duodenal carcinoma. World J Gastroenterol 2010;16(25):3206-10.
[11] Foo FJ, Gill U, Verbeke CS, et al. Ampullary carcinoma associated with an annular pancreas. JOP 2007;8(1):50-4.
[12] Faust DB, Mudgett CS. Aberrant pancreas, with review of the literature and report of a case. Ann Intern Med 1940;14(4):717-28.
[13] Horgan EJ. Accessory pancreatic tissue report of two cases. Arch Surg 1921;2(3):521-34.
[14] Straatman J, Meester RJ, Grieken NCTV, et al. Clinical picture: multiple sites of ectopic pancreatic tissue. Springerplus 2015;4:293.
[15] Habener JF, Kemp DM, Thomas MK. Minireview: transcriptional regulation in pancreatic development. Endocrinology 2005;146(3):1025-34.
[16] Lau J, Kawahira H, Hebrok M. Hedgehog signaling in pancreas development and disease. Cell Mol Life Sci 2006;63(6):642-52.
[17] Klieser E, Swierczynski S, Mayr C, et al. Differential role of Hedgehog signaling in human pancreatic (patho-) physiology: an up to date review. World J Gastrointest Pathophysiol 2016;7(2):199-210.
[18] Kim SK, MacDonald RJ. Signaling and transcriptional control of pancreatic organogenesis. Curr Opin Genet Dev 2002;12(5):540-7.
[19] Gandhi V, Gautam GP, Pai N. Recurrent acute pancreatitis and the reverse 'S'-shaped pancreatic duct. BMJ Case Rep 2019;12:e226492.
[20] Bang S, Suh JH, Park BK, et al. The relationship of anatomic variation of pancreatic ductal system and pancreaticobiliary diseases. Yonsei Med J 2006;47(2):243-8.

[21] Turkvatan A, Erden A, Turkoglu MA, et al. Congenital variants and anomalies of the pancreas and pancreatic duct: imaging by magnetic resonance cholangiopancreaticography and multidetector computed tomography. Korean J Radiol 2013;14(6):905-13.
[22] Patra DP, Basu A, Chanduka A, et al. Annular pancreas: a rare cause of duodenal obstruction in adults. Indian J Surg 2011;73(2):163-5.
[23] Thomford NR, Knight PR, Pace WG, et al. Annular pancreas in the adult: selection of operation. Ann Surg 1972;176(2):159-62.
[24] Nayak BS, Yooi OM. A classic case of annular pancreas and its clinical implications. Int J Morphol 2011;29(2):559-61.