A rare case of polysplenia syndrome presenting with biliary obstruction in adulthood

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A 23-year-old previously well woman presented with right-upper-quadrant pain and obstructive jaundice. Her bilirubin on presentation was 4.85 mg/dL. Magnetic resonance cholangiopancreatography (MRCP) revealed severe central intra-hepatic biliary tree dilatation with a 6mm calculus in the left intrahepatic duct (Fig. 1a, white arrow). There were multiple calculi in the gallbladder (Fig. 1b, white arrow), without evidence of extra-hepatic ductal dilatation. Interestingly, there was evidence of multiple aberrant accessory spleens (Fig. 1a, 1b & 1c, dotted arrows). MRCP also revealed a retrocaval course of the inferior vena cava (Fig. 1a, arrowhead) and partial agenesis of the pancreas with truncation at the level of the proximal pancreatic body (Fig. 1b, arrowhead). This constellation of imaging findings is consistent with a diagnosis of polysplenia syndrome (PSS). Endoscopic retrograde cholangiopancreatography revealed a 1cm high-grade stricture at the bifurcation of the intra-hepatic ducts, and extensive calculi within the biliary system that were unable to be extracted. Trans-thoracic echocardiography showed mild atrial septal redundancy with no intra-atrial shunting. She underwent open cholecystectomy, common bile duct exploration and hepaticojejunostomy with Roux-en-Y reconstruction as definitive management. Intra-operative findings included common hepatic duct stricturing, extensive calculi within the extra-hepatic and intra-hepatic ducts as well as congenital retroperitonealisation of her proximal jejunum.

Polysplenia syndrome is a rare congenital syndrome of situs ambiguous characterized by left isomerism. The syndrome's hallmark feature is the presence of multiple small aberrant splenic nodules, but is also associated with a vast range of abnormalities, including cardiac, vascular, gastrointestinal, biliary, pulmonary and genitourinary malformations. These abnormalities usually result in death or requirement for liver transplantation in early childhood. Of the small proportion of patients that survive into

adulthood, most are diagnosed incidentally on imaging. While biliary atresia has a recognized association with PSS in children, presentation with biliary obstruction in adulthood with previously undiagnosed PSS is extremely rare. To our knowledge, only 5 prior cases have been reported with a similar presentation. These patients usually require definitive surgical management to relieve their obstruction.

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