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

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Received date: July 20, 2019; Accepted date: August 18, 2019; Published date: September 18, 2019.

Citation for this Article: lokesh rana, Portal Annular Pancreas with portal cavernoma formation with associated dorsal pancreatic agenesis-A rare case report, J Clinical Imaging and Interventional Radiology, Doi: 10.31579/JCIIR.02/007

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Abstract
Portal annular pancreas is an 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 causing 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 tissue1,2.

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 of agenesis of dorsal pancreas in this patient.

Images

a) b)
Type II-Pancreatic divisum
Type III-Anteporal 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\textsuperscript{21,22}.

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.

References


