Sudanese Journal of Paediatrics 1984; 3: 42 - 45

NEONATAL HEPATITIS AND BILIARY ATRESIA: A COMMON AETIOLOGY.

Z. A. Karrar,
Department of Paediatrics and Child Health, Faculty of Medicine, University of Khartoum

Abstract A Sudanese girl presented at 2½ months of age with persistent jaundice - starting at age 7 days - and hepatosplenomegaly. Extrahepatic biliary atresia with absence of the gall bladder was confirmed surgically; whereas an open liver biopsy showed a typical appearance of neonatal hepatitis. It is suggested that this case supports the hypothesis that neonatal hepatitis and biliary atresia have a common aetiology.

Key words Infant newborn; Hepatitis; Bile duct/abnormalities.

INTRODUCTION

Biliary atresia and neonatal hepatitis are the commonest causes of obstructive jaundice in the neonatal period, biliary atresia being slightly commoner[1-3]. The incidence of biliary atresia is estimated to be 1 in 20-30,000.

The aetiology remains obscure. One hypothesis is that biliary atresia is a congenital failure of canalization of the biliary duct, while neonatal hepatitis is mainly a perinatal insult to the hepatocytes. An alternative hypothesis is that neonatal hepatitis and biliary atresia have a common aetiology and they represent different manifestations of the same insult[3,4]. In favour of this hypothesis the following evidence is put forward:

a. The onset of both is usually at 2-3 weeks of postnatal life.
b. There were reported cases of atresia after documented postnatal patency[5].
c. Bile duct recanalization after surgical confirmation of atresia in the neonatal period have been reported[1].
d. Both are common in trisomy 17-18 suggesting a perinatal aetiology leading to chromosomal damage as well as hepatic insult[6]. This is a case report which strongly favours the hypothesis that both have a common aetiology.

Case report

R.T., a 2½-month-old girl, presented on 3.10.76 with persistent jaundice and vomiting. The jaundice was noticed at the age of 7 days and was getting deeper. The stools were persistently pale and greasy. Antenatal history was uneventful. There was neither history of drug intake nor that of irradiation. The delivery was normal. She had a 2½-year-old female sibling who was healthy.

On examination, she was deeply jaundiced, not anaemic, with a distended abdomen. The liver was 7 cm below the costal margin, at the anterior axillary line, very firm with a sharp margin. Spleen was enlarged 4 cm below the costal margin and soft.

Investigations showed excessive bilirubin in the urine. There were neither reducing substnaces, aminoaciduria nor owl-eye cells consistent with cytomegalovirus infection. Haemogram and clotting profile were normal. Serum bilirubin was 11.2 mg/dl, conjugated 8 mg/dl, cholesterol 355 mg/dl, alkaline phosphatase 26 K A units, total proteins 6.7 g/dl, albumin 4.3 g/dl, serum glutamic oxaloacetic transaminase (SGOT) 80 units. Khan and Wasserman tests were negative, maternal blood group O-ve and child's blood group O-ve. Histopathology of a percutaneous needle liver biopsy showed few hepatocytes, deeply bile stained, and the architecture could not be indetified.

The parents gave consent for an explorative labaratomy and this was performed on 29.10.1977. At operation, the bile duct was completely artritic with complete

absence of the gall bladder. An open liver biopsy was done and the histopathological examination revealed a typical picture of giant cell hepatitis with peri-portal infiltration.

Her postoperative course was uneventful. She was put on vitamines K, A & D parenterally. Her weight gain on milk feeds containing medium-chain triglycerides was unsatisfactory. She was persistently restless and had difficulty in sleeping. The serum bilirubin rose to 16.5 mg/dl and the liver increased in size to 12.5 cm.

DISCUSSION

In this case extrahepatic biliary atresia with absence of the gall bladder was confirmed surgically, While the liver biopsy was showing a typical appearance of neonatal hepatitis.

It is unfortunate that we could not perform all the virology and serology studies that we hoped to do regarding the aetiology, but the case strongly suggests a common aetiological factor.

ACKNOWLEDGEMENT

I am grateful to Dr. Saddig M. A. Wahab for performing the laboratory investigations, to Professor Ahmed M. El Hassan for examining and reporting on the liver biopsies and to the doctors and nursing staff of ward C2 for their help.

REFERENCES

1. Alagille D. Clinical aspects of neonatal hepatitis. *American Journal of Diseases of Children* 1972;123:287.
2. Danks DM, Campbell PE. Extrahepatic biliary atresia: comments on the frequency of potentially operable cases *Journal of Pediatrics* 1966;69:21.
3. Danks DM. Prolonged neonatal obstructive jaundice. A survey of modern concepts. *Clinical Pediatrics* 1965; 4:499-510.

4. Landing BH. Considerations of the pathogenesis of neonatal hepatitis, biliary atresia and choledochal cyst: the concept of infantile obstructive cholangiopathy. In: Bill AH, Kasai M, eds, Pediatric Surgery. Philadelphia: University Park Press, 1974; 113-139.
5. Holder TM, Aschcraft KW. The effect of bile duct ligation and inflammation in the foetus. *Journal of Pediatric Surgery* 1967;2:35.
6. Alpert LI, Strauss L, Hirschhorn K. Neonatal hepatitis and biliary atresia associated with trisomy 17-18 syndrome. *New England Journal of Medicine* 1969; 280:16-20.