Preduodenal Portal Vein And Polysplenia: A Case Report and Review Of Literature

Saudia M. McCarley
*Reading Hospital - Tower Health*

Anas Qatanani
*Drexel University College of Medicine*

Jaclyn Malat
*Philadelphia College of Osteopathic Medicine*

P. Kurt Bamberger
*Reading Hospital - Tower Health*

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Keywords
Preduodenal, portal vein, polysplenia, anterior portal vein, multiple spleens
ABSTRACT

Preduodenal portal vein (PDPV) is a rare anomaly in which the portal vein courses anterior to the second part of the duodenum. PDPV is often associated with other congenital anomalies such as polysplenia, malrotation and pancreatic anomalies. We report an elderly male with Stage IIB esophageal adenocarcinoma. Incidental findings on staging computed tomography (CT) included an anomalous preduodenal and presplenic portal vein and polysplenia, though the patient did not present with any symptoms related to these findings. Post-neoadjuvant chemoradiation, the patient underwent an Ivor Lewis esophagectomy. Appreciating the anomalous tract of the portal vein anterior to the pancreas and duodenum from the preoperative images, we were able to perform duodenal mobilization (Kocher manuever) without vessel injury. Since, PDPV is often an asymptomatic and incidental finding in adults, serious surgical complications may occur if not appreciated on preoperative imaging or intraoperatively. Heightened awareness of PDPV and other associated anomalies remains vital to a safe procedure in all ages.

BACKGROUND

PDPV is a rare congenital vascular disorder in which the portal vein courses anterior, rather than posterior, to the second part of the duodenum. PDPV is thought to arise from abnormal involution of the fetal vitelline veins. PDPV is best diagnosed with delayed contrast CT. While PDPV may be an isolated anomaly, it is often associated with congenital cardiac and gastrointestinal anomalies, such as situs inversus, atrial isomerism, bowel malrotation, polysplenia, annular pancreas, and anomalies of the inferior vena cava (IVC) and azygous vein. Up to two-thirds of cases are diagnosed in the first week of life due to symptoms of intestinal obstruction. Neonates present with refractory bilious vomiting and feeding intolerance, and abdominal imaging reveals intestinal obstruction secondary to gastrointestinal anomalies. PDPV is found in 4% of all cases of duodenal obstruction, which is likely caused by anomalies associated with PDPV rather than isolated PDPV. In adults, PDPV is usually asymptomatic and is often diagnosed incidentally on routine or pre-operative imaging or intra-operatively. When complications occur, treatment involves a duodenoduodenostomy. In most cases diagnosed in adulthood, PDPV does not warrant treatment or surgical intervention.

We present the case of an elderly man whose presentation and management provides insight into the proper surgical preparation for patients with PDPV, particularly when excising masses in the lower thoracic or upper abdominal cavity.

Correspondence to Saudia McCarley
saudia.mccarley@towerhealth.org

Disclosure Statement: The authors have no conflicts of interest to declare.
An elderly man presented with esophageal adenocarcinoma Stage IIb, cT2N0M0, which was diagnosed on Esophagastroduodenoscopy (EGD). He initially declined surgical intervention and was treated with neoadjuvant chemotherapy and was without evidence of malignancy on repeat EGD. Surveillance EGD six months later revealed recurrence of the poorly differentiated esophageal adenocarcinoma near the gastroesophageal (GE) junction. A positive emission tomography (PET) scan revealed distal esophageal mucosal thickening without enlarged lymph nodes. CT prior to surgery revealed notable findings, which were the presence of an anomalous preduodenal and presplenic portal vein, polysplenia with three lobes, and an enlarged azygous vein. The patient was scheduled for diagnostic laparoscopy, pyloromyotomy, Ivor-Lewis esophagectomy, and esophagogastric anastomosis. Intraoperatively during the Kocher maneuver (duodenal mobilization), the portal vein was appreciated tracking anterior to both the pancreas and duodenum. Due to preoperative image review and close attention to detail during the procedure, the Kocher maneuver was performed without inadvertent vessel injury. The procedure was completed without any complications, and the patient recovered well. Final pathology revealed ypT1bN0M0.

First described by Knight in 1921, PDPV is a rare anomaly with fewer than 100 cases reported in the literature.7 PDPV is usually diagnosed in the neonatal period due to evident symptomatology secondary to PDPV and its associated congenital anomalies. Most cases in the literature document pediatric neonatal patients undergoing surgical intervention for complications, most often duodenal obstruction.1,2,9 Reports of PDPV in adults are sparse, as PDPV is rarely symptomatic and therefore infrequently diagnosed. However, duodenal obstruction secondary to PDPV in adult patients has been reported.8 Although PDPV is especially rare in adults, adequate awareness is warranted to avoid potential surgical complications. If PDPV is not noted on pre-procedural imaging, the presence of several associated anomalies should heighten suspicion of its presence.10 Intra-abdominal anomalies, particularly polysplenia, malrotation, and anomalies of the pancreas, IVC, and azygous veins, and cardiac anomalies, like situs inversus, dextrocardia, and atrial isomerism, may indicate a possible PDPV. With knowledge of these anatomical associations, intraoperative injury can be avoided.

Preoperative consideration is particularly important in asymptomatic adults with PDPV undergoing surgical intervention on gastrointestinal structures. PDPV poses a risk of damage to the biliary tract, duodenum, azygous vein, and surrounding structures. In particular, caution should be taken with interventions involving the biliary tract because a PDPV must be bypassed to access the ampulla of Vater. Inadvertent damage to the portal vein may cause serious surgical complications,
including intraoperative hemorrhage and associated postsurgical complications.\textsuperscript{11}

Our patient was diagnosed pre-operatively via Computed Tomography (CT) scan with polysplenia with three distinct portions, which raised suspicion for PDPV that was found on imaging. Additional findings included an enlarged azygous vein on imaging, which was confirmed intra-operatively. While duodenal obstruction secondary to PDPV is often treated with duodenoduodenostomy, our patient was not symptomatic and was undergoing radical resection of the lower esophagus and gastric cardia. An intraoperative assessment was taken to determine if intervention was warranted. Due to the asymptomatic nature of the patient's PDPV, intervention was not undertaken.

**CONCLUSION**

PDPV is a rare anomaly in which the portal vein courses anterior to the duodenum. Although PDPV is most often asymptomatic in adults, we suggest searching for PDPV pre-operatively or intraoperatively if associated cardiac or gastrointestinal anomalies are discovered. Operative complications can be prevented through adequate awareness of PDPV, and alternations in surgical approaches may be warranted.

**REFERENCES:**


