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Case Report

AUTOIMMUNE TYPE-2 HEPATITIS IN A 5-YEAR-OLD CHILD: A CASE REPORT

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ABSTRACT

Autoimmune hepatitis (AIH) is a long-term noncommunicable disease with unknown etiology. Depending on the antibody profile, it is classified into two types: type-1 AIH and type-2 AIH. Type-1AIH is frequently identified in adults, while type-2 is typically diagnosed in children. We report a case of 5 years old female child who presented with complaints of jaundice for the last three months, was diagnosed as AIH type-2, treated with prednisolone and responded well.

INTRODUCTION

Autoimmune type-II hepatitis (AIH type-2) is an inflammation of the liver that emerges when the body's immune system attacks its hepatocytes. Genetic and environmental factors influence the etiology; however, the exact cause is still unknown. The clinical manifestations of autoimmune hepatitis may vary from person to person but appear suddenly. Depending upon the stage, the most frequently reported symptoms include: dark urine, pale stools, ascites, and hepatomegaly. Besides, jaundice, itching, fatigue, nausea, vomiting, skin rashes, abdominal discomfort, spider angiomas can also be seen in the patient. The two types of autoimmune hepatitis are type-I and type-II.[1] Type-I occurs more frequently and can present at any age, whereas type-2 occurs more commonly in children and young people but is reported in adults also. The distinguishing feature between the two is that type-1 AIH is characterized by an elevated level of ANA(anti-nuclear antibodies) and ASMA(anti-smooth muscle antibodies). However, increased levels of anti-LKM (anti-liver/kidney antibodies) and ALC-1 (anti-liver cytosol 1 antibody) can be seen in type-II AIH. If left untreated, AIH can result in liver scarring or cirrhosis, leading to liver failure. The most frequently reported complications associated with autoimmune hepatitis include: esophageal varices, liver failure, and in some cases, liver cancer.[2]

Case report

A 5-year-old female child presented to the pediatrics in-patient department with chief complaint of yellow discoloration of skin and sclera along with anorexia for the last three months. There was no history of fever, abdominal pain, clay-colored stool, pruritis, or blood transfusion. Also, there was no history of hematemesis and weight loss or similar complaint in the family. On examination, icterus was present with hepatomegaly, but there were no signs of portal hypertension. As per the guardian, the immunization of the child was up to date. Birth history revealed normal vaginal delivered child, who cried immediately after birth (CIAB). There was no history of bronchial asthma, tuberculosis, hypertension, and epilepsy.

On examination, the general condition was fair; respiratory and heart rates were 26 and 80 beats per minute, respectively. Investigations showed high levels of serum glutamic oxaloacetic transaminase (SGOT), serum glutamic pyruvic transaminase (SGPT), conjugated bilirubin, globulin, and alkaline phosphatase (ALP). Immunoserological investigations showed positive LKM (liver-kidney microsome) antibody and immunoglobulin-G and a negative anti-nuclear antibody (ANA) and anti-smooth muscle antibody (ASMA). The prothrombin time (PT) was also increased. Ultrasound sonography (USG) of abdomen revealed normal liver size with hypoechoic appearance, i.e., periportal cuffing in both lobes. Color doppler ultrasound of both lower limbs was normal. Anti-hepatitis C virus (HCV), hepatitis-B surface antigen (HBsAg), hepatitis-A, and ELISA were negative.

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Table 1 shows abnormal laboratory values on admission

Test	Patient result	Reference range
SGOT	2100 IU/L	6-40 IU/L
SGPT	1250 IU/L	6-40 IU/L
Bilirubin	5.8 mg/dl	0.1-0.2 mg/dl
Conjugated	2.0 mg/dl	0-0.3 mg/dl
ALP	343 U/L	25-110 U/L
Globulin	3.8 gm/dl	1.5-3.5 gm/dl
Test-PTI	24 sec	11-16 sec
INR	1.8	≤1.1
LKM	1:40 (Positive)	1:40-1:640

Clinical characteristics and laboratory tests eventually revealed autoimmune hepatitis type –II disease.

Initially, the patient was treated with ursodeoxycholic (150mg, OD) and injections of vitamin-K 10mg with four units of FFP. Methylprednisolone (Pulse therapy) was given at 175mg with 30ml normal saline (NS) over 30 minutes for two days. After a few days, the patient's general condition was good; liver function tests and PT-INR showed improvement, so the child was discharged under satisfactory conditions. (Table no.02 shows the improved laboratory values at the time of discharge) The patient was advised medications tablet prednisolone (10mg, BD), lansoprazole (15mg, OD), ursodeoxycholic acid (150mg, OD), silymarin (70mg, OD), hepamerz (1 tablet, BD), and syrup zincovit (5ml, OsD) at the time of discharge.

DISCUSSION

In the pediatric population, the AIH is divided into type-1 and type-2 based on antibody profile. The cardinal feature of type-1 AIH is characterized by positive anti-SMA or ANA, whereas type-2 is positive for anti-LKM antibody, mainly due to the deficiency of IgA antibody. 75% of affected children are female; female to male ratio is 4:1 in type-1, and 10:1 for type-2 AIH. Liver biopsy is necessary to assess the severity of AIH and to decide the treatment. Moreover, if there is a cholestatic profile, it is crucial to rule out the possibility of autoimmune sclerosing cholangitis (ASC). Although AIH affects 1lakh-2lakhs individuals in the United States, the prevalence is less in India. Besides, the prevalence and prognosis of AIH remain unclear.[3]A study reported that the prevalence of clinical manifestations in children with AIH was pallor (26%), abdominal pain (38%), anorexia (47%), non-specific weakness (57%), and jaundice (58%).[4]

The diagnosis of AIH in pediatric population uses the established IAIHG scoring system. The scores are assigned to the scoring system variables, including clinical, laboratory, and histological features. The scores are further divided into three categories: if a score is more than 17 before treatment that provides a definite diagnosis, the score between 15-17 is considered probable, and less than 15 exclude from the diagnosis of AIH.[5]The only noticeable difference between the score used for pediatric and adult is the amount of titer considered as significant for autoimmune antibodies. Autoimmune positivity is extremely rare, and a titer of 1:20 is considered significant, especially in the pediatric population.[1]
The laboratory findings in our patient were consistent with AIH, and she had a long-term history of jaundice. She was immediately started on methylprednisolone. Prednisolone with immunosuppressive drugs is the mainstay of treatment. In addition, the steroid-sparing agent can be used. Within 6-9 months, 75-90% of the affected children normalize their biochemical profiles. However, most cases will rapidly progress to cirrhosis and liver failure without treatment, and after that, the only treatment left is liver transplantation.

In 1970, the clinical trial results revealed that the standard treatment for AIH includes corticosteroids. Generally, two main treatment regimens are either prednisolone alone or prednisolone and azathioprine. Tacrolimus, cyclophosphamide, and mycophenolate mofetil are other options in patients unresponsive to these regimens.[6] Combination therapy of prednisolone and azathioprine is well supported by the American Association for the Study of Liver Disease (AASLD) and the British Society of Gastroenterology (BSG).[7]

CONCLUSION

Autoimmune hepatitis is uncommon in the pediatric population. AIH is treatable and measures should be taken to prevent complications like cirrhotic liver and liver failure.

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