Introduction

Congenital absence of portal vein with a complete loss of liver portal perfusion leads to developmental and functional alterations that predispose liver to focal or diffuse hyperplastic or dysplastic changes, also there is increased incidence of hepatic tumors or tumor like conditions in these patients.

This poster presents 2 cases of congenital absence of portal vein in very young ages and with some degrees of liver failure. Both patients are in liver transplant list. Due to complex vascular anatomy they need to have a whole graft.

Case report #1

Case # 1: is a 4 year old boy with portal vein agenesis, regenerative nodular hyperplasia of the liver, and persistent low grade cholestasis. MRV of the abdomen showed abnormal draining of superior mesenteric vein to the IVC, just inferior to the IVC/right atrium junction. The splenic vein could not be clearly seen in this patient. The liver nodules are better seen in the ultrasound exam.

Case report #2

Case # 2: a 2 year old girl with pulmonary hypertension, bilateral pulmonary arteriovenous malformation and malrotation. She presented with feeding intolerance and emesis. MRV of the abdomen showed abnormal drainage of portal vein and superior mesenteric vein with an abnormal course draining to the left sided IVC. There was IVC Azygus vein continuation. The splenic vein joins the SMV to drain into left sided IVC.

Discussion

Once considered a rare event, with advances in imaging techniques the number of detected cases of congenital absence of portal vein (CAPV) are increasing.

The first case of an absent portal vein with a congenital mesentero-caval shunt was discovered by John Abernethy in an autopsy of a 10 month old female (1).

Later CAPV was classified in type 1 and type 2 (2) and also type 1 was subclassified as type 1a and 1b(3), (table 1).

Case report #1: MRA of the abdomen shows a prominent size of hepatic artery as the only source of blood supply of the liver.

Case report #2: MRV of the abdomen shows abnormal drainage of superior mesenteric vein to the IVC, just inferior to the IVC/right atrium junction.

Case report #2: Coronal CT scan image of the chest, shows Azygus continuation of the IVC.

References


Classification of CAPV

| Type 1a | The splenic vein and superior mesenteric vein drain separately. |
| Type 1b | The splenic vein and superior mesenteric vein drain together forming a common trunk. |
| Type 2 | Part of the mesenteric blood drains into the systemic circulation through a partial shunt. |