

**Portal Annular Pancreas with portal cavernoma formation with associated dorsal pancreatic agenesis-A rare case report**

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**Abstract**

Portal annular pancreas is a uncommon congenital anomaly resulting from fusion of the pancreatic parenchyma around the portal vein. Its causing portal cavernoma formation and association with dorsal pancreatic agenesis is rare

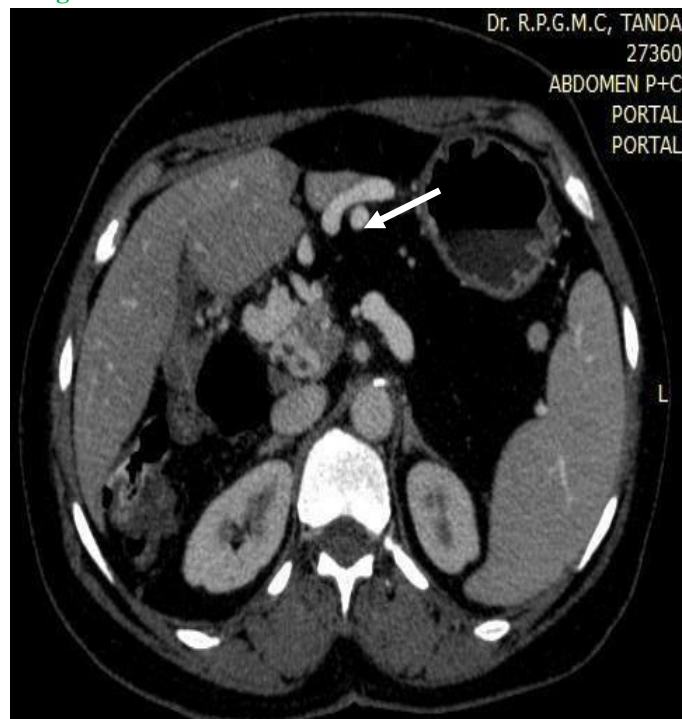
Case report We report a 51-year-old female who underwent contrast enhanced computed tomography for vague right hypochondrial pain.On CECT abdomen images there was presence of rind of pancreatic tissue around the portal vein causin its luminal narrowing with proximal dilation of portal vein tributaries with cavernoma formation.There was also presence of agenesis of dorsal pancreas in this patient.Conclusion This variant of portal annular pancreas with cavernoma formation associated with dorsal pancreatic agenesis has not yet been reported and we propose a new CT classification of the same.

Portal annular pancreas (PAP) is an uncommon and under-recognized congenital anomaly of the pancreas and with cavernoma formation with associated dorsal pancreatic agenesis is rarest and not been reported in the literature. In contrast to a conventional annular pancreas in which the pancreatic tissue encircles the second part of the duodenum, portal annular pancreas is characterized by encasement of the portal vein by rind of pancreatic parenchymal tissue<sup>1,2</sup>.

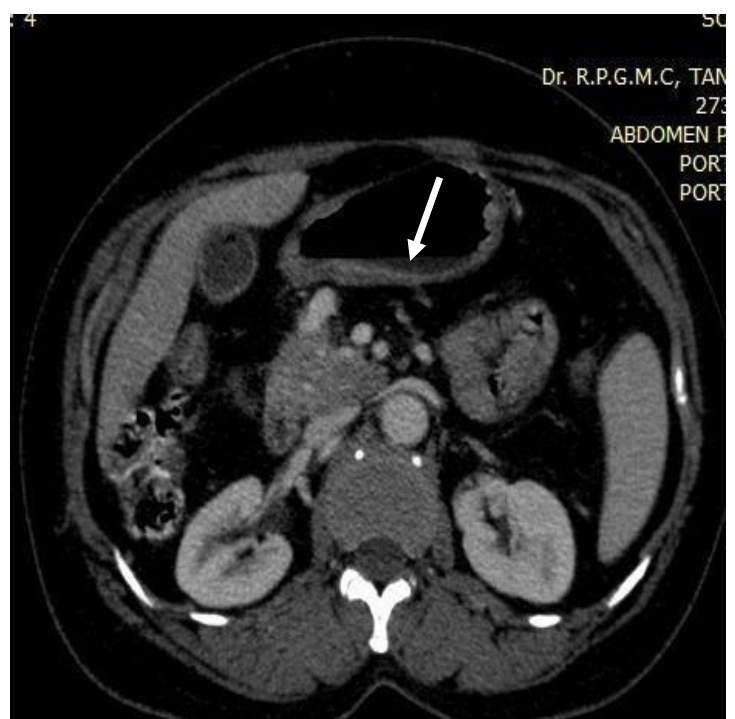
We noted, on an axial abdominal computed tomography study of a 69-year-old female with chronic vague pain in right hypochondriac region, anomalous pancreatic parenchyma encircling the main portal vein with proximal dilation of the portal vein tributaries with consequent cavernoma formation. There was also presence

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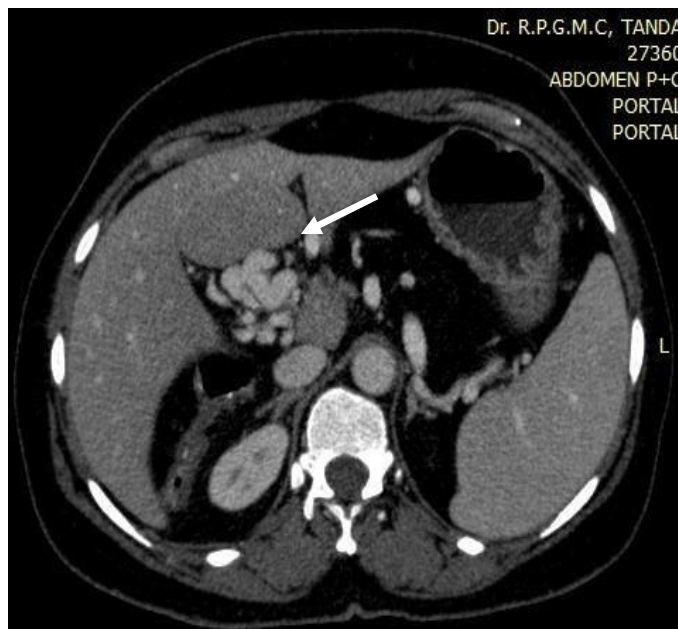
**Images**



a)



b)



c)

CECT images portal venous phase of 31 year old female showing (fig a) a ring of pancreatic tissue around portal vein (arrow) causing its luminal narrowing. There is presence of dorsal agenesis of pancreas (fig b) with dependant viscera sign (arrow). This is Type IV portal annular pancreas according to the new classification described in the text. Fig c shows dilated proximal portal channels with cavernoma formation (arrow).

Pancreas develops from a ventral and a dorsal bud of the duodenum<sup>2,8,9</sup>. The ventral bud forms the major part of the head and the uncinate process, whereas the dorsal bud forms upper part of the head, the body, and tail of the pancreas. The ventral bud rotates posteriorly during the 7<sup>th</sup> week of gestation to fuse with the dorsal bud so as to form the fully mature gland<sup>10</sup>. Rarely, this fusion occurs to the left of the mesenteric or portal vein, resulting in an encircling pancreatic parenchymal tissue around the portal vein. This has been referred to as the portal annular pancreas. Karasaki *et al*<sup>1</sup> and Ishigami *et al*<sup>13</sup> in their studies concluded that the prevalence of portal annular pancreas is not extremely low but is not readily recognized on preoperative imaging due to lack of adequate knowledge and awareness of this uncommon variant but its association with dorsal pancreatic tissue has not been reported yet<sup>11,12,13</sup>.

Imaging plays a pivotal role in the diagnosis of portal annular pancreas and contrast-enhanced multi-detector computed tomography (MDCT) is considered sufficient enough to establish the diagnosis. Joseph *et al*<sup>14</sup> have classified PAP into 3 types. In type I the ventral bud of the pancreas fuses with the dorsal bud posterior to the portal vein with a retroportal pancreatic duct (as is seen in the present case); type II has concomitant pancreas divisum; and type III is when the uncinate process alone is involved and the pancreatic duct is seen anterior to the portal vein (anteportal pancreatic duct)<sup>3,7,14,16</sup>.

Another classification describes this entity as suprasplenic (commonest), infrapleural, and mixed type based on the fusion of uncinate process with the body posteriorly above or below the level of the splenoportal confluence. It may be associated with the abnormal course of pancreatic duct (retroportal pancreatic duct) or pancreatic divisum<sup>17,18</sup>.

We propose a new classification of this entity with two groups.<sup>1,3,19,20</sup>

Group A-Portal annular pancreas(PAP) without complications

Group B-Portal annular pancreas(PAP) with complication of portal hypertension, pancreatitis, obstructive biliopathy etc.

Type I-Ventral bud fuses with the dorsal bud posterior to portal vein forming a ring of pancreatic tissue around portal vein

Type II-Pancreatic divisum

Type III-Anteportal pancreatic Duct

Type IV-Associated dorsal pancreatic agenesis

Our case is type IV and computed tomography scan is usually adequate for diagnosing this anomaly, which can be demonstrated by continuity of the extension of the uncinate process into the body of the pancreas in more than 2 slice however, the presence of the anomaly of pancreatic ducts can also be suggested by demonstrating the abnormal course of the pancreatic duct when present which can be depicted on MRCP or ERCP<sup>21,22</sup>.

### Conclusion

PAP is a rare anomaly and cavernoma formation with associated dorsal pancreatic agenesis more rarer. We emphasize on the new classification system which lay stress on complication associated with it and co-existent congenital anomalies of pancreas.

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