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Biliary atresia with rare associations: a case report

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Abstract

Background It is not often written in medical journals that preduodenal portal vein, biliary atresia, intestinal malrotation, and situs inversus totalis are all related. This is a rare association.

Case reports A 2-month-old female infant had biliary atresia type III, situs inversus totalis, midgut malrotation, and a preduodenal portal vein. She had been operated on by the Kasai procedure (hepato-portoenterostomy).

Discussion It is important to carefully look into the relationship between the preduodenal portal vein and biliary atresia because the patient is at risk of injury from this aberrant vein during operative intervention.

Conclusion The relationship between biliary atresia and other congenital anomalies like preduodenal portal vein, intestinal malrotation, and situs inversus must be taken into consideration to avoid other diverse effects during surgical intervention.

Keywords Preduodenal portal vein, Portal vein anomalies, Kasai operation, Intestinal malrotation, Situs inversus totalis, Heterotaxia syndrome

Background

Preduodenal portal vein (PDPV) is a rare congenital vascular malformation in which the portal vein's primary trunk runs in front of the duodenum rather than behind it [1]. Biliary atresia (BA) is a blockage of the intrahepatic and extrahepatic biliary tree that gets worse over time and leads to cholestasis and cirrhosis [2].

It is important to carefully look into the relationship between PDPV and BA. PDPV is unintentionally discovered in 5–10% of infants with BA during surgery [3]. The infants are in danger of iatrogenic injury and bleeding from this aberrant vein because of this unusual anatomical position [4]. The Kasai portoenterostomy is the initial surgical treatment of BA for neonates younger than 90 days. The most frequent reason for pediatric liver transplantation worldwide is still BA [4, 5].

PDPV is a rare anomaly; fewer than 100 cases have been documented in the literature. However, very few cases of BA associated with PDPV have been reported [4, 6–10]. Herein, a case combined with BA with PDPV, midgut malrotation, and situs inversus totalis is presented.

Case presentation

A full-term female infant, G1P1, weighing 3450 g at birth, was referred to our department at the age of 2 months old with severe jaundice and poor feeding. The mother reported yellow sclera and skin, lightning-colored stool, and dark urine a few days after birth, which were progressively aggravated. The infant had not received any drugs or medical treatment before, and the parents were relatives.

On physical examination, the infant had an olive-green colored sclera and skin with an itching mark, a pale clay stool in the diaper, a low-grade fever of 37.8 °C, a weight of 3770 g, a pulse rate of 138 b/m, and a respiratory rate of 38 b/m. The heart sounds and breath sounds were

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normal. The abdomen was soft and lax on examination. The liver was two fingers below the costal margin. The intestinal sound was normal.

The laboratory investigation revealed that total bilirubin was elevated at 7 mg/dl, direct bilirubin was elevated at 4 mg/dl, gamma-glutamyl transferase (GGT) was elevated at 157.1 U/L, alkaline phosphatase was elevated at 279 U/L, and albumin was decreased at 3.4 g/dL.

Abdominal ultrasonography revealed a triangular cord sign in the liver hilum, an absent gall bladder, and a normal-sized spleen on the right side behind the liver. A chest X-ray revealed dextrocardia. The HIDA scan revealed a failure of radioisotope excretion in the duodenum. Non-contrast magnetic resonance cholangiopancreatography (MRCP) showed that the common hepatic duct and the common bile duct could not be seen. A preoperative percutaneous liver biopsy reported liver fibrosis, bile ductule proliferation, and cholestasis.

An experienced pediatric surgeon obtained the decision for exploratory laparotomy by right subcostal abdominal incision. The spleen is located in the right upper quadrant below the liver (Fig. 1). Extracorporealization of the liver is done (Fig. 2). The gall bladder was rudimentary. On further exploration, we also found intestinal malrotation (IM) and PDPV. All extrahepatic bile ducts were absent; type III portal atresia. The portal plate was dissected easily, and the rudimentary gall bladder was removed (Fig. 3).

Widening of the narrow base of the intestinal mesentery, then reconstruction of the retro-colic Roux-en-Y limb of the jejunum, and finally hepato-portoentostomy (Kasai operation) were done. A wedge liver biopsy obtained reported ductular proliferation, hepatic fibrosis, and bile plugs; a feature suggestive of biliary obstruction.

The post-operative follow-up of the patient in the pediatric ICU revealed features of bile drainage in the intestine; the stool returned to its normal brown color; and the yellow skin color was slightly improved. Total bilirubin was 4 mg/dl, and direct bilirubin was 2 mg/dl. The patient was discharged on the tenth postoperative day.

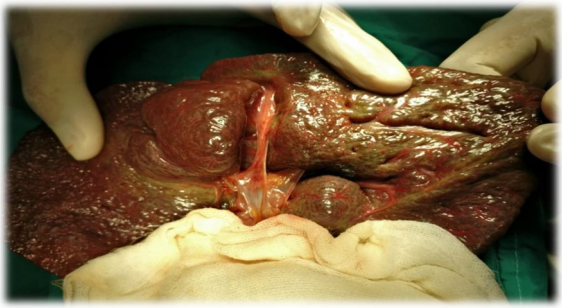

Fig. 2 Our policy is to extra-corporealize the liver

Unfortunately, 3 months later, the patient developed the manifestations of liver cell failure and ascites. The patient died in the intensive care unit.

Discussion

The incidence of BA varies from 1/15,000 up to 1/19,000. Females had a slightly higher prevalence of BA than males [2, 5, 11]. Untreated infants develop rapidly progressing fibrosis and cirrhosis, resulting in portal hypertension, end-stage liver disease, and death within the first years of life [5]. Only 10% of newborns with BA have additional congenital anomalies such as PDPV, situs inversus, midgut malrotation, and cardiovascular deformities, or it might be a symptom of polysplenia syndrome or heterotaxia syndrome. Ninety percent of cases have an isolated BA [5, 12].

The first report of BA connected to PDPV was provided by Pernkopf, who performed an autopsy on a new infant [6]. PDPV originates from the persistent primordial yolk vein as a result of the faulty formation of the portal vein from the fetal vitelline veins and is related to

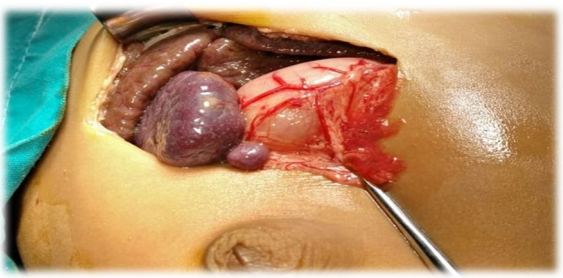

Fig. 1 A surprise is the presence of the spleen in the right upper quadrant below the liver

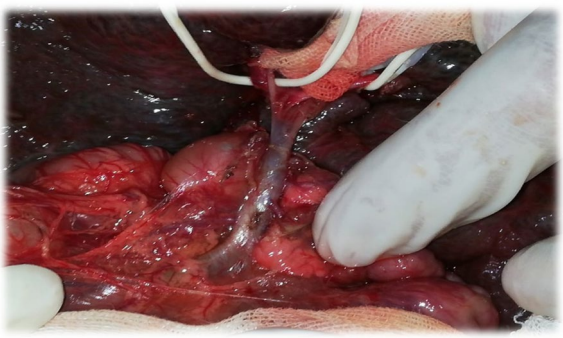

Fig. 3 The portal plate was dissected without extra effort than usual. It is suspended in white rubber in the porta-hepatis region, below it is the pre-duodenal portal vein

abnormal midgut malrotation. Because PDPV sometimes goes unnoticed, it is impossible to know its true incidence [1, 4]. PDPV in our case was asymptomatic and did not cause duodenal obstruction, so there was no need for bypass surgery.

The term "situs inversus totalis" describes a condition in which the viscera in the thoracic and abdominal cavities are transposed in a mirror image form, including dextrocardia [13]. This condition affects 1 in 10,000 people, making it extremely rare. A busy surgeon might only expect to see something like this once or twice in their whole life [14]. So, the surgeon must take care when handling this condition. Our case is considered part of the heterotaxia syndrome because both the liver and spleen are present on the right side in the presence of IM. Kouwenberg et al. (2008) report five cases with PDPV associated with IM; however, one case only had heterotaxia syndrome (situs inversus, PDPV, IM, and severe cardiac abnormalities; no spleen could be identified during surgery) [15].

According to the Kasai classification, it is type III [11]. Magnetic resonance cholangiography (MRCG) in conjunction with a hepatobiliary iminodiacetic acid (HIDA) scan are two non-invasive methods for diagnosing BA [16, 17]. MRCG has a sensitivity of 100% and a specificity of 96% [15]. HIDA scan has a sensitivity of 98.7% and a specificity of 70.4% for the diagnosis of BA.14 ERCP is still an uncommon procedure in children [18].s

When carried out prior to the patient's 60th day, the Kasai procedure (portoenterostomy) can save the patient's life, but liver transplantation is strongly recommended during the first year of life [2, 4–6, 12, 18]. Many authors hypothesize that the syndromic form of biliary atresia is associated with poor success of the Kasai portoenterostomy procedure, and thus it requires liver transplantation in infancy [19–21]. Liver transplantation is a challenge in this case because of the small size of the infant, the presence of associated anomalies, and PDPV. This infant developed a manifestation of liver cell failure and died. We believe that the Kasai operation does not eliminate the need for liver transplantation in BA patients but rather gives them more time to find a liver donor. Francesco et al. (2019) described two patients who benefited from PDPV reconstruction that facilitated the graft implantation, but in two older infants 9.5 and 13 months old and whose weights were 4.3 and 9.2 kg.

Conclusion

Although it is unknown how these anatomical relationships would affect an infant's prognosis for biliary atresia, recognizing this abnormal anatomy is crucial both for the first exploration of hepato-portoenterostomy and for liver transplantation.

Abbreviations

PDPV	Preduodenal portal vein
BA	Biliary atresia
MRCG	Magnetic resonance cholangiography
HIDA	Hepatobiliary iminodiacetic acid
IM	Intestinal malrotation

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None.

Authors' contributions
SMA contributed to writing the manuscript and was the operative assistant. TAS was the main surgeon, the pioneer of the case, and the reviewer. BA was a reviewer. All authors read and approved the final manuscript.

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Declarations

Ethics approval and consent to participant
Approved.

Consent for publication
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Competing interests
The authors declare that they have no competing interests.

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