



Idiopathic Neonatal Hepatitis- A Case Report

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ABSTRACT

Idiopathic neonatal hepatitis represents a heterogenous group of neonatal cholestatic diseases of unknown origin characterized by prolonged obstructive jaundice associated with giant cell transformation of hepatocytes. The disorder occurs predominantly in neonates and represents 10% to 15% cases of neonatal hepatitis. We report a case of Idiopathic neonatal hepatitis in a 4 month old male child and describe the histopathological features, differential diagnosis and role of liver biopsy in diagnosing it.

KEYWORDS : Idiopathic neonatal hepatitis, neonatal cholestasis, giant cell hepatitis.

INTRODUCTION:

Idiopathic neonatal hepatitis also called as Neonatal giant cell hepatitis is an important diagnostic consideration in neonates who present with jaundice. Neonatal hepatitis may be caused by metabolic diseases, viruses or genetic disorders¹. In Idiopathic neonatal hepatitis, however, the cause of inflammation remains unknown. The affected neonates have jaundice, dark urine, light or pale stools and hepatomegaly². We report a case of Idiopathic neonatal hepatitis in a 4 month old male child on liver biopsy after excluding all other causes of neonatal hepatitis.

CASE REPORT:

A 4 month male child presented with complaints of fever and convulsions since 1 day. He had a past history of jaundice 1 month back and was admitted to Neonatal intensive care unit for 8 days, during which the cause could not be evaluated. He had no significant family history. On general physical examination, the child was febrile, unconscious and responded to painful stimulus. Icterus was present. Per abdominal examination showed hepatomegaly, 2cm below right costal margin. Laboratory examination showed raised liver function tests. Radionucleotide hepatobiliary scanning was negative. TORCH group of infections were negative. Under aseptic precautions, true cut liver biopsy was done and sent for histopathological examination. On gross examination, specimen consisted of single linear bit of tissue measuring 1.5cm in length. On microscopy, the section studied showed lobular disarray and cholestasis (Figure 1) along with hepatocytes undergoing ballooning degeneration and giant cell transformation (Figure 2). Portal tract showed fibrosis and lymphocytic infiltration (Figure 3).

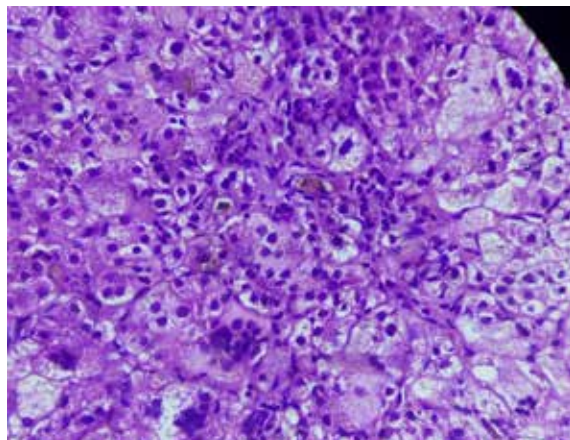


Fig 1.

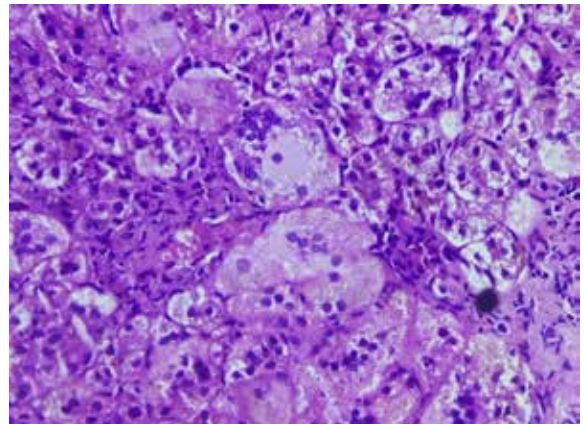


Fig 2.

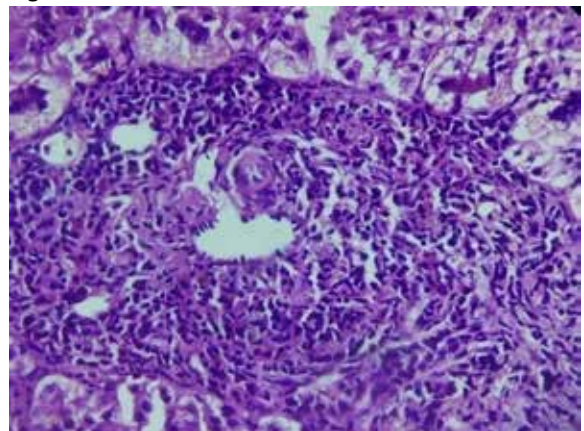


Fig 3.

DISCUSSION:

Idiopathic neonatal hepatitis is one of the more important causes of neonatal cholestasis. It is regarded as one of the clinical presentations of 'idiopathic obstructive cholangiopathy', just like extrahepatic biliary atresia³. The onset of idiopathic neonatal hepatitis is normally within the first 3 months of birth. Male babies are affected slightly more than the female babies⁴. Idiopathic neonatal hepatitis occurs in two forms, Sporadic (75%) and familial (25%). Based on this our case falls into sporadic form.

It is not possible to discriminate between intrahepatic and extrahepatic causes of neonatal cholestasis, or idiopathic neonatal hepatitis and metabolic, infectious, genetic or toxic causes by using clinical or laboratory parameters. Liver histology is slightly more helpful³. In our case also, liver biopsy played a critical role in diagnosis. The multinucleated giant cells are formed from the syntitial fusion of several mononucleated hepatocytes. The giant cell transformation of parenchymal cells is considered as a common response of the neonatal hepatocytes to various types of injury and its occurrence in adults is very rare⁵.

Idiopathic neonatal hepatitis should be differentiated from other causes of neonatal hepatitis and biliary atresia⁶.

Other known causes of neonatal hepatitis are ruled out by doing clinical examination, blood tests, workup of infectious causes, hepatobiliary scans, possible metabolic and genetic testing, and possibly liver biopsy.

Neonates with biliary atresia present with neonatal cholestasis, but exhibit normal birth weight, postnatal weight gain, associated more frequently with pale stools, treated surgically and have poor prognosis. Liver biopsy shows features of bile ductular proliferation, bile duct and ductular bile plugs and portal fibrosis⁷.

In idiopathic neonatal hepatitis more than 90% make a complete biochemical and clinical recovery⁸.

CONCLUSION:

Idiopathic neonatal hepatitis is a diagnosis attained by excluding all other causes of neonatal hepatitis. It should be included in the differential diagnosis in a child with prolonged obstructive jaundice. Liver biopsy is considered as the most reliable method to differentiate idiopathic neonatal hepatitis from biliary atresia⁹.

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