

INTESTINAL MALROTATION WITH SHORT PANCREAS, PREDUODENAL PORTAL VEIN, AZYGOS IVC AND POLYSPLENIA**¹Dr. Shivshankar Mishra, ²Dr. Rajeev Mehta, ³Dr. Tariq Patel and ⁴Dr. Zubair Kazi**¹DMRD, Junior Resident, Department of Imaging Sciences, Saifee Hospital, Mumbai-400004, India.²MD, Chairman & Head, Department of Imaging Sciences, Saifee Hospital, Mumbai-400004, India.³DM, Senior Consultant, Department of Gastroenterology, Saifee Hospital, Mumbai-400004, India.⁴DNB, Senior Consultant, Department of Imaging Sciences, Saifee Hospital, Mumbai-400004, India.***Corresponding Author: Dr. Shivshankar Mishra**

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ABSTRACT

Polysplenia syndrome is a rare situs ambiguous anomaly related to multiple spleens and anomalies of chest and abdominal organs. We report a case of polysplenia syndrome found in 34-year-old adult. CT scan is useful in the diagnosis by showing the situation of spleen, location of organs within the abdomen, and identification of associated anomalies. In this article, we illustrate anomalies of polysplenia syndrome and concentrate on abdominal organs.

KEYWORDS: Intestinal malrotation, short pancreas, polysplenia, preduodenal portal vein.**INTRODUCTION**

Polysplenia syndrome is a rare anomaly with a reportable incidence of one per 250 000 live births.^[1] It is related to numerous visceral anomalies such as multiple spleens, impaired visceral localization, inborn heart diseases, vascular abnormalities and azygous continuation of the inferior vena cava.^[1] However, short pancreas and preduodenal vein (PPV) are uncommon inborn anomalies in association with polysplenia syndrome.^[2] A patient with varied inborn anomalies as well as short pancreas and PPV in association with polysplenia is mentioned in this article.

CASE REPORT

A 34-year-old male complained of discomfort in the right upper quadrant of the abdomen with increased frequency of stools since past 5 months. At a local diagnostic centre, he was noted to have fatty liver and mildly enlarged mesenteric lymph nodes on an abdominal ultrasonogram 3 months ago. A definitive diagnosis, however, couldn't be made at that point. He visited our hospital for further evaluation. Colonoscopy done at our hospital suggested normal findings. Contrast-enhanced abdominal CT scan revealed intestinal malrotation with small bowel loops on the right and large bowel loops to the left of midline with inversion of SMA-SMV axis. Infrahepatic interruption of the IVC

with azygos continuation. Hepatic veins drained directly into the right atrium. There are numerous little spleens, one to three cm in diameter, within the left hypochondrium. The portal vein was positioned anterior to the duodenum and short pancreas with complete non-visualization of its body and tail.



Figure 1: SB-small bowel loops, LB-large bowel loops: intestinal malrotation with small bowel loops on the right and large bowel loops on the left.



Figure 2: SMA SMV inversion. SMA-Superior mesenteric artery, SMV-Superior mesenteric vein.

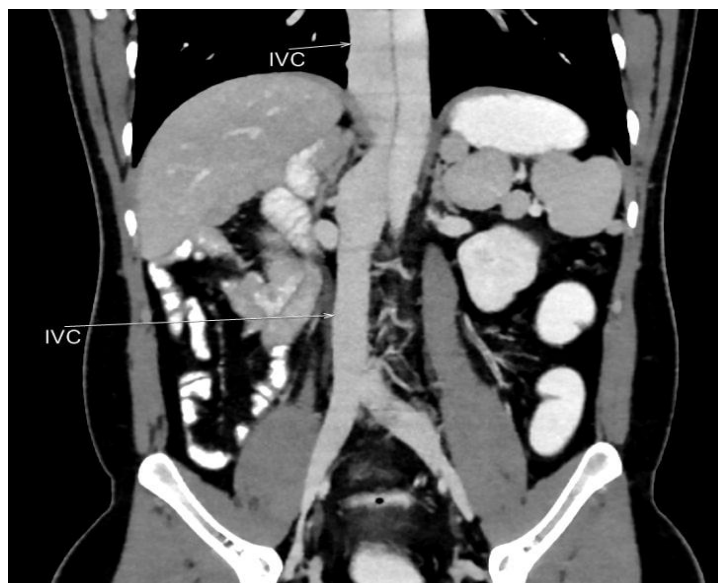


Figure 3: Azygous interruption of IVC in coronal view.

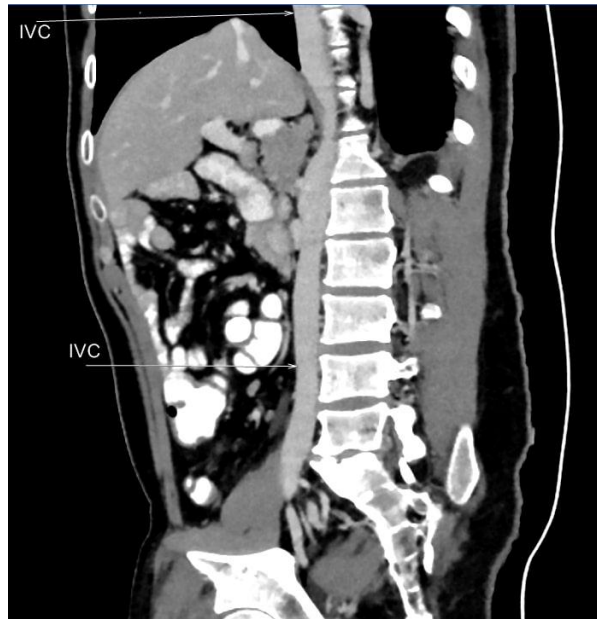


Figure 4: Azygos interruption of IVC in sagittal view.

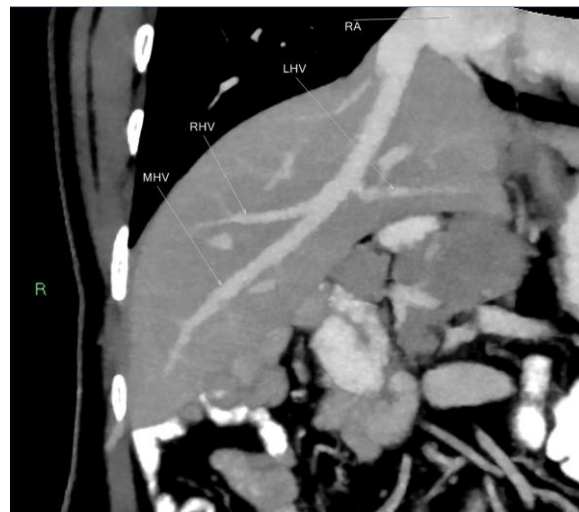


Figure 5: Hepatic veins draining directly into right atrium. RA-right atrium, RHV-right hepatic vein, MHV-middle hepatic vein, LHV-left hepatic vein.

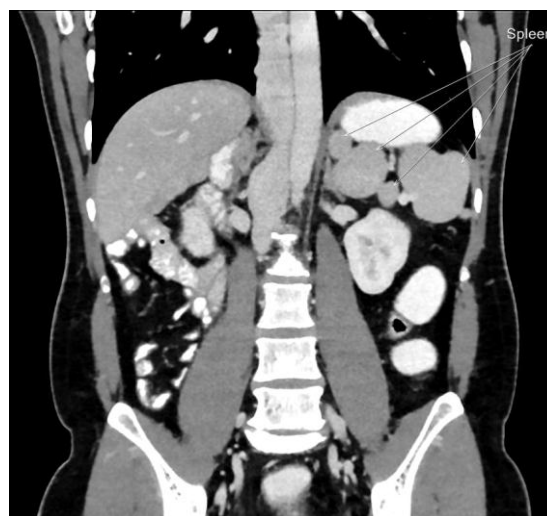


Figure 6: Polysplenia in coronal view.

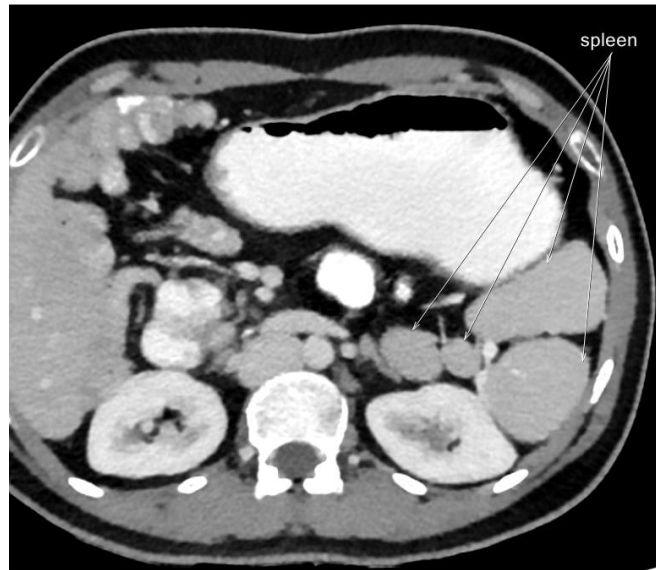


Figure 7: Polysplenia in axial view.

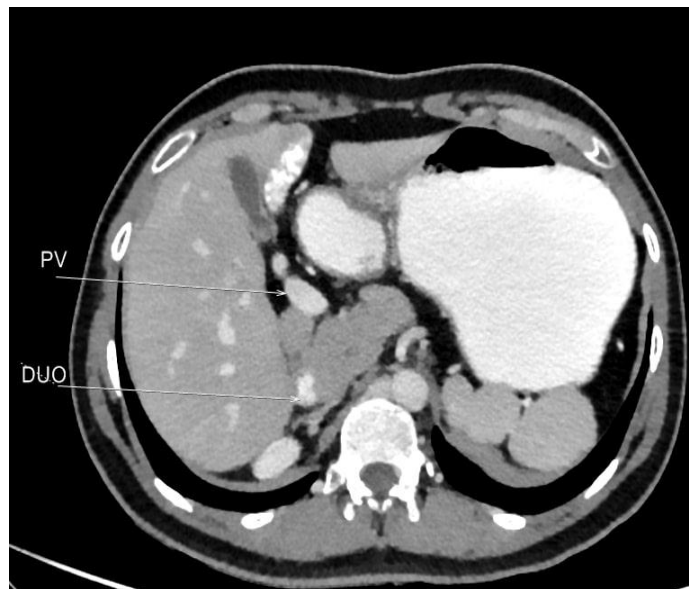


Figure 8: Preduodenal portal vein. PV-portal vein, DUO-duodenum.



Figure 9: Short pancreas P-pancreas.

DISCUSSION

Polysplenia syndrome is commonly associated with cardiopulmonary, gastrointestinal, genitourinary and central nervous system anomalies.^[3] Anomalies related to polysplenia are important from the embryogenesis point of view. Since splenic anomalies like splenic agenesis, hypogenesis and polysplenia, are well-known to be related to several inborn anomalies of other visceral organs, the spleen is considered to play a vital role within the normal development and localization of visceral organs.^[2] The current patient had polysplenia, a short pancreas, a PPV, intestinal malrotation and azygous continuation of the IVC. Though number of these anomalies are seen in association with polysplenia syndrome, the mix of these varied visceral anomalies as well as short pancreas, PPV and azygous continuation of the IVC as seen in this case has not been documented earlier. Specially, short pancreas and PPV are uncommon associated anomalies. Most of the people with this syndrome have symptoms in childhood as a result of associated cardiac anomalies. Other anomalies could also be noted incidentally in adulthood as a result of them not showing any pathognomonic symptoms. The association between abdominal pain and polysplenia syndrome in this patient isn't clear, however malrotation of the intestine could be a factor.

The association of polysplenia with inborn short pancreas was 1st reported in 1984 by Hatayama and Wells. Only few case reports of a short pancreas in polysplenia syndrome have been reported thereafter. The current patient showed a short pancreas. Pathologic conditions including chronic pancreatitis, ischemic atrophy of the pancreatic tail, kwashiorkor and generalized viral infection are also known to cause short pancreas.^[2] None of those pathologic conditions were found in this patient. The explanation for inborn short pancreas has been attributed to be agenesis of the dorsal duct gland. Tanaka et al reported that endoscopic retrograde cholangiopancreatography (ERCP) reveals the presence of accessory pancreatic duct in two-thirds of patients with short pancreas, suggesting that short pancreas may be caused by hypoplasia rather than complete agenesis of the dorsal pancreas.^[4] While most patients with short pancreas in normal situs had diabetes mellitus occasionally accompanied by pancreatitis, none of the patients with short pancreas in polysplenia syndrome had diabetes mellitus and pancreatitis. Therefore, the embryogenesis of short pancreas in polysplenia syndrome could be totally different from that of typical inborn short pancreas. Therefore, it's important for the doctor doing ERCP to bear in mind the presence of inborn short pancreas beside pathologic conditions like pancreatic cancer and pancreatitis, once abrupt cut-off or short pancreatic duct is noted.

PPV is a rare anomaly where the portal vein passes anteriorly to the duodenum. PPV is usually found together with numerous inborn anomalies like visceral malrotation, situs inversus, duodenal atresia, annular

pancreas, biliary abnormality and splenic anomalies.^[5] Clinically, PPV is an anomaly of considerable surgical importance. It's unlikely that PPV itself might cause small intestine obstruction as a result of it's a thin wall and slow flow. The indication for surgery is commonly due to other anomalies like obstruction caused by malrotation, annular pancreas, duodenal web or biliary atresia. The presence of PPV causes technical difficulties at the time of abdominal surgery. Failure to acknowledge PPV throughout operation simply ends up in serious complications like thrombosis or intraoperative bleeding. Correct operative identification of PPV is important to avoid potential hazards throughout operation.

Azygous continuation of the IVC is a common vascular anomaly in association with polysplenia syndrome. Helpful signs of this IVC anomaly on chest film are convexity within the right tracheobronchial angle on the posteroanterior view and an absence of the IVC shadow on the lateral view. On CT examination, the enlarged azygous vein might mimic retroperitoneal lymph node or paravertebral mass. However, the azygos vein can be easily identified as a tubular structure with same degree of intense contrast enhancement as other vascular structure and the continuity with the SVC through azygous arch.

CONCLUSION

Polysplenia syndrome is associated with a wide spectrum of abnormalities. A CT-scan with multiplanar reconstruction is useful in visualizing these anomalies. As the wide spectrum of anomalies in polysplenia syndrome don't have pathognomonic radiologic findings, the information of varied radiologic features plays a crucial role within the identification of this rare syndrome. Additionally, correct identification might decrease the mortality and morbidity. Low degree of suspicion of this syndrome results in detailed investigations as well as CT angiography and even unnecessary thoracotomy due to its associated uncommon anomalies. Surgeons need to be aware with the spectrum to avoid complications during abdominal surgery.

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