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Case Report

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The Congenital Absence of the Portal Vein

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ABSTRACT

Congenital absence of the portal vein (CAPV) is a rare defect often accompanied by other abnormalities such as heart defect, skeletal anomalies and / or liver tumors [1].

We describe here a case of CAPV revealed by an upper gastrointestinal tract hemorrhage in a child aged 02 years and 8 months. The esogatro-duodenal fibroscopy revealed the presence of Esophageal varices grade II, abdominal ultrasound revealed a large liver with no visualization of the spleen. Subsequent abdominal computed tomography revealed the presence of a spleno-mesaraic trunk measuring 7 mm in anteroposterior diameter receiving the inferior and superior mesenteric vein and a small splenic vein draining by dilated peri-gastric and peri-esophageal leads. Associated with splenic hypoplasia and portal trunk atresia. The rest of the malformative assessment was negative.

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Introduction

Congenital absence of the portal vein (CAPV) is a congenital anomaly in which the intestinal and splenic venous drainage bypasses the liver and drains into the inferior vena cava (IVC), renal vein, or iliac vein through a congenital portosystemic shunt [2]. CAPV can be diagnosed without invasive techniques. It has often been associated with other congenital abnormalities, such as cardiac defects, biliary atresia, and polysplenia . At present, there is no explanation for these associated cardiac and skeletal malformations [1].

Case Report

A boy child aged 02 years and 8 months, admitted to the pediatric department for moderate melena and Hematemesis. The child is from a consanguineous marriage, he had a history of a similar episode a year ago, no fever or transit disorder. His past medical health, including birth history, immunizations, and other medical problems is unremarkable. The child's vaccination was appropriate for his age.

On initial examination, the weight was 17 kg and the height 90cm; weight-for-age was between 50 and 85th percentile, height-for-age was between 5th and 15th percentile, these anthropometric results show stunted growth.

On admission, the child was alert, tired and slightly pale appearing, heart rate was 120 beats per minute, respiratory rate 32 cycles per minute, and blood pressure 100/640 mmhg. Cardiovascular examination was normal, percussion and pulmonary auscultation were without abnormalities. His mucous membranes were dry and pale. The abdominal examination was without visceromegaly or ascites. His skin shows no lesions, bruises or petechiae. Upper and lower extremities were normal. His neurological exam was normal.

The complete blood count showed hemoglobin at 10.5 g / dl, MCV at 71.6 fl, TCMH at 23.9 pg and platelets at 289.10'3 / mm3. The prothrombin level was 90%, and hepatic and renal tests were normal.

Upper gastrointestinal endoscopy revealed the presence of esophageal varices grade II. Abdominal ultrasound (US) showed a large liver with no visualization of the spleen. Abdominal computed tomography revealed the presence of a spleno-mesaraic trunk measuring 7 mm in anteroposterior diameter receiving the inferior and superior mesenteric vein and a small splenic vein draining by associated dilated peri-gastric and peri-oesophageal dilated associated to splenic hypoplasia and portal atresia. The cardiac ultrasound was without abnormality.

Therapeutic management consisted of endoscopic band ligation to treat esophageal varices with medical supervising [3].

Discussion

We now discuss different possible clinical presentations and congenital anomalies associated with Abernethy syndrome, diagnostic workup and treatment options.

Abernethy's malformation, also called a congenital portosystemic shunt, is defined by the congenital absence of the portal vein, results from the malformation of the splanchnic venous system. Congenital portosystemic shunts are divided into extra and

intrahepatic shunts [2, 4]. Two shunts were defined: Type I is characterized by the complete diversion of portal blood into the vena cava with associated congenital absence of the portal vein. Type II is defined as an intact but deviated portal vein via a lateral extrahepatic connection to the vena cava.

Congenital intrahepatic portosystemic shunts may be present during the neonatal period with growth retardation, galactosemia, neonatal cholestasis and hepatic encephalopathy. Many patients are diagnosed with congenital malformations associated with heart defects, which are present in up to 60% of patients, spinal abnormalities or liver tumors. Secondary complications such as hypoglycemia, hyperammonemia, encephalopathy and heart failure may be transient and may resolve spontaneously [5]. Other patients may be asymptomatic throughout their lives.

In the long term, other lesions such as nodular regenerative hyperplasia, partial nodular transformation, hepatoblastoma, hepatocellular carcinoma and adenoma may appear [6]. The two possible explanations for the development of these lesions are: a) the diversion of hepatotrophic substances like insulin and glucagon away from the liver leading to impaired development, function and regenerative capacity of the liver and b) increased blood loss. hepatic arterial flow [7].

Our patient has no evidence of hepatic neoplasia (Alpha-fetoprotein was normal) during his last hospitalization, however, given the increased risk of hepatocellular carcinoma and adenoma, we advised regular radiology and serological monitoring every six months.

Conclusion

Congenital absence of the portal vein might be more common than previously thought. Although the clinical presentations of CAPV are various, the imaging characteristics of CAPV show the absent portal vein and the portosystemic shunt. CAPV can be diagnosed without angiography (1). Regardless of the clinical presentation, it is important to first detect the absence of the portal vein by US and then proceed to further diagnostic examination.

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