Portal Annular Pancreas - An Underreported and Unexpected Intraop Catastrophe - Review of Anatomy, Classification and Management

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Abstract: Portal annular pancreas (PAP) is an anatomic variation due to aberrant fusion of uncinate process of the pancreas which arises from the ventral bud and extends to fuse with the dorsal pancreatic bud by encircling the portal vein or superior mesenteric vein. In this article, we present a case report in a 59 year old male who presented with obstructive jaundice and features of G. O. O and diagnosed as having moderately differentiated adenocarcinoma on endoscopic biopsy from second part of duodenum. He was planned for an elective Whipple procedure, during pancreatic transection tuft of pancreatic tissue encircling the portal vein and SMV axis and joining the body of pancreas posteriorly was present. Portal annular pancreatic anomaly was identified intraoperatively. The retroportal tissue was soft in consistency and suture ligated with 3-0 prolene. Post operatively there was no POPF but developed ascites which was resolved spontaneously and discharged on post op day 13.

Keywords: PAP-portal annular pancreas, G. O. O – Gastric outlet obstruction

1. Introduction

There are three types of pancreatic fusion anomalies: Annular pancreas, pancreas divisum and portal annular pancreas.

Portal annular pancreas also known as circumportal pancreas is the rarest of these and is mostly asymptomatic with a varying incidence reported in literature ranging from 0.8 to 2.5%.1-4. One of the earliest reports of this fusion anomaly was reported bySugiuira et al. in 1987. Portal annular pancreas may develop during aberrant embryogenesis, with the ventral and dorsal pancreatic primordium fusing over the portal vein (PV) /superior mesenteric vein (SMV) due to which a rind of pancreatic parenchyma, encircles the portal vein (PV) or the superior mesenteric vein (SMV) and fuses with the body of the pancreas. It is an asymptomatic condition and is usually an incidental finding on abdominal imaging. This unusual anomaly is underreported in radiological studies due its rarity and can even be misinterpreted or overreported as locally advanced pancreatic cancer with surrounding SMV in a normal anatomical variant. It is important for surgeons because the postoperative pancreatic fistula (POPF) rates following pancreatic surgeries are higher in cases with portal annular pancreas than those with normal pancreas. The reported POPF rates was vs 16.5 % in normal anatomic variant after Whipples procedure (2). Pancreatic surgeons and GI radiologists should be aware of this rare anomaly even with low index of suspicion to prevent complications because POPF in pancreaticoduodenectomy is responsible for most of the morbidity and mortality in post operative period. The complexity associated with surgery in PAP is due to the varying MPD anatomy and two pancreatic resection surfaces. Adequate clinical knowledge about this anomaly during preoperative assessment will avoid any intraoperative surprise and catastrophe.

2. Case Report

A 59 year old male presented with painless progressive jaundice and pruritis with history of vomiting after food intake, he has loss of appetite and has lost 12 kgs inthe last three months. On examination he was icteric and palpable gall bladder was present. He has earlier consulted a gastroenterologist for the same for which endoscopy was done and showed ulcerative growth in second part of duodenum biopsy was done from the lesion and scope could not be passed beyond. Nasojejunal tube was placed for feeding in view of outlet obstruction features.
He was later imaged with contrast enhanced CT scan which showed 3* 2.8 cm ill defined hypodense mass lesion in Pancreaticoduodenal groove region – possibility of Groove pancreatitis with Infiltration in to medial duodenal wall at level of D1 and D2 causing significant narrowing. The mass partially encases the SMV in the region of uncinate process of pancreas and the MPD diameter was 4mm. He was planned for an elective whipples procedure. Pancreatic neck was transacted and during uncinate dissection tuft of pancreatic tissue was found encircling the portal vein simulating residual malignant tissue in the uncinate process and was an R2 resection. This tissue was extending posteriorly and joining the body of pancreas, but different in consistency with rest of malignant tissue in the head of pancreas. Anomaly of circumportal pancreas was interpreted at this stage of operation and then reviewed with the pre op images (Fig.1) confirming portal annular pancreas with absent duct in the retroportal tissue and a single anteportal main pancreatic duct. This has led to increased intra operative time and blood loss during dissection near PV-SMV junction. Portal vein is hooked with vascular loop rotated to left and the retroportal tissue is dissected from portal venous attachments, there was no duct in this tissue with single MPD identified in the anteportal portion of pancreas. Pancreatic stump was mobilised for about 4cm, the retroportal tissue was suture ligated with 3-0 prolene, by continuing suture technique. Pancreatic reconstruction was done by dunking technique of pancreaticogastrostomy and the annular portion dissected was not included in the anastomosis. Antecolic H-J (hepaticojejunostomy) done and antecolic G-J (gastrojejunostomy) done. Two abdominal drains were placed one in morisson’s pouch and other near PG site. Post operatively he had delayed PPH (Post Pancreatectomy haemorrhage) on POD-3 with intraluminal bleed in ryles tube aspirate which was treated conservatively with double dose of I. V Pantoprazole and resolved by post operative day 5. There was increased drain output which was serous in nature 350 ml, 200 ml and 100ml on post operative day one, two and three respectively which was gradually resolved. Drain fluid amylase on Post operative day 3, 5, 7 was 87, 56, 52 IU respectively and within normal limits without any POPF, (post operative pancreatic fistula). He had grade A DGE-delayed gastric emptying, oral liquids were allowed on POD-6and he was discharged on POD-13. HPE was moderate to poorly differentiated pancreatic ductal adenocarcinoma with PDAC and pTNM staging – T3N2Mx (5/8 LN)

![Image](image.jpg)

**Figure 1:** PRE OP-CECT showing MPD ante portal to portal vein with PAP – Type 3A-ANTEPORTAL MAIN PANCREATICDUCT
Embryogenesis of fusion anomalies of pancreas

There are three types of pancreatic fusion anomalies:
1) Annular pancreas-Incomplete and Complete
2) Pancreas divisum
3) Portal annular pancreas.

Portal annular pancreas is the rarest among all fusion anomalies

Prevalence and literature
PAP or circumportal pancreas the possibility of this anomaly during pancreatic head resection was first described by Sugiura et al. Complete fusion of the uncinate process with the body of the pancreas was described by Hamanaka et al. during a resection of the pancreas [1]. Karasaki et al. [2] reported the identification rate of portal annular pancreas to be 1.14%. Yilmaz and Celik [3] has reported prevalence of 0.8% and Ishigami et al [4] reported as 2.4% among general population. The prevalence of portal annular pancreas in general population was 25 out of 1000 patients i.e. 2.5% with a slight female predominance in a retrospective study conducted Rettu John and Simon et al [4]

They reviewed all the CT abdominal images conducted in a 13 month period most of which are non hepatobiliary pancreatic pathologies there was an increased frequency of this anomaly in females; female: male ratio was 1: 2.1. Karasaki et al reported prevalence among female to male was 1.3:1, whereas Harnossetal reported equal prevalence among males and females. This condition is clinically asymptomatic and undetected in most cases or can be incidentally detected when cross sectional imaging like CT/MRI done for some other intraabdominal condition.

Classifications and Subclassification
Joseph et al [3] has classified portal annular pancreas in to three subtypes

Type I-Is the fusion of the ventral bud of the pancreas with the body and retroportal MPD (RMPD)
Type II-when type I is associated with pancreas divisum with both RMPD+AMPD (ante portal main pancreatic duct)
Type III-when the uncinate process alone is involved in the encasement of the vessels and fusion with only AMPD (ante portal main pancreatic duct)

Each of the type is further divided in to a, b and c (suprasplenic, infra-splenic and mixed) depending on its relationship to the splenic vein

Figure 2: Intra operative image with annular pancreatic tissue above the SMV-SV confluence
a) PAP – portal annular pancreas Type A (karasaki et al)
b) MPD-main pancreatic duct
c) SMV-PV confluence
Review of literature and meta analysis of portal annular pancreas in patients undergoing pancreatic resections by Manish S. Bhandare, Sharles V. Shrikhande et al. [7] reviewed 29 articles which were retrospectively studied and total reported cases in literature were53 and of which the type 3A was the most common variant identified in 25 cases out of 53 reported cases among the different types. Our case in the present study was also type 3A anomaly and there was no POPF. Regarding POPF in PD, POPFs were reported for a mean overall fistula rate of 21.3%. According to a systematic review conducted by Harnoss et al which included 21 studies the pancreatic fistula rate in patients with PAP (12 pancreaticoduodenectomies and 3 distal pancreatic tomies being 46.7% as per ISGPS classification. [8] Retrospective metanalysis conducted by Manish S. Bhandare, Sharles V. Shrikhande et al [8] which included 29 studies with 53 patients identified 42.55% rate of POPF and 34% CR-POPF [clinically relevant] which was double the rate of POPF in PD without circumportal pancreas, after pancreaticoduodenectomy with out PAP the reported rate of POPF is highly variable, ranging from 2%-20% [10, 11]
POPF-related mortality rate was 1% after PJ and 0.8% PG after PD in systematic review and analysis of the POPF-related mortality rate in 60, 739 patients retrieved from the English literature published between 1990 and 2015 by Sergio Pedrazzoli [9].

If preoperative suspicion of portal annular pancreas during surgery for pancreas all these cases have to undergo MRI with MRCP to delineate proper pancreatic ductal anatomy, the threshold should be at the minimum to undergo an MRCP as pancreaticoduodenectomies or distal pancreatic resections with circumportal pancreas has high POPF rate compared to resections without this anomaly. There are 53 reported cases of portal annular pancreas in pancreaticoduodenectomy or in other pancreatic resections, the case from present study will be 54th reported case. Because of the rarity of the anomaly there are no large number case series, and most of them are published case reports.

The non-dominant cutting plane, i.e., the plane without MPD can be either sutured or stapled. (12) It has also been suggested that PG with vagnimation of the two resected pancreatic planes together into the stomach after PD helps to minimise resected volume of the pancreas as well as possibly reduce chance of POPF (13).

Portal annular pancreas seems to be associated with increased risk for POPF after pancreatic resection because of additional section plains and variable courses of the pancreatic ductal system. An intraoperative pancreatography might be useful for confirmation in selected cases. In PD, a shift of the resection plain to the left should be considered. After PD, pancreaticojenunostomy in types 1 and 2 is technically difficult because of the retroportal anastomosis; intype 3, ligation of a retroportal branch duct of the unciate process is possible. In the suprasplenic and infrasplenic type, an additional resection is required to liberate portal or supra mesenteric vein, respectively.

Preoperative or intraoperative identification of PAP is extremely essential so as to adapt to a different strategy during pancreatic resection as well as stump reconstruction to decrease the devasting complication of POPF.

Surgical strategies to minimise the rate of CRPF during PD should involve
1) Extended resections to left of SMV to have single MPD
2) Suture closure of the retroportal tissue
3) Pancreatocagastrotomy or PJ by dunking depending on the type of PAP.

### 3. Conclusion

PAP is frequently underreported by radiologists. It may also be overreported as locally advanced pancreatic mass which was actually a normal anatomic variant. Precise knowledge regarding this rare anomaly to HPB surgeons is of utmost importance during pancreatic resections in addition to the aberrant or replaced arterial vasculature to avoid intraop surprise

<table>
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<tr>
<th>Sr. no</th>
<th>Author</th>
<th>Patient</th>
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<th>Surgery</th>
<th>PAP type</th>
<th>Non MPD pancreas stump</th>
<th>PJ, PG</th>
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</table>
| 29    | 79/F              |         | Ampullary            | PD       | 3/B      | Staped                | PJ     | B    

**Table 1: Published cases with pancreatic resection in PAP (7)**
Non MPD pancreas stump—pancreas stump in PAP without the main pancreatic duct and its management
PD- Pancreatic Duodenectomy
PG- Pancreatic Gastrostomy
PJ- Pancreatic Jejunostomy
TP- Total Pancreatectomy
Multivisceral 14-subtotal gastrectomy+ right hemicolectomy+PD,
DP+splenectomy+L nephrectomy+ hemicolectomy
RAMPS radical antergrade modular pancreaticosplenectomy
HPD hepato-pancreaticoduductectomy
DP distal pancreatocutaneous
DPSP distal pancreatojejunostomy spleen preserving
CAR coeliac axis resection
PAP type Joseph/Karasaki
PDAC- Pancreatic ductal adenocarcinoma
IPMN- Intraductal papillary mucinous neoplasm
NET- Neuroendocrine tumour
RCC- Renal cell carcinoma.

Conflict of interest
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Consent
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