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Choledochal cyst, polysplenia and situs ambiguous: A rare and new association

Francesca Destro [a,*], Luciano Maestri [a], Marcello Napolitano [b], Milena Meroni [a], Andrea Pansini [a], Giovanna Riccipetitoni [a]

[a] Department of Pediatric Surgery, Buzzi Children's Hospital, Via Lodovico Castelvetro, 32, 20154 Milano, Italy
[b] Department of Pediatric Radiology and Neuroradiology, Buzzi Children's Hospital, Via Lodovico Castelvetro, 32, 20154 Milano, Italy

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ABSTRACT

Introduction: Situs ambiguous (SA) is a rare clinical entity that includes a spectrum of abnormalities in which organs and vessels are opposed to their normal arrangement. SA is frequently associated with other malformations such as splenic alterations, biliary atresia and vascular abnormalities. We report two cases of patients with SA, polysplenia and choledochal cyst. This association has never been described to date.

Case reports: Two patients, both males, came to our attention at the ages of 7 and 8 months after ultrasound identification of choledochal cysts. One patient, who had a prenatal suspicion of situs ambiguous and dextrocardia, presented with failure to thrive and increased hepatic enzymes. The other patient was asymptomatic, but prenatal evaluation showed gastric malposition. Post-natal laboratory tests were normal. Preoperative radiological diagnostic work-up showed polysplenia, intestinal anomaly of rotation and fixation, pre-duodenal portal vein in one baby. Both patients underwent surgical correction by cyst excision and Roux-en-Y hepaticojejunostomy. Follow-up (3 months and 3.5 years respectively) was uneventful. Biochemical hepatic parameters normalized soon after surgery.

Discussion: Situs anomalies in children are often associated with other malformations. The most common are splenic and cardiac defects but intestinal malformations and hepatico-biliary abnormalities have been reported too. Above all, the BASM (biliary atresia splenic malformation) syndrome has been extensively examined, obtaining important information on the etiology of the cystic form of biliary atresia.

Conclusions: A careful evaluation of patients with situs ambiguous is important to exclude associated anomalies and to plan the most appropriate surgical approach. The association with choledochal cyst should be taken into account.

1. Introduction

Disturbances of body symmetry may occur as independent anomalies, but they are frequently associated with other affections, typically splenic malformations. In 1993 Davenport introduced the term Biliary Atresia Splenic Malformation Syndrome (BASM) to indicate a congenital cystic clinical variant of biliary atresia with developmental origin and poor prognosis. The syndrome includes an alteration of intra-hepatic biliary ducts, a cystic dilatation of the external biliary tree, splenic malformation (polysplenia, asplenia), situs inversus, absence of inferior vena cava and pre-duodenal portal vein. We report two cases of patients in whom situs ambiguous, polysplenia and pre-duodenal portal vein were associated with a coledochal cyst. This is the first time this association is described, to the best of our knowledge.

2. Case reports

2.1. First case

The first case was the one of a male patient born at 39 weeks of gestational age by elective cesarian section with a birth weight of 3165 g. The boy had a prenatal identification of situs ambiguous and dextrocardia. After birth he was fine and he started

* Corresponding author.
E-mail address: francesca_destro@hotmail.com (F. Destro).

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breastfeeding, after appropriate radiological assessment. Abdominal ultrasounds (US) showed polysplenia and right aorta in front of the inferior vena cava. The upper gastrointestinal series showed midgut malrotation (Fig. 1). Echocardiogram identified an azygous continuation, a right-sided aortic arch, an inter-atrial defect with left-right shunt, a ductus arteriosus (that spontaneously closed in 48 h). He was discharged in healthy conditions and he was fine for the first 18 months of life, when he presented with failure to thrive associated with distended abdomen and hepatomegaly. Laboratory tests showed increased markers of hepatic necrosis and cholestasis; both infective and metabolic analyses were normal. He never had icterus or itches. The US enlightened a dilatation of intra-hepatic biliary ducts, left hepatic lobe hypertrophy, a choledochal cyst (16 × 8 mm oval image) and a pre-duodenal portal vein. A cholangio-MRI confirmed the presence of the choledochal cyst (Fig. 2) that was described as an oval mass at the choledochal duct reached by the cystic duct. The gallbladder was in median position. The pancreas was stubby without ductal dilatations (Fig. 3). Polisplenia and pre-duodenal portal vein were also evident (Fig. 4). The boy underwent surgery: we performed a cholangiography together with hepatic biopsy and choledochal cyst removal with Roux-en-Y loop to restore the intestinal continuity and to allow biliary drain. During surgery we found a pre-duodenal portal vein and common hepatic artery, venous ectasia of vascular branches in front of the pylorus, incomplete intestinal fixation, and incomplete anular pancreas. The extra-hepatic biliary tree had a diverticulum in its proximal part. Histological examination showed neither biliary stasis in the hepatic parenchyma nor mineral deposits. The picture was that of an inflammatory reaction to extrahepatic biliary obstruction. The choledochal cyst had epithelium without signs of inflammation or subepithelial cicatrix and scarce mural muscular cells. Ectopic pancreatic tissue was found in the removed diverticulum. Biochemical parameters improved soon after surgery. No complications occurred. Hospital stay lasted 13 days. The boy is doing fine four years after surgery.

2.2. Second case

The second child was a boy, born at 38 weeks of gestational age (birth weight of 3575 g). US pregnancy screening enlightened an abnormal gastric position without evidence of the gallbladder. Radiological investigations performed after birth (thorax x-ray, abdominal US, thoraco-abdominal MRI, upper gastrointestinal tract contrast x-ray and barium enema) showed situs ambiguous with left isomerism characterized by increased liver volume with left gallbladder, right stomach, left duodenum, right colon and polysplenia (Figs. 5 and 6). Intrahepatic biliary ducts were dilated and there was a Type I cholodochal cyst (12 × 20 mm) according to the Todani classification (Fig. 7). The inferior vena cava was on the left side of the aorta. He was asymptomatic and he had normal laboratory tests. At 8 months of age, after completing the radiological, clinical and laboratory assessment, the boy underwent choledochal cyst removal and bilio-digestive anastomosis in a Rou-en-Y fashion. During surgery we identified a pre-duodenal portal vein that was not obstructing the intestine (positive air test) and a mesenterium commune with retromesocolic hernia. The intestinal loops were freed, together with the duodeno-jejunal corner and the Treitz, the colon was placed in the right abdomen and the ileum in the left part. The appendix was removed. The gallbladder and the cystic duct were isolated together with the choledochal cyst that was resected. Hepatic ducts were not dilated. The jejunal loop was prepared 25 cm far from the Treitz (the length was conditioned by the anatomical mesenteric fashion). The distal part was

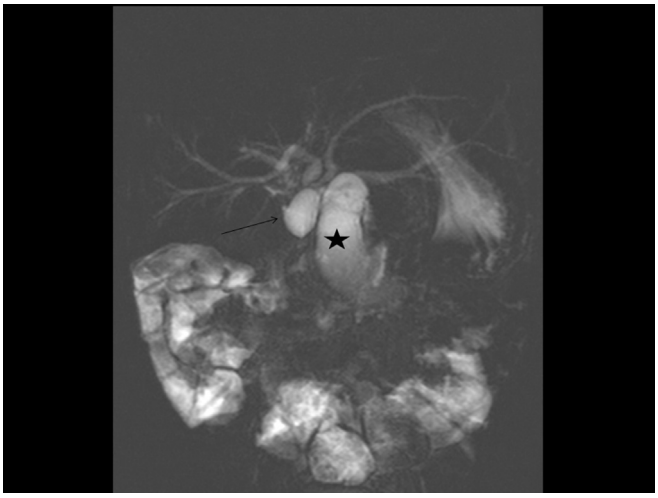

Fig. 2. Magnetic resonance cholangiography shows a choledochal cyst (thin arrow) and dilatation of intrahepatic biliary ducts. Gallbladder is in the midline (black star).

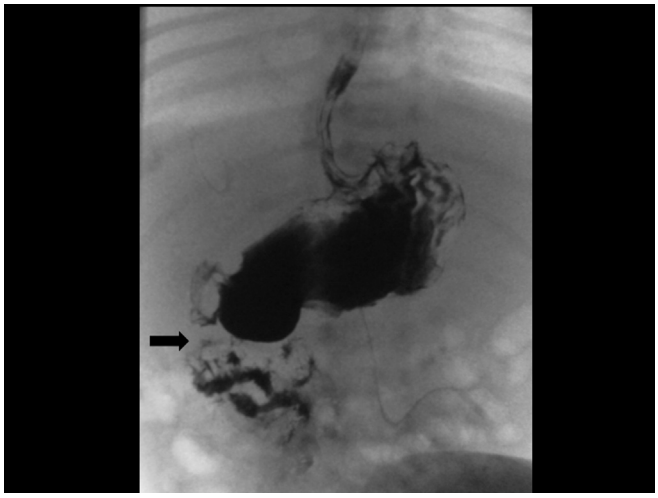

Fig. 1. Upper gastrointestinal series show abnormal position of duodenojejunal junction (thick black arrow) with both duodenum and jejunum on the right side of the vertebral column.

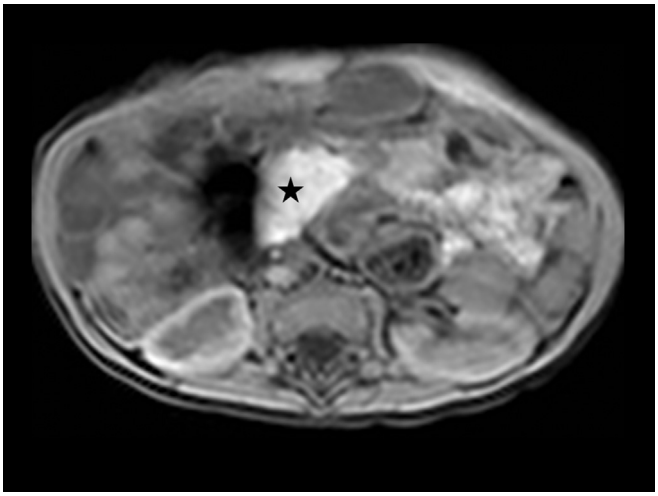

Fig. 3. Axial T1 weighted MR with fat saturation shows congenital short pancreas with only a globular pancreatic head visible (black star).