Case Report

Role of imaging in congenital extrahepatic portosystemic shunt-a rare entity in clinical practice

Athiyappan Kumaresh[1], Rajoo Ramachandran[2*], Sellappan Rajamanickam Babu[3], Subramanian Ilanchezhian[4]

[1,2,3]Assistant Professor, [4]Sr. Resident, Department of Radiology, Sri Ramachandra University, Porur, Chennai-600116, Tamil Nadu, INDIA.
Email: kumaresha@gmail.com, drrajoor@gmail.com, baburad@gmail.com, msilan1984@gmail.com

Abstract

Congenital extra-hepatic porto-systemic shunts are extremely rare vascular malformations and not often reported in English literature. Here, we report two cases of congenital extra hepatic portosystemic shunt and discuss the imaging findings related to this rare condition which aided in further management.

Keywords: Congenital, Extrahepatic portosystemic shunts, Computed tomography.

[*]**Address for Correspondence:**
Dr. Rajoo Ramachandran, Assistant Professor, Department of Radiology, Sri Ramachandra University, Porur, Chennai-600116, Tamil Nadu, INDIA.
Email: drrajoor@gmail.com

Received Date: 21/09/2014 Revised Date: 30/09/2014 Accepted Date: 03/10/2014

Access this article online

Quick Response Code:

Website: www.medpulse.in

DOI: 04 October 2014

INTRODUCTION

Congenital extrahepatic porto systemic shunts (CEPS) are rare vascular anomalies which are either discovered in a symptomatic infant or incidentally seen in a child who undergoes Ultrasonography (US) for other reasons. CEPS (Congenital Extrahepatic Porto systemic) shunt develops because of excessive involution of vitelline veins that embryologically constitute the portal vein.[1] The first case of CEPS was reported in a 10 month old girl by John Abernethy in 1793 who described the postmortem examination that revealed termination of portal vein into the Inferior Vena Cava (IVC) at the level of renal veins.[2] Based on gross anatomy, it has been classified into two types; Type I shows an absent intrahepatic portal vein with shunting of entire portal blood to the systemic circulation. Type II shows an intact portal vein with partial shunting of portal blood into systemic circulation. The role of the radiologist is to classify the type and make an accurate diagnosis which aids in therapeutic management.

CASE PRESENTATION

Case 1

A 4 year old girl came with complaints of bleeding per rectum for 3 months. The bleeding was painless, intermittent and of a small volume, each lasting for 2 to 3 days. No fever or jaundice was seen. The patient had a past history of right vesicoureteric junction reflux for which she underwent ureteric implantation. Colonoscopy and biopsy was done which revealed mild chronic non specific colitis. Initial laboratory investigations showed low hemoglobin. And the rest of the blood investigations like leukocyte - total count, differential count, platelet count and liver function tests were within normal limits. The child was subjected to USG of the abdomen which showed absent portal vein (Figure 1a].The child was advised to undergo CT(Computed Tomography) examination for detailed evaluation of portosystemic circulation. CECT (Contrast Enhanced Computed Tomography) of the abdomen showed an absent portal vein (Figure 1b) and its branches with an abrupt ending of the superior mesenteric vein (SMV) and the splenic vein confluence. A large (~20mm calibre) inferior mesenteric vein (IMV) from the splenic vein was seen extending craniocaudally into the pelvis and draining into the left internal iliac vein. Multiple perirectal collaterals were seen (Figure 2a, 2b and 3). No focal liver lesions were seen. The bowel loops were normal. No other associated anamolies were found. No clinical feature of hepatic encephalopathy was seen and blood ammonia and galactose levels were within normal limits. A radiological diagnosis of congenital absence of portal vein with inferior mesenteric-caval shunt and multiple perirectal

How to site this article: Athiyappan Kumaresh, Rajoo Ramachandran, Sellappan Rajamanickam Babu, Subramanian Ilanchezhian. Role of imaging in congenital extrahepatic portosystemic shunt-a rare entity in clinical practice. *MedPulse – International Medical Journal* October 2014; 1(10): 613-616. http://www.medpulse.in (accessed 10 October 2014).

collaterals was made. Since no features of any complication related to the shunt or no focal liver lesion was seen, the child was given haematinics and was advised regular follow-up. In this case the differentiation between the type I and type II Abernethy malformation can be made only by liver biopsy which the parents refused.

Case 2

A 4 day old female baby presented with cyanosis and tachycardia. Echocardiography showed situs ambigus and atrioventricular septal defect. The child was advised to undergo CT of the chest. MDCT (Multi detector Computed tomography) with cardiac angiogram was done to evaluate for cardiac anamolies and to look for any associated extra cardiac abnormalities. CECT of the chest showed multiple cardiovascular abnormalities like, atrio ventricular septal defect (figure 4a), bilateral hyparterial bronchus (left sided isomerism) (Figure 4b), coarctation of aorta (Figure 4c and bilateral superior vena cava (Figure 4d). The visualized section of the upper abdomen showed a shunt (~6mm calibre) between the splenic vein and the left renal vein causing dilatation of the latter (figures 5a and 5b). The right portal vein was small and hypoplastic. A relative enlargement of the left lobe of liver was seen possibly due to increased blood flow to the left lobe (Figure 6a). Dilated left hemiazygos vein (figure 6b) was seen communicating inferiorly with the dilated left renal vein and superiorly draining into the left SVC. The dilatation was due to the hyper dynamic flow caused by port systemic shunt. The colon was predominantly seen on the left side of the abdomen and the small bowel loops on the right side. The SMV was seen anterior to SMA -features which were suggestive of intestinal mal-rotation. The child was planned for cardiac surgery but unfortunately she succumbed due to cardiac failure. Since the portal vein was present and a part of the portal flow was shunted into the left renal vein this is a case of type II Abernethy malformation. Type II abernethy malformations are usually isolated and association with multiple cardiovascular anamolies is rare as seen in our case.

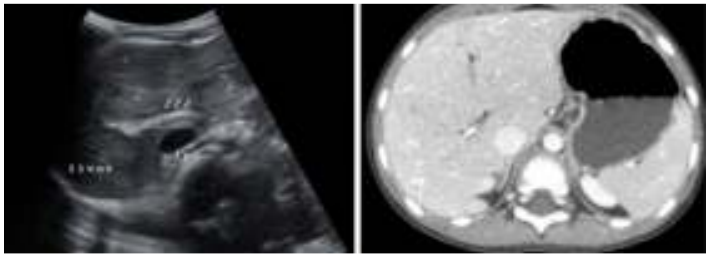

Figure 1a and 1b: Ultrasonography of the abdomen (transverse view at the level of liver) shows absent portal vein (slanted lines) and a echogenic band at the site of portal vein. The corresponding axial CECT - abdomen in venous phase shows absent portal vein

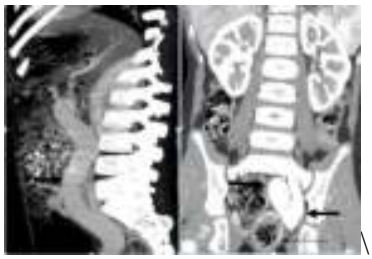

Figure 2a and 2b: CECT of the abdomen in venous phase with sagittal (MIP) and coronal (MPR) images show a large inferior mesenteric vein (right arrow) from the splenic vein (left thin arrow) extending craniocaudally into the pelvis and draining into the left internal iliac vein (left thick arrow), perirectal collaterals are seen (dotted arrow)

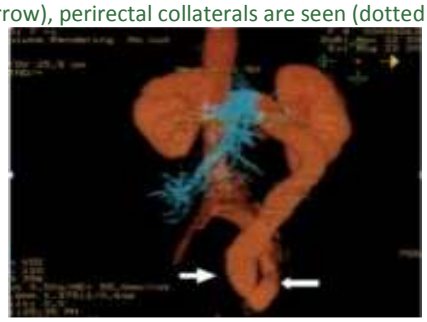

Figure 3: CECT of the abdomen , volume rendered images in venous phase show completely absent portal vein and the branches with a large inferior mesenteric vein(right arrow) draining into the internal iliac vein(left arrow) The superior mesenteric vein is shaded in blue

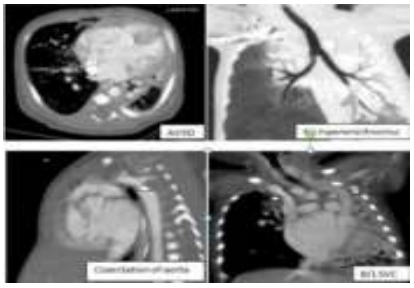

Figure 4a, 4b, 4c and 4d: MDCT - Cardiac angiogram with coronal and sagittal reformations show multiple cardiovascular abnormalities - atrio ventricular septal defect *(black arrow*), bilateral hyparterial bronchus (left sided isomerism), coarctation of aorta *(arrow head)* and bilateral superior vena cava *(asterix)*

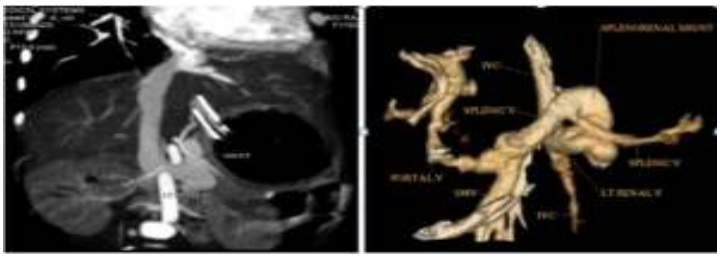

Figure 5a and 5b: Coronal CECT of the upper abdomen and the volume rendered images in venous phase shows a shunt *(dotted arrow)* between the splenic vein and the dilated left renal vein. Significant difference is seen in the calibre of the splenic vein proximal and distal to the shunt

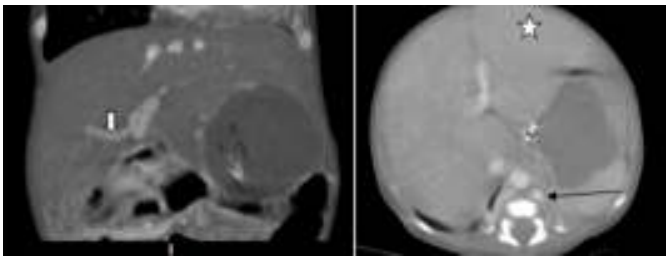

Figure 6a and 6b: CECT of the upper abdomen in coronal and axial views show a small and hypoplastic right portal vein (vertical arrow) with relative enlargement of the left lobe of liver (star). Enlarged hemiazygous vein (left arrow) is seen communicating with the left renal vein and draining into the left SVC

DISCUSSION

Congenital extra hepatic portal systemic shunt (CEPSS or Abernethy mal formation) is of two types. In type I (side to end anastomosis) complete agenesis of portal vein (CAPV) occurs and entire portal blood is shunted into systemic circulation. In type II (side-to-side anastomosis) part of the portal circulation enters the liver and rest of the blood is shunted into the systemic circulation. CAPV can be further divided into type A and type B.[3] In type A the superior mesenteric vein and the splenic vein do not join and drain separately into a systemic vein. Whereas in type B both the veins join together and drain into a systemic vein. Type I Abernethy malformation is usually seen in young females and associated with other cardiac anomalies like situs ambiguous, dextrocardia and septal defects; skeletal anomalies like hemivertebra; gastrointestinal anomalies like polysplenia, duodenal atresia, malrotation, annular pancreas, biliary atresia and renal tract anomalies like multicystic dysplastic kidney, vesicoureteric reflux and crossed fused renal ectopia.[4,5] Type II Abernethy malformation is usually present in a slightly older age group with no sex predominance and no congenital abnormalities. The portal vein develops from selective involution of right and left vitelline veins. Congenital absence of portal vein occurs due to excessive involution of vitelline veins. Due to the proximity of the portal vein to the inferior vena cava and since the development occurs during the same period, the portosystemic shunt in CAPV is commonly with the inferior vena cava.[6,7] Other rare sites of drainage can be into the left renal vein, azygos vein and internal iliac vein. Complications can be due to the structural or functional abnormalities of liver or related to the shunt. Liver dysfunction with atrophy and fatty changes can occur due to the reduced portal blood flow to the liver. Complications related to the shunt are elevated blood ammonia, bilirubin, galactose and rarely elevated insulin and androgen due to reduced clearance by the liver .Hepatic encephalopathy is often seen at an older age due to increased susceptibility of the brain to the toxic

ammonia.[8] Focal liver lesions like nodular regenerative hyperplasia (NRH), focal nodular hyperplasia, adenoma or hepatoblastoma are associated with CEPS. NRH is the commonest focal lesion in CEPS which appear hyper intense on T1-weighted MRI images and show homogenous and increased prolonged enhancement in the arterial and venous phase images.[9] USG shows abnormal liver size with absent portal vein or increased periportal echogenicity. It is used in the follow-up of focal liver lesions. Doppler helps to see the flow direction of the shunt and other vessels. MDCT angiography is fast and can be done in non-cooperative patients. It clearly depicts the shunt and has good resolution for the small vascular branches. MR angiography is the first choice of investigation because it is non- ionizing and serves the following purpose; it is used to visualize the abnormal vasculature and to characterize the liver lesions if present. Transrectal portal scintigraphy with[123] I iodoamphetamine is done to calculate the portosystemic shunt index. Shunt index >5% is abnormal. Shunt index>60% causes spontaneous encephalopathy.[10, 11] Conventional angiography can be done in type II shunt for coil embolization, transvenous liver biopsy and for the measurement of pressure gradient.[12] Liver biopsy is done in cases of absent portal vein on imaging. Type I shunts are reclassified as type II shunts when portal venules are seen under microscopy.[12] Patients should be on clinical, biochemical and radiological follow-up and those with liver dysfunction, hepatic encephalopathy and shunt index >60% should be intervened. Liver transplantation is the only surgical treatment for type I shunt. Coil embolization or surgical closure of the shunt can be done for type II shunt.[12]

CONCLUSION

Congenital extrahepatic portosystemic shunt is a rare vascular malformation and the clinician should consider this is as a differential diagnosis in children who present with non specific liver dysfunction. Knowledge of the imaging findings of CEPS is very important for accurate diagnosis which further aids in best therapeutic approach and management.

REFERENCES

1. Stringer, M.D. The clinical anatomy of congenital portosystemic venous shunts. Clin Anat. Mar 2008; 21: 147–157
2. Abernethy J. Account of two instances of uncommon formation in the viscera of the human body. Philos Trans R Soc Lond 1793; 17:292–299.
3. Ankur Gadodia, Raju Sharma, Harsh Kandpal, Rajinder Prashad. Congenital absence of portal vein with large inferior mesenteric - caval shunt. Tropical gastroenterology 2011; 32(3):223-22
4. Howard ER, Davenport M. Congenital extrahepatic portocaval shunts: the Abernethy malformation. J Pediatr Surg 1997; 32(3):494–497.
5. Murray CP, Yoo SJ, Babyn PS. Congenital extrahepatic portosystemic shunts. Pediatr Radiol 2003; 33 (9):614–620.
6. Komatsu S, Nagino M, Hayakawa N, Yamamoto H, Nimura Y. Congenital absence of portal venous system associated with a large inferior mesenteric-caval shunt: a case report. Hepatogastroenterology, 1995; 42:286–90.
7. Niwa T, Aida N, Tachibana K, Shinkai M, Ohhama Y, Fujita K, *et al*. Congenital absence of the portal vein: clinical and radiologic findings. J Comput Assist Tomogr. 2002; 26:681–6.
8. Kandpal H, Sharma R, Arora NK, Gupta SD. Congenital extrahepatic portosystemic venous shunt: imaging features. Singapore Med J 2007; 48(9): e258–e261
9. Brancatelli G, Federle MP, Grazioli L, Golfieri R, Lencioni R. Benign regenerative nodules in Budd- Chiari syndrome and other vascular disorders of the liver:radiologic-pathologic and clinical correlation.RadioGraphics 2002;22(4):847–862.
10. Kashiwagi T, Azuma M and Ikawa T, *et al*. Portosystemic shunting in portal hypertension: evaluation with portal scintigraphy with transrectally administered I-123 IMP. Radiology 1988; 169(1):137–140.
11. Gallego C, Miralles M, Marín C, Muyor P, González G, García-Hidalgo E. Congenital hepatic shunts. RadioGraphics 2004; 24(3):755–772.
12. Eduardo Alonso-Gamarra, Manuel Parrón, Ana Pérez, Consuelo Prieto, Loreto Hierro, Manuel López-Santamaría, Clinical and Radiologic Manifestations of congenital extrahepatic portosystemic shunt: A Comprehensive Review. RadioGraphics, 2011, Vol.31: 707-722.

Source of Support: None Declared
Conflict of Interest: None Declared