Polysplenia syndrome with preduodenal portal vein

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Polysplenia syndrome is a heterogeneous disease that primarily affects the asymmetric organs, including the heart, lungs and bronchi, liver, intestines, and spleen [1]. It manifests mainly in childhood, 40% of the patients reach 2 years of age and the majority dies before 5 years of the age due to cardiac anomalies [2]. 5-10% of the patients lack cardiac involvement, which allows them to reach adulthood [3]. The precise etiology of polysplenia is unknown. Embryonic, genetic and teratogenic components have all been implicated as causative factors in polysplenia [4]. Although polysplenia syndrome has a wide range of abnormalities, there is no single pathognomic abnormality that characterizes this rare entity. The range of anomalies include multiple spleens of equal volume, visceral heterotaxia, right-sided stomach, a left-sided or large midline liver, malrotation of the intestine, a short pancreas, preduodenal portal vein and inferior vena cava anomalies [5].

We report a 50-year-old female presented to our outpatient department with chief complaints of right upper quadrant pain for the last 3 months. At a local hospital, she was noted to have polysplenia and cholelithiasis. She was referred to our institution for further evaluation and treatment. An abdominal computed tomography (CT) and ultrasonography showed cholelithiasis with polysplenia, the portal vein was located anterior to the duodenum and there was associated malrotation of gut (Fig. 1A). The surgical procedure for cholelithiasis began with a thorough exploration of the abdomen. The portal vein was detected in front of the first part of the duodenum (Fig. 1B). The gallbladder was hugely distended with a 2 cm stone impacted at the neck. The common bile duct was located posterior to the portal vein. The presence of multiple small spleen and one normal size spleen was confirmed on the left side of the upper abdomen. There was malrotation of the gut with the entire right colon located in the left upper quadrant along with left the colon in normal position. The pancreas was short with deficient body and tail. The gallbladder was opened at the fundus, stone removed, cystic artery and duct identified, ligated and cut between ligature and gallbladder removed through liver bed. Ladd’s procedure was added to correct malrotation of gut. The postoperative course was uneventful, and the patient was discharged on 5th postoperative day.

Reports indicate that most cases of preduodenal portal vein (PDPV) in adults involve surgery for cholelithiasis leading to the hypothesis that PDPV may be responsible for the formation of gallstones due to chronic compression of the common bile duct by the portal vein, leading to stasis of bile. A similar opinion was also expressed by Seo et al [6] and Low et al [7]. When surgery is required, care must be exercised, especially for procedures involving the upper abdomen. If PDPV is not detected prior to surgery, it can cause severe complications, such as hemorrhage and vascular ligation [8]. Such accidents
can be prevented by performing careful diagnostic imaging in advance, such as CT, and especially noting the possibility of PDPV in cases of polysplenia syndrome [8].

References