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CASE REPORT

Abernethy Malformation: A Rare Case Report

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Abstract

Abernethy malformation also known as congenital extrahepatic portosystemic shunt(CEPS) are rare (1 in 50,000 people) vascular anomalies of the splanchnic venous system and results from persistence of embryonic vessels. We present a rare case of abernethy malformation in a 4 year old girl. First case was reported by John Abernethy in 1793 and till now 83 cases have been reported till now.

Key words

Abernethy Malformation, Congenital Extrahepatic Portosystemic Shunt(CEPS), Vascular Anomalies

Inroduction

Abernethy malformation is a congenital extra hepatic portosystemic shunt (CEPS) that develops between the porto-mesenteric vasculature and the systemic veins. [1]

The basic abnormality is a vascular aberration in which the splanchnic venous return drains directly into the systemic veins (IVC or the left renal, left iliac or left hepatic vein), diverting the mesenteric circulation away from the liver. The portal vein is either hypoplastic or completely by-passes the liver.

The portosystemic shunt disrupts the enterohepatic circulation with deranged metabolism of various substances, leading to adverse clinical manifestations. The clinical presentation varies from hypergalactosemia to hyperbilirubinemia to hyperammonimia due to delayed hepatic metabolism of these metabolites as they bypass the liver in the first instance.

Pulmonary venous congestion results in hepatopulmonary syndrome, presenting clinically with dyspnea due to pulmonary hypertension. Hepatic encephalopathy may develop in longstanding cases, clinically manifesting with tremors, extrapyramidal symptoms, irritability and altered sensorium. Regenerative nodular hyperplasia of the liver can also result from the liver's abnormal response to the absent portal flow and can progress to a hepatic tumor in the form of adenoma, hepatoblastoma or hepatocellular carcinoma.

Case Report

A four year old girl presented to **Pediatric** OPD of NIMS Jaipur with history of fever, limping due to flexion deformity at left hip joint from past 10 days. Blood work up showed increased ESR (110), CRP(185) and low Hb (6gm). On further workup ultrasonography showed normal size of liver and spleen, loculated heterogenous collection of approx. 31*91*51 mm(AP*CC*Tr) in the left para colic gutter, perisplenic and perinephric region was also noted which showed communication with the left psoas muscle and was seen extending to the left upper thigh muscles.

An abnormal communication between the extrahepatic portal vein and IVC was incidentally noted on color Doppler sonography. The left and right branches of the portal vein was not visualized.

Furthermore, CT Angiography was advised.

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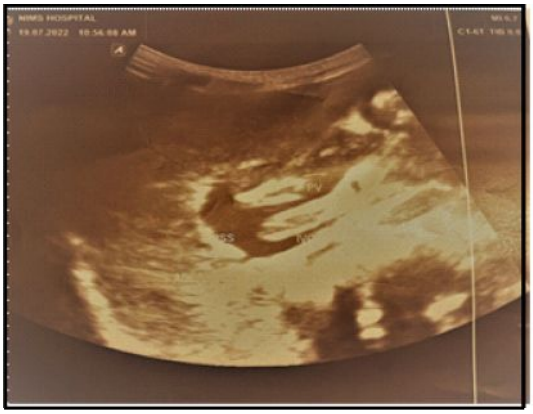

Fig 1 : USG Image showing a shunt between main portal vein and IVC

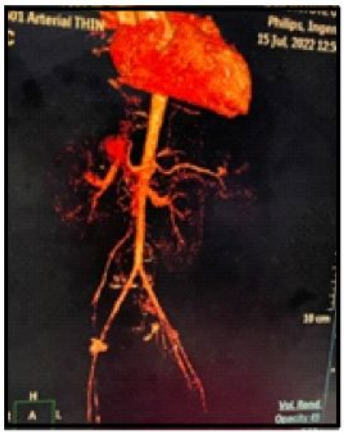

Fig 3 : 3D Image

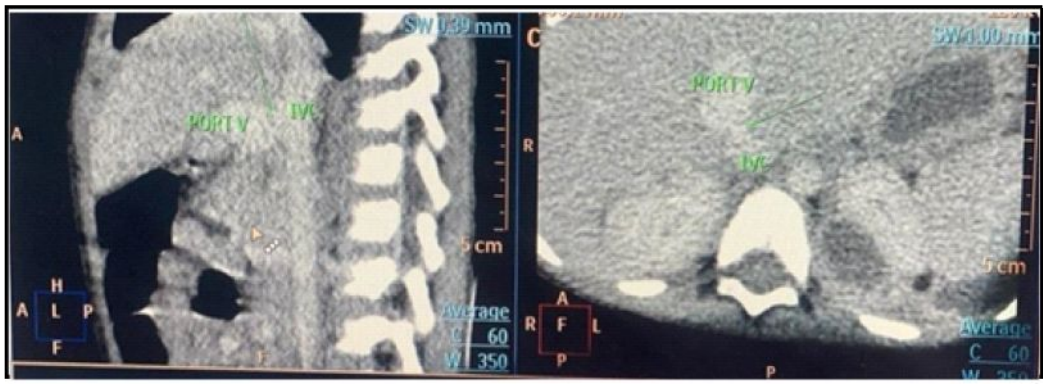

Fig 2 : CT Angiography sagittal and axial images depicts the shunt between the main portal vein and the IVC, also pruning of intra hepatic portal branches

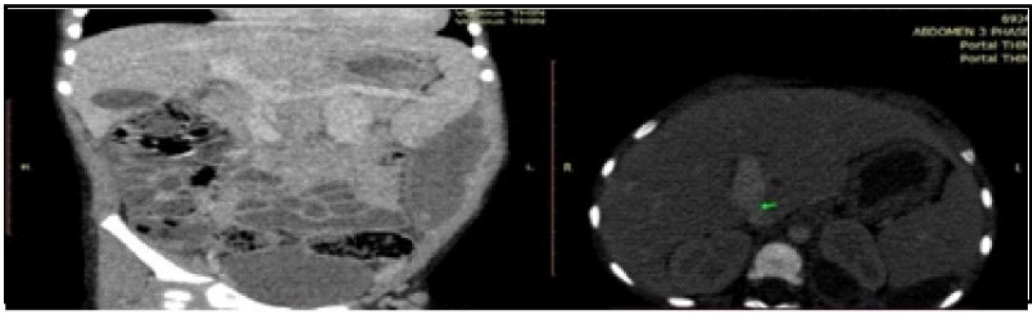

Fig. 4 : Coronal and Axial Ct Angiography Images showing Abnormal Communication Between main Portal Vein And IVC.

On CT Angiography, the splenic and superior mesenteric vein joins to form portal vein, appears prominent measuring approx 12 mm near the IVC , and at this region portal vein and IVC appears to communicate with pruning of intrahepatic portal branches suggestive of **Type II** abernethy malformation.

Discussion

Portal vein anomalies should be included in the differential diagnosis of pulmonary hypertension or pulmonary arteriovenous malformations. Based on careful assessment of the anatomy and testing of portal vein, hemodynamics interventional therapy of complex Abernathy malformations can be performed successfully in specialized centers.

Embryology

The development of the portovenous system occurs in close relation with the umbilical venous system. There are 2 vitelline veins, which arise from the yolk sac, travel along thethird part of the duodenum, and drain into primitive hepatic sinusoids. These vitelline veins are

connected by 3 transverse communicating channels located at the level of transverse hepatic fissure (cranial most), dorsal (middle), and ventral (caudal most) to the duodenum. During the process of embryonic development, there occurs complex intricate involution of these transverse communicating channels resulting in the fully developed adult portal system.[2]

If the vitelline veins fail to establish anastomosis with hepatic sinusoids, type I Abernethy malformation results. The persistent right vitelline vein results in an abnormal shunt between the PV and the retrohepatic inferior vena cava (IVC).Similarly, the persistence of the left vitelline vein leads to an abnormal shunt between the PV and the suprahepatic IVC or right atrium.There is another network of cardinal veins in the proximity ofthe PV which is the precursor of IVC. In the embryonic stage, there are multiple anastomotic channels between vitelline and subcardinal veins. These anastomotic channels undergo involution with fetal development, and any abnormal persistence of these channels results in type II Abernethy malformation.[2,3]

Classification

The most commonly used clinical classification was proposed by Morgan and Superina. It divides the Abernethy malformation into 2 types: type I and type II. [11] Type I refers to the total aplasia of intrahepatic portal venousbranches with complete extra hepatic shunting of portovenous blood into systemic veins. Type I is further divided into type Ia (SMV and SV drain separately into a systemic vein) and type Ib (SMV and SV unite to form a common vein before draining into a systemic vein).

Type II Abernethy refers to hypoplastic intrahepatic portal venous branches with partial extrahepatic shunting of portovenous blood into a systemic vein.[4,5] Different types of Abernethy malformation. Type 1 represents end-to-side shunting of PV into IVC bypassing the liver. It is further classified into type 1a (the SV and SMV drain separately) and 1b (both veins drain together after forming a common trunk).[6-8] Intrahepatic portal venous branches are absent. Type 2 represents side-to-side anastomosis of PV and systemic vein. Intrahepatic portal venous branches are presentbut hypoplastic. PV, portal vein; SV, splenic vein; SMV, superior mesenteric vein; HV, hepatic veins; IVC, inferior vena cava. The presence or absence of intrahepatic portal vein branches forms the basis of this classification.[9-10]

Conclusion

Our case ,4 year old girl with bilateral psoas abscess and abnormal portal venous communication was concluded to be type 2 abernethy malformation. Abernethy malformation is a rare anomaly with multiple clinical associations. Most often, children present with dyspnea, encephalopathy, and abdominal complaints. The purpose of imaging is to identify and classify the shunt, with identification of accompanying anomalies. It is essential to distinguish this entity from intrahepatic shunts and acquired extrahepatic shunts. Careful monitoring is recommended if the patients are asymptomatic or have mild metabolic abnormalities. Any complication warrants appropriate therapeutic intervention. Radiologists must be familiar with imaging features of this rare anomaly for early diagnosis and therapeutic guidance, leading to better patient outcome.

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