

Case Report

Hepatic vascular shunts in a newborn

Shivachev P*., Kiryazov K., Stefova M. and Georgieva M.

Abstract

Department of Pediatrics, University
Hospital "Sveta Marina", Varna,
Bulgaria

*Corresponding Author's E-mail:
drshivachev@yahoo.com

Hepatic Vascular Shunts (HVSs) are rare anomalies that consist of abnormal communications between the hepatic arteries, portal veins, and the hepatic or systemic veins and can be associated with severe morbidity and high mortality rate. We report a newborn patient with hepatic vascular shunts and discuss their etiology and management. We review the presentation, course of disease and outcome in a male newborn who developed hepatomegaly, gastrointestinal bleeding, severe anemia, signs of disseminated intravascular coagulopathy, hypoproteinemia, hypoglycemia and jaundice two hours after birth. Multiple intra- and extrahepatic portosystemic shunts and intrahepatic arteriovenous malformation were diagnosed using Doppler ultrasound and contrast-enhanced CT scan. Echocardiography showed enlarged hepatic veins and subsystemic pressure in the right ventricular. Despite vigorous intensive treatment for progressing liver failure the patient developed cerebral edema, pulmonary hemorrhage renal and heart failure, and died 11 days after birth. Autopsy was not performed due to the religious beliefs of the parents. Multiple HVSs in the newborn can lead to fulminant hepatic failure and fatal outcome. When practically the entire hepatic circulation is disturbed the only possible treatment option is allogeneic liver transplantation which is further limited due to the lack of size appropriate organ donors in that age group.

Key words: Congenital vascular malformations, Hepatic Vascular, Liver failure, Liver neoplasms, Shunts, Newborn

INTRODUCTION

Hepatic Vascular Shunts (HVSs) are rare anomalies that consist of abnormal communications between the hepatic arteries, portal veins, and the hepatic or systemic veins and can be associated with severe morbidity and high mortality rate (Gallego et al., 2004). We report a newborn who developed fulminant hepatic failure due to HVSs several hours after birth.

Case report

We present a 2-day old male newborn, born from second uncomplicated pregnancy via natural delivery, at 39th week of gestation, with body weight of 3300 g. His Apgar scores were 9 and 9 at 1st and 5th min, respectively. Initial physical examination was unremarkable with no

dysmorphic features, neurologic abnormalities, skin lesions, rash, or edema of the extremities.

Two hours after delivery the newborn's condition deteriorated. He was acutely ill, with pale skin and petechial rash on the face. The infant had mild respiratory distress. A 2/6 systolic ejection cardiac murmur was noted. The abdomen was distended with liver spanning 2.5 cm under the right costal margin. Lab results showed hypoglycemia, anemia, thrombocytopenia, hypoxia and hypercapnia with mild respiratory acidosis. Serum electrolytes were normal. Chest radiography revealed clear lung fields and slightly increased heart size. Abdominal radiography was abnormal only for hepatomegaly. Echocardiography demonstrated normal heart function and a small patent ductus arteriosus. Antibiotic, oxygen, haemostatic (vit. K) and fluid therapy



Figure 1. External view of the newborn.



Figure 2. Plain radiography of the chest and abdomen, 2nd day after birth, revealed clear lung fields, slightly enlarged heart size, and hepatomegaly

was initiated. He was twice transfused with packed red blood cells.

On 28th hour after birth the child was transferred to our department due to clinical deterioration with further liver enlargement, progressive of jaundice and gastrointestinal bleeding. At admission the child was critically ill. He was alert, with normal muscle tone and reflexes. Vital signs were: temperature – 36.6°C, body weight – 3300 g, respiratory rate – 38 respirations/min, heart rate – 150 beats/min, blood pressure – 66/28 mm

Hg, oxygen saturation – 95% under oxygen tent. Skin was pale, jaundiced, with normal turgor, and petechial rash on the face. Chest examination showed shallow irregular breathing without rales and wheezing. Regular heart rhythm with 2/6 systolic ejection murmur was noted. He had abdominal distention with hepatomegaly (liver spanning about 5 cm below right costal margin, with soft-elastic consistency and smooth surface) and splenomegaly (spleen distended about 2 cm below left costal margin) (Figure 1).

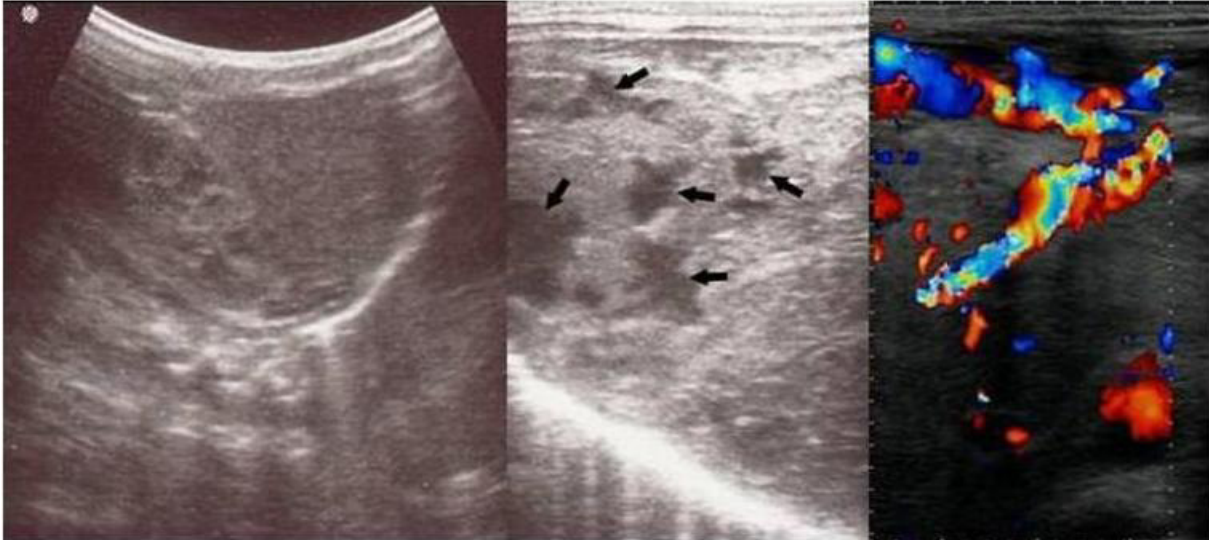


Figure 3. Abdominal ultrasound. A, 2nd day after birth, showing heterogeneous structure of the liver. B, 5th day after birth showing heterogeneous liver structure with multiple anechoic zones with irregular shape and different size (arrows). C, Color Doppler US image demonstrating the vascular nature with turbulent flow of the aforementioned lesions

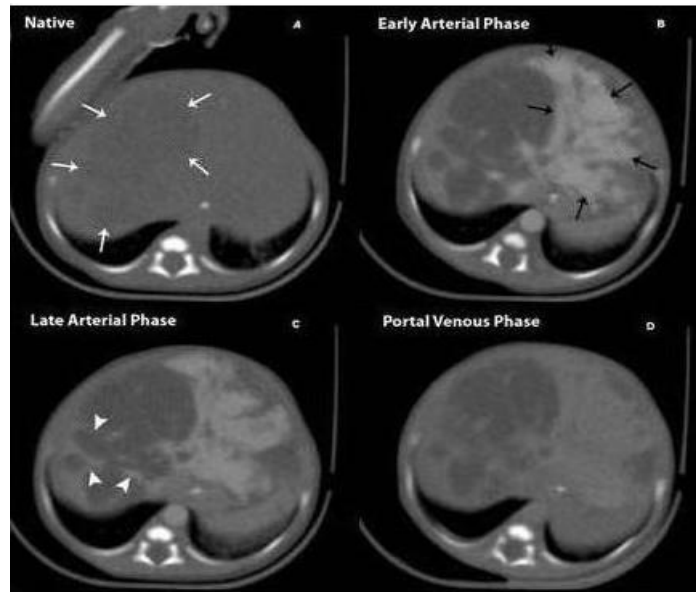


Figure 4. CT scan. A, Native CT scan showing a large hypodense, lobulated lesion with multiple interconnected lacunas in the right lobe (white arrows). B, C and D, Contrast enhanced multiphase CT showing heterogeneous structure (black arrows) in the left hepatic lobe, which increases its density and resembles a conglomerate of unevenly dilated, tortuous and branching vascular structures (B). The aforementioned structure does not increase its density in the cystic zones, unlike the internal septa (white arrowheads), which increase their density significantly and appear to be connected to the hepatic hilus (C).

CBC was notable for anemia and thrombocytopenia. Arterial blood gases, acid base balance and electrolytes were within normal ranges. C-reactive protein was slightly elevated at 8.8 mg/L. Complete sepsis work up and

screening for congenital infections were all negative. Renal function tests indicated the presence of renal failure. Liver function tests revealed blood glucose – 2.7 mmol/L (2.8-5.0), total protein – 53 g/L (46-74), albumin –

34 g/L (25-34), ammonia – 82,6 $\mu\text{mol/L}$ (21-95), total bilirubin - 125 $\mu\text{mol/l}$ (< 123.6), conjugated bilirubin – 12 $\mu\text{mol/L}$. Clotting studies were prolonged with a prothrombin time of 33,9 seconds (10,4-12,6), activated partial thromboplastin time of 50,6 seconds (22.1-28.1), and fibrinogen of 0.5 g/L (1.8-3.5). Liver biochemistry tests were all elevated (ALT – 592 U/L, AST – 1845 U/L, GGT – 153 U/L, Alkaline phosphatase - 443 U/L). The alpha-fetoprotein (a-FP) level was normal at 16 247 ng/mL.

Plain radiography of the chest and abdomen performed on the 2nd day after birth showed no significant differences from the first x-ray series. Echocardiography was notable for subsystemic pressure in the right ventricular (60 mmHg) and enlarged hepatic veins. Abdominal ultrasound of the liver performed at the same time showed heterogeneous structure (Figure 2).

A follow up abdominal ultrasound performed at the 5th day after birth showed heterogeneous liver structure with multiple anechoic zones with irregular shape and different size (Figure 3).

Doppler ultrasound and contrast-enhanced CT scan performed on the 5th day after birth demonstrated multiple intra- and extrahepatic portosystemic shunts and intrahepatic arteriovenous malformation (Figure 4).

Despite vigorous intensive treatment for progressing liver failure, coagulopathy, anemia and renal failure the patient developed cerebral edema, pulmonary hemorrhage and heart failure and died 11 days after birth. Autopsy was not performed due to the religious beliefs of the parents.

DISCUSSION

We have described a newborn that became acutely ill two hours after birth with signs and symptoms of acute liver failure. Considering the presentation, the clinical course and the initial laboratory and imaging results our differential diagnosis included infections (bacterial, viral, fungal), metabolic diseases (glycogen storage disease, mucopolysaccharidosis, galactosemia, neonatal hemochromatosis), neoplasms with sufficient vasoproliferative component and congenital vascular malformations.

Most cases of acute liver failure in the newborn are the result of either metabolic or infectious diseases (Suskind and Murray, 2006). The sepsis workup was negative, as was the workup for congenital viral infections. Metabolic conditions could not be completely excluded by enzymatic determination because of this neonate's fulminant course. However the possibility of storage was less probable in view of absence of significant acidosis or persistent hypoglycemia.

The progression of the changes in the liver demonstrated on Doppler ultrasound, and contrast-enhanced CT imaging narrowed down the differential

diagnosis to HVSs due to neoplasms or congenital vascular malformations. Following Mulliken and Glowacki classification HVMs can be classified as fast-flow (arteriovenous malformations (AVMs), arterioportal fistulas), slow-flow (portosystemic shunts (extra- and intrahepatic), venous and lymphatic malformations) and combined forms (Mulliken and Glowacki, 1982).

Neoplasms with sufficient vasoproliferative component that can lead to the development of arteriovenous or portovenous shunting are hemangioendothelioma, hepatoblastoma, angiosarcoma, hamartoma and metastatic neuroblastoma (Gallego et al., 2004). The fetal and neonatal hemangioma, mesenchymal hamartoma, and hepatoblastoma have survival rates of 75%, 64%, and 25%, respectively (Isaacs, 2007).

Liver neoplasms are very rare during the neonatal period (only 5% of all tumours in the fetus and newborn) and accurate diagnosis can be challenging (Makin and Davenport, 2010). The most common are benign infantile haemangioendotheliomas, cavernous haemangiomas, mesenchymal hamartomas and malignant hepatoblastomas. Differential diagnosis can be difficult because of non-specific clinical symptoms, misleading imaging, inconsistent expression of tumor markers and sometimes similar histological findings (Von Schweinitz, 2003). Congenital neuroblastomas with massive hepatic metastases are exceedingly rare and can mimic congenital infections, primary liver tumors, and storage disease (Thompson et al., 2008). Urine studies for vanillylmandelic acid and homovanillic acid were ordered to rule out metastatic neuroblastoma, but were never sent due to his extreme oliguria. Hepatoblastoma is the most common malignant liver tumor, but less than 10% are said to occur in the neonatal period (Isaacs, 2007; Von Schweinitz, 2003). The α -FP level in our case was normal. However about half of neonatal hepatoblastomas may not produce elevated levels of α -FP (De Ioris et al., 2008; Isaacs, 2007; Makin and Davenport, 2010). Bone marrow biopsy was recommended but could not be performed due to the newborn's coagulopathy, but the absence of blast in peripheral blood made the diagnosis of congenital leukemia in our case less likely. Rapid growth, poor response to medical treatment, and metastasis are considered reliable markers of malignancy even in presence of benign histology (Nazir and Pervez, 2006).

The etiology of HVSs may be either congenital or acquired. Color Doppler US complemented by multiphasic CT allows early diagnosis and evaluation of HVSs, and suitable treatment (Dessouky et al., 2011). Infantile hemangioma and liver AVMs, both exhibit "fast flow" arteriovenous shunting, have a similar presentation but a different natural history (Boon et al., 1996). On the contrary to hemangioendothelioma AVMs are present at birth (Lima et al., 2005). The cystic transformations of the parenchyma of the liver, the portosystemic shunts in the area of the hilus and arteriovenous shunts in the area of

the left hepatic lobe, combined with the laboratory studies and course of disease in our case, narrowed down the differential diagnosis to partially thrombosed vascular neoplasm of the liver (either primary or secondary) or congenital defect in the development of the liver and its vascular structures.

Conflict of Interest

The authors report no conflicts of interest.

CONCLUSION

Multiple HVs are extremely rare clinical entity that can lead to fulminant hepatic failure and fatal outcome. A precise diagnosis is sometimes very difficult. Whether the shunts in the reported patient are primary vascular malformations or secondary to neoplasm we can only presume due to the lack of pathological examination of the lesions in our case.

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