Autoimmune hepatitis: a rare cause of acute hepatic failure in male infant

***Madiha Zainab**[1]

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Introduction

Autoimmune hepatitis (AIH) is a chronic inflammatory disease of the liver of unknown aetiology characterised by circulating autoantibodies, hypergammaglobulinaemia, necro-inflammatory changes in liver histology and dramatic response to immunosuppresants[1]. AIH has varied presentations ranging from asymptomatic cases to fulminant forms affecting all age groups from infancy to elderly. Here we report a male infant who presented as acute hepatitis, progressed to fulminant hepatic failure and succumbed.

Case report

An 8 month old male infant, born of a non-consanguineous marriage, was admitted with a history of low grade fever for one month and yellowish discolouration of sclera for 12 days with lack of appetite. Jaundice was insidious in onset and gradually progressive without any pruritus or clay coloured stools. There was no history of bleeding manifestations, altered sensorium, rash, pain in the abdomen, drug intake or blood transfusion. There was no significant family history. The baby was developmentally normal. On admission, pallor and jaundice were present. The height, weight and head circumference were between the 3rd and 50th centile. The liver was 2cm, non-tender, liver span was 7cm, spleen was not palpable and there was no ascites. A provisional diagnosis of infective hepatitis was made. Initial investigations revealed a haemoglobin (Hb) level of 6.3g/dl, microcytic hypochromic anaemia and normal platelet and white blood cell (WBC) counts. Reticulocyte count was 1.2%. Viral serology, human immunodeficiency virus (HIV) 1 and 2, malarial parasite dual antigen (MPDA), and Widal test were all negative. Blood and urine cultures were normal. Coagulation profile was normal. Liver function tests showed a total serum bilirubin (TSB) level of 15.5 mg/dl (direct 10.7 mg/dl, indirect 4.8 mg/dl), serum albumin 2.6g/dl, globulin 3.5g/dl, alanine transaminase (ALT) 593U/L, aspartate transaminase (AST) 1588U/L gamma glutamyl transferase (GGT) 29U/L and alkaline phosphatase (ALP) 300IU/L.

On day 3, high grade fever persisted, the child had 2 episodes of haematemesis and the sensorium was altered. Liver span became 9cm, spleen 1cm and ascites was present. Repeat investigations revealed a Hb level of 5.4g/dl, a positive direct Coombs test (DCT), a TSB of 17.8 mg/dl (direct10.7 mg/dl, indirect 7.1 mg/dl), globulin 3.8 g/dl, AST 584U/L, ALT 309U/L, international normalised ratio (INR) no coagulum, prothrombin time (PT), and activated partial thromboplastin time (APTT) prolonged. Child went into hepatic failure.

Wilson disease was excluded as urinary copper and caeruloplasmin levels were normal and on slit lamp examination Kaiser-Fleischer (KF) ring was absent. Doppler ultrasonography was suggestive of liver parenchymal disease. Autoantibodies antinuclear antibody (ANA), anti-smooth muscle antibody (ASMA), anti-mitochondrial antibody (AMA) were negative but anti-liver kidney microsomal antibodies (LKM-1) were positive (4+). Steroids were started but the child succumbed on day 6 of admission. Liver biopsy could not be done due to coagulopathy and poor general condition of the patient.

Discussion

AIH is a rare cause of chronic liver disease in India and often presents late[2]. Prevalence of paediatric AIH in India has not been documented[3]. Case reports have been published where AIH has presented with dermatological manifestations like erythema nodosum[4], autoimmune haemolytic anaemia and sickle cell anaemia. However, fulminant hepatic failure is a rare presentation and that too in a male infant.

AIH should be considered in the diagnostic work up of any patient with cryptogenic liver disease. Diagnosis basically refers to exclusion of other possible aetiologies like chronic viral infection and Wilson disease and the use of biochemical and histological criteria[5]. A simplified scoring system (2008) by AIH Society group has lower sensitivity but higher specificity in diagnosing AIH[6]. Anti

[1]*Vivekananda Institute of Medical Sciences, India*
**Correspondence:* madiha_zainab@ymail.com
https://orcid.org/0000-0003-2993-7481

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LKM 1 serum reactivity defines AIH 2 which most commonly occurs in infants and children and is commoner in females[7]. AIH 1 with positive ANA and SMA are common in adults. Increase in globulin or IgG is found in 85% patients with AIH and may be normal in acute onset AIH. In our case, the child presented with acute hepatitis which turned into fulminant liver failure and DCT was positive so that associated autoimmune haemolytic anaemia was responsible for the pallor which was undiagnosed before. Weinster *et al* reported that a 5 month old female infant, diagnosed as AIH at 5 months of age, developed liver disease at 6 years with anti SMA positive[8].

AIH is usually associated with other autoimmune disease like thyroiditis and type 1 diabetes. AIH may also be associated with primary sclerosing cholangitis and cholestasis also known as overlap syndrome[9]. Treatment for AIH is steroids with or without azathioprine depending on the response. In non-responders, other immunosuppressive drugs like cyclosporine may also be used. AIH should be considered as an alternative diagnosis of cryptogenic liver disease and hepatic failure irrespective of age and sex of the patient. Early diagnosis and treatment improves patient survival and reduces the need for liver transplantation.

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