(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 7, Issue 3 , Page No: 407-409
May-June 2024

IJMSCR

A Case Of Autoimmune Hepatitis

[1]Dr. Lahari Sri, [2]Dr. Prithi Inamdar
[1]Junior Resident, [2]Associate Professor,
Department of Paediatrics, DY Patil Medical College

***Corresponding Author:**
Dr. Lahari Sri
Junior Resident, Department Of Paediatrics, DY Patil Medical College

Type of Publication: Case Report
Conflicts of Interest: Nil

Abstract

Autoimmune hepatitis is a chronic inflammatory process of the liver, secondary to loss of immune tolerance against liver antigens resulting in progressive destruction of hepatic parenchyma and subsequent fibrosis. There are 3 types of autoimmune hepatitis- based on circulating antibodies. AIH is rare and very aggressive in children, progresses rapidly unless immunosuppressive treatment is started promptly. With appropriate treatment some 80% of patients achieve remission and long-term survival. Here we present a 9 year old female child with complaints of fever since 20 days, vomiting since 6 days and jaundice since 6 days with history of similar complaints in the father and uncle (jaundice) lasting for 1 month. Investigations suggested of increasing trends of conjugated bilirubinemia, serum aminotransaminase and deranged coagulation profile. Viral hepatitis markers, Weil Felix test were negative. Serum ceruloplasmin levels were within normal limits. Suspecting autoimmune hepatitis, ANA was found to be weakly positive and ASMA was positive. On further evaluation liver biopsy was done confirming the diagnosis of autoimmune hepatitis type 1. Child was started on steroid-prednisone which decreased the trend of bilirubin and normalized values of prothrombin and APTT. Now the child is doing well and is on tapering doses of prednisolone. A good clinical suspicion and an early diagnosis and treatment has improved the quality of life and prevented the need of liver transplantation.

Keywords: Autoimmune hepatitis, immunosuppressive drugs, liver transplant

Introduction

Autoimmune hepatitis is a rare, progressive inflammatory liver disorder characterized serologically by high transaminases and immunoglobulin G (IgG) levels, presence of autoantibodies, and a histological picture of interface hepatitis (i.e. dense inflammatory mononuclear/plasma cell infiltrate of the portal tracts that invades the parenchyma), in the absence of a known aetiology. Incidence of autoimmune hepatitis is 0.9-2/1,00,000 population per year and prevalence of 11-25/1,00,000 population per year.

Based up on the circulating antibodies there are 3 types of autoimmune hepatitis. Type 1 is distinguished by presence of anti-smooth muscle antibodies (ASMA) with/without anti-nuclear antibody, affecting both children and adults. Patients present with impaired hepatic, synthetic function [prolonged prothrombin and hypoalbuminemia], raised IgA levels, and has cirrhosis as initial presentation on liver biopsy along with interface hepatitis suggesting a more chronic disease compared to other types AIH-2 is predominantly a pediatric condition affecting age of 2-14 years presents with positive anti liver kidney microsome [LMK]antibody and or anti liver cytosol antibody IgG is usually raised. But IgA deficiency is common in AIH-2 and presents more frequently with fulminant hepatic failure. In both types, about 20% of patients have associated autoimmune disorders- including thyroiditis, vitiligo, type 1 diabetes,

inflammatory bowel disease, and nephrotic syndrome—and about 40% have a family history of autoimmune disease. Type 3 is characterized by presence of anti-soluble antigen in the serum, directed against cytokeratin 8 and 18.this is the rarest type and affects adult age. AIH usually responds to immunosuppressive treatment, which should be instituted as soon as the diagnosis is made. If left untreated, AIH generally progresses to liver failure requiring transplantation. We present a case of autoimmune hepatitis with timely diagnosed has prevented liver transplantation

Clinical Presentation

We present a 9-year-old female child, born of non-consanguinous marriage with complaints of fever since 20 days, vomiting since six days, yellowish discoloration of eyes since six days and high colored urine since six days with history of a jaundice in father and uncle, which lasted for one month. On presentation, child had signs of some dehydration with decreased oral acceptance, with icterus and no signs of liver cell failure hence started on symptomatic treatment with IV fluids and on hepatic drip. On deep palpation liver was felt in right hypochondrium 7 cm below the right subcostal margin, in mid clavicular line, with smooth surface and rounded edges, firm in consistency and non-tender and liver span of 10 cms.

On bimanual palpation spleen was palpable which was 2 cm with sharp edges. Relevant investigations were send to rule out infective etiology in which viral markers were negative, and Weil Felix test was negative. Serial serum Bilirubin monitoring was done, in which Conjugated hyperbilirubinemia was found to be on increasing trends.

Total bilirubin- 15.3>16.1>20.3>21.6mg/dl [N- 0.05-0.4mg/dl]

Direct bilirubin- 11.1>12.50>14.9>20.11 [N= 0.05-0.2mg/dl]

Indirect bilirubin- 4.2>3.60>5.4>1.5 [N= 0.2-0.8mg/dl]

Serials Serum aminotransaminases were on higher range exceeding greater than 1000 IU/L

Sgot-1077>1365>1232>1126IU/L [N= 21-44IU/L]

Sgpt- 1132>809>720>658IU/L [9-25IU/L]

Alp- 454>357>279>292 IU/L [N= 100-320IU/L]

Serial coagulation profile was deranged, hence vitamin k was given for 3 consecutive days

Prothrombin time [17.2-20-16.4]

Aptt [39.71— 44.68]

Serum Ammonia was sent, which was within normal limits 72.50 [30.7 to 122.6]. To prevent hepatic encephalopathy, patient was started on ursodeoxycholic acid and L-ornithine and L aspartate medications. Fund examination was done to rule out Kayser Fleischer ring -which was negative and serum ceruloplasmin was 19.7 mg per dl [N= 20.5 to 40.5] suspecting auto immune hepatitis ANA by immunofluorescence was done, which was weakly positive.

Anti-smooth muscle antibody was positive, suggesting the diagnosis of type 1 auto immune hepatitis. LKM1, pANCA, cANCA were negative. Confirmation was done by liver biopsy, suggestive of chronic -hepatitis with cholestasis with peripheral fibrosis. Moderate portal inflammation, with lymphocytes, plasma cells and few eosinophils were present. Child was started on Oral Prednisolone 2mg/kg/day and by three weeks there was decreasing trends of serum bilirubin and serum transaminase. Patient responded well to the treatment and is on tapering dose of steroids.

Discussion

Autoimmune hepatitis is a clinical constellation that suggests an immune-mediated process; it is responsive to immunosuppressive therapy. It typically refers to a primarily hepatocyte-specific process, whereas autoimmune cholangiopathy and sclerosing cholangitis are predominated by intra- and extrahepatic bile duct injury. Overlap of the process involving both hepatocyte and bile duct directed injury may be more common in children. It is a dense portal mononuclear cell infiltrate invades the surrounding parenchyma and comprises T and B lymphocytes, macrophages, and plasma cells. The immunopathogenic mechanisms underlying autoimmune hepatitis are unsettled. Triggering factors can include molecular mimicry, infections, drugs, and the environment (toxins) in a genetically susceptible host. Several human leukocyte antigen class II molecules, particularly DR3, DR4, and DR7 isoforms, confer susceptibility to autoimmune hepatitis. Heterozygous mutations in the autoimmune regulator gene (AIRE), which encodes a transcription factor

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controlling the negative selection of autoreactive thymocytes, can be found in some children with autoimmune hepatitis types 1 and 2 AIRE mutations also cause autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy (also called autoimmune polyen-docrinopathy syndrome) in which autoimmune hepatitis occurs in approximately 20% of patients. The histologic features include interface hepatitis, bridging and piecemeal necrosis in 25-30% of patients with autoimmune hepatitis, particularly children, the illness mimics acute viral hepatitis. Patients can be asymptomatic or have fatigue, malaise, behavioral changes, anorexia, and amenorrhea, sometimes for many months before jaundice or stigmata of chronic liver disease are recognized. Extrahepatic manifestations can include arthritis, vasculitis, nephritis, thyroiditis, Coombs-positive anemia, and rash. Some patients' initial clinical features reflect cirrhosis (ascites, bleeding esophageal varices, or hepatic encephalopathy). There may be mild to moderate jaundice, oedema and ascites in severe cases. It is a clinical diagnosis based on certain diagnostic criteria; no single test will make this diagnosis. Diagnostic criteria with scoring systems have been modified for children. Important positive features include female gender, primary elevation in transaminases and not alkaline phosphatase, elevated y-globulin levels, the presence of autoantibodies (most commonly antinuclear, smooth muscle, or liver-kidney micro-some), and characteristic histologic findings. Important negative features include the absence of viral markers (hepatitis B, C, D) of infection, absence of a history of drug or blood product expo-sure, and negligible alcohol consumption. Common conditions that might lead to chronic hepatitis such as Wilson's disease, alpha anti-trypsin deficiency, peri cholangitis, has been ruled out.

Prednisone, with or without azathioprine or 6-mercaptopurine, improves the clinical, biochemical, and histologic features in most patients with autoimmune hepatitis and prolongs survival in most patients with severe disease. The goal is to suppress or eliminate hepatic inflammation with minimal side effects. With prednisone (2 mg /kg /day) serum transaminase and serum bilirubin values returned to normal within 3 weeks and prothrombin time within 2 weeks

Patient responded well to treatment and advised to follow up

Conclusion

We are presenting this case to highlight the importance of early recognition and treatment of auto immune hepatitis to reduce morbidity and mortality, and the need of liver transplantation.

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