Asian Journal of Clinical Pediatrics and Neonatology Case Report

Acute Liver failure in a neonate –A devastating complication of sepsis

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Abstract

Acute liver failure in neonates is incidentally rare with very high mortality in spite of aggressive management. Causes include sepsis, metabolic, hematological, congenital vascular abnormalities and drugs. Early aggressive treatment, multidisciplinary involvement and liver transplantation has some hope in this regard. We are presenting a neonate with acute liver failure who succumbed despite all possible management available.

Key words Liver failure, Neonate, Sepsis

INTRODUCTION

Acute liver failure is a multisystem disorder characterized by an acute severe impairment of liver function with or without encephalopathy with hepatic necrosis in a patients with no recognizable chronic liver disease.[1] The commonest cause of acute liver failure in neonates is septicemia secondary to Escherichia coli, Staphylococcus aureus, or Herpes simplex.[2] Other causes include Adenovirus, Echovirus, and Coxsackie virus infection. Pediatric acute liver failure is a rare but life threatening disease.[3] When hepatic involvement is advanced with massive hepatic necrosis, liver transplantation is the only answer and mortality may be >70% without liver transplantation.[4] The treatable condition like sepsis should be kept in mind as a cause of neonatal liver cell failure.

CASE PRESENTATION

A term male baby was brought to emergency room on day 7 of life with history of lethargy, seizures and discoloration of skin. Baby was born by LSCS and said to have cried immediately after birth, APGAR at 1 and 5 minute were 8 and 9 respectively. On examination, baby was lethargic with poor respiratory effort and noticed to have petechial rashes. Septic markers done showed increased CRP with blood culture growing Klebsiella pneumonia. On admission to our NICU, baby was sick and septic. Baby was managed appropriately with required measures in the form of ventilation, Inotropes and blood products. Baby was investigated in detail to rule out sepsis induced organ dysfunction, including hematological, hepatic and renal parameters. Investigations revealed DIC like picture and massive liver cell failure as depicted in table 1. Supportive measures like Ventilation, Inotropes, and blood products were started. In view of rising liver enzyme levels, hepato protective measure like N-acetyl cysteine was started.Antiviral and antifungal drugs were administered and stopped subsequently after the results came negative. As a part of sepsis management and deteriorating condition baby also underwent exchange transfusion

Investigations were also carried to rule out other causes of liver dysfunction. TORCH panels, HSV PCR, Dengue panel, metabolic workup, and serum complement factors were normal. Serum ferritin was high. In view of high ferritin level, neonatal hemochromatosis was thought of and Punch lower lip biopsy was performed which turned out to be normal (no hemosiderin pigments). Despite all the above measures including exchange transfusion, baby succumbed to death. Post mortem liver biopsy was consistent with massive hepatic necrosis, Pas stain was negative. As liver transplantation is the last option to manage liver failure in neonates, option was discussed with parents but baby succumbed to illness before the transplantation could be carried out.

DISCUSSION

Septicemia in newborn period is common and still adds on to mortality despite various measures to prevent and treat it. Although, sepsis induced organ dysfunction or its cascade is well known, to an extent sepsis induced massive liver cell failure is rare. Neonatal ALF is not defined; however, data is contemplated from pediatric definition. The Pediatric Acute Liver Failure Study Group (PALFSG) defined ALF as "biochemical evidence of liver injury, with no history of known chronic liver disease with coagulopathy not correctable by vitamin K administration and INR greater than 1.5, if the patient had encephalopathy or greater than 2.0, if the patient does not have encephalopathy".[2] As per the above mentioned definition, our case satisfies the criteria for ALF.This baby had presented with features of sepsis and coagulopathy, which in general, is a presentation of any neonatal sepsis. An attempt was made to rule out sepsis induced liver dysfunction as coagulopathy was severe and persistent despite all corrective measures. The neonate was supported by other hepatoprotective measures like N-Acetyl cysteine infusion which is used in non-acetaminophen liver cell failure.[2,5,7] Secondary causes of liver dysfunction were also ruled out. The common causes of liver failure in neonates are Galactosemia, Tyrosinemia, Neonatal haemochromatosis, Haemophagocytic lymphohistiocytosis congenital leukaemia, Septicemia and shock.[9,10] Appropriate tests should be done for each entity to pinpoint any possible causes. Serum ferritin should also be considered in babies with liver failure, however, relying on ferritin levels when associated sepsis is hard. As ferritin is an acute

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Table 1 – Laboratory investigations of the patient

	Day 1	D ay 2	Day 3	Day 4	D ay 5	Day 6	Day 7	D ay 8
PT	74.5	120	46.2	37.9	58.3	35.3		>120
INR	9.0		4.94	3.9	6.6	3.5		>9
aPTT	>120	107	82.5	86.8	91.7	66.6		>120
Platelet count	35000	21000	40000	46000	36000	30000	25000	14000
SGOT	1493				303			
SGPT	1040				213			
Bilirubin Total	16.3				10.9			
Direct	3.9				4.6			
Alb umin	2.4				3.0			2.9
Cr eatinine	0.49				0.48			0.41
S. Ferr itin							15925	
CRP	20				12.8			6.7

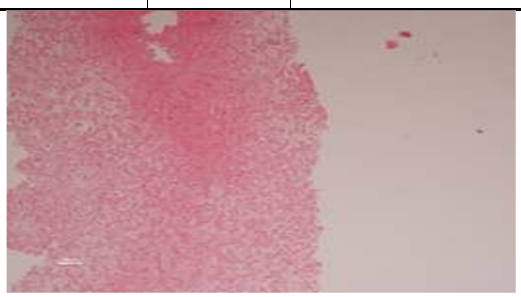

Figure 1: Liver biopsy-massive hepatic necrosis

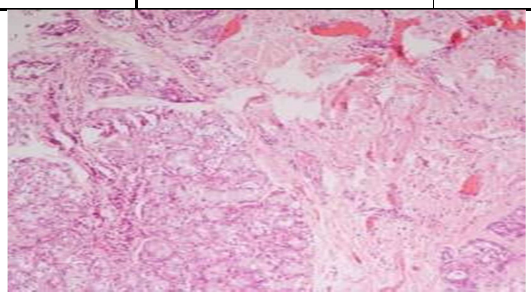

Figure 2: Buccal biopsy-Normal, no evidence of neonatal hemochromatosis, salivary glands seen.

phase reactant WHO recommends always CRP along with the serum ferritin. Whatever may the etiology, the management should be aggressive and intensive. The management should include treating coagulopathy, infection (including possible viral and fungal infection) and management of all possible complications. Possible complications may be hemodynamic instability and shock requiring inotropic support, renal failure may require dialysis. Liver transplantation for neonates has increased the hope for survival. The decision for liver transplantation should be taken early in the disease process for optimum result.[8]

CONCLUSION

Acute liver failure could be a major threat in any neonate with sepsis. The management includes treating the sepsis with multiple supportive measures. In these babies liver transplantation may be a ray of hope which should be discussed as early as possible once the diagnosis is made.

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Funding: None

Conflict of Interest: None stated