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Case Report

Autoimmune Hepatitis in Children: A Case Report and Literature Review

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Abstract:

Autoimmune hepatitis (AIH), is an relatively uncommon cause of liver disease, especially in children. It has wide range of presentation like asymptomatic rise of liver enzyme to advance stages of liver disease. When other causes of liver disease ruled out, AIH should always to taken into account. We presenting the case of 9-year-old girl, who had manifestations of liver disease and we diagnosed her as a case of AIH.

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Introduction:

Autoimmune hepatitis (AIH), a chronic inflammatory condition with unknown etiology, The hallmarks of the disease include hypergammaglobulinemia and/or circulating auto-antibodies, profound changes in hepatic histology, and a marked response to immunosuppressive treatment[1]. Women are affected more frequently than men, 75% of patients of AIH are female, and the disease is seen in all ethnic groups and at all ages.[2] In Bangladesh, Benzamin et al. found that, about 8% of children with chronic liver disease with hepatomegaly and or splenomegaly are due to AIH.[3]

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In accordance with serological profile, AIH are two types; AIH-1 is defined as positive for antinuclear antibody (ANA) or anti-smooth muscle antibody (SMA) or both; AIH-2 is defined as positivite for anti-liver kidney microsomal type 1 antibody (anti-LKM-1) or for anti-liver cytosol type 1 antibody (anti-LC-1) or both.[4]

Case Report:

A 9-year old immunized girl, 2nd issue of non-consanguineous parents, got admitted at Pediatrics Department, Sylhet M AG Osmani Medical College Hospital, with complaints of jaundice for 3 months, abdominal distension for same duration and nausea with anorexia for 6 months. She had no history of gastrointestinal bleeding, abdominal pain, altered sleep pattern, rash, arthralgia, blood transfusion, any surgical procedure, family history of such type of illness, and any offending drug exposure. On examination she had jaundice and splenomegaly. Her vital signs were within normal limit and anthropometrically she was well thriving. Investigations showed raised serum bilirubin, serum ALT and thrombocytopenia. Ultrasonography showed coarse hepatic parenchyma with splenomegaly. Investigations for hepatitis B virus, hepatitis C virus and

Wilson's disease were negative. ANA was positive, anti LKM1 was negative. Serum TSH was elevated but serum T3 and T4 were normal. Endoscopy of upper gastrointestinal tract showed Grade II esophageal varices (fig1) and histopathology of liver tissue compatible with autoimmune hepatitis with cirrhosis. So finally we diagnosed her as a case of chronic liver disease (CLD) with autoimmune hepatitis with portal hypertension with subclinical hypothyroidism. We treated her with Tab. Prednisolone 2mg/kg/day and Tab. Azathioprine 1-2mg/kg/day were added 2 weeks later of prednisolone started. Tab. Propranolol 1mg/kg/day was given as primary prophylaxis of portal hypertension and Tab. Levothyroxine 1.5 mcg/kg/day was started for subclinical hypothyroidism.

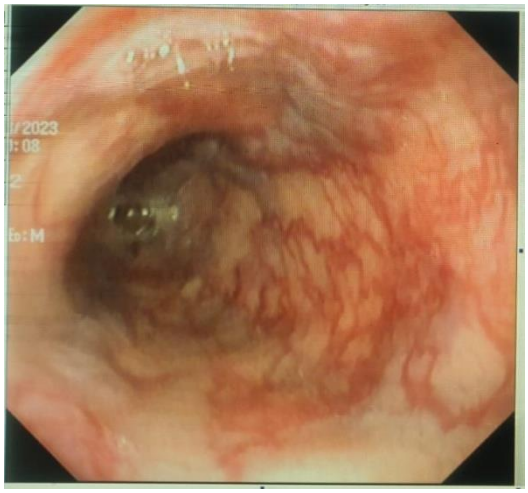

Fig 1: Oesophageal varices

Discussion:

AIH has a variety of clinical presentations. Range of presentations includes non-specific symptoms like progressive fatigue, malaise, nausea, anorexia, vomiting, joint pain, amenorrhea, headache, abdominal pain to acute hepatitis, relapsing jaundice or fulminant hepatic failure or chronic liver disease, complication of cirrhosis and portal hypertension without history of previous liver disease and jaundice. Non specific symptoms may last for 6 months to few years before diagnosis. Incidental finding of raised ALT without sign symptoms is another presentation of AIH.[4]Our case presented with non specific symptoms such as anorexia, nausea for 6 months followed by jaundice for 3 months. With these symptoms she visited to several physicians and evaluated for common liver diseases, but remained undiagnosed. Suspicion of autoimmune hepatitis always to be considered after excluding all causes of acute and chronic hepatitis. The diagnosis of AIH requires compatible histological findings and is further supported by the following features: (1) elevated serum aminotransaminase levels (ALT/AST); (2) elevated serum IgG level and/or positive serological marker(s); (3) exclusion of viral, hereditary, metabolic, cholestatic, and drug-induced diseases that may resemble AIH. Initial serological testing should include ANA, SMA, anti-LC-1 and anti-LKM1 in children; consider additional autoantibody tests if warranted to secure the diagnosis.[5]In our case, we found elevated serum ALT, ANA positive, histology suggestive of AIH and we excluded viral causes and Wilson disease. She also had portal hypertension, as evidenced by oesophageal varices. Histopathology of liver tissue is one of the key diagnostic tools for diagnosis of AIH. Regardless of serologic type, the classic histological findings on liver biopsy are similar. Biopsy shows portal and lobular inflammation, composed of lymphocytes and, most commonly, conspicuous plasma cells. The portal inflammation typically creates "interface hepatitis" where the limiting plate is disrupted and the inflammation involves the adjacent liver parenchyma; concomitant hepatocyte necrosis may or may not be present. Lobular inflammation can range from mild to severe, with either scattered necrotic hepatocytes, or in more severe cases centrilobular and/or bridging necrosis. Rosette formation and emperipolesis are two uncommon characteristics.[6]In our case, histopathology of liver tissue was consistent with autoimmune hepatitis. In both types, a family history of autoimmune disease is frequent (40%) and approximately 20% of patients have associated auto immune disorders either present at diagnosis or developing during follow-up, including thyroiditis, inflammatory bowel disease (IBD), hemolytic anemia, vitiligo, celiac disease, insulin-dependent diabetes, Behchet disease, Sjo¨gren syndrome, glomerulonephritis, idiopathic thrombocytopenia, urticaria

pigmentosa, hypoparathyroidism, and addison disease (mainly in AIH-2).[4]Our case have no family history of autoimmune disease but she had hypothyroidism. But autoimmune thyroiditis was not rule out. After diagnosis, prompt treatment should be started. AIH is responsive to immunosuppressive drug. Treatment consists of two parts- induction of remission and maintenance phase. With the exception of a fulminant presentation with encephalopathy, AIH responds satisfactorily (remission rate of up to 90%) to immunosuppressive treatment whatever the degree of liver impairment. Conventional treatment of AIH consists of prednisolone 2mg/ kg/day (maximum 60mg/day), which is gradually decreased during a period of 4 to 8 weeks, in parallel to the decline of transaminase levels, to a maintenance dose of 2.5 to 5mg/day. In most patients, an 80% decrease of the aminotransferase levels is achieved in the first 2 months, but their complete normalization may take several months.[4]In our case, Tab. Prednisolone 2mg/kg/day and Tab. Azathioprine 1-2mg/kg/day was added 2 weeks after starting prednisolone. On follow up after 2 month, her bilirubin was 3.6 mg/dl and ALT was 103 U/L. That's indicating partial improvement.

Investigations	Findings	Normal
CBC (02/08/2023)	Hb 13.1 gm/dl MCV 82.9 fL MCH 27.7 pg WBC 4800 per cumm. Platelet 100000 per cumm.	Male- 13-17 gm/dl , Female 12-15 gm/dl 83-101 fL 27-32 pg 4000-11000/cumm. 150000-400000/cumm
PBF	No evidence of haemolysis, Thrombocytopenia	
SGPT	154U/L	Female <35U/L
S.Bilirubin	4 mg/dl	Female: mg/dl
Prothrombin time with INR	Patient 14 Sec. Control 12 Sec. Index 85.7% Ratio 1.16:1 INR 1.20	
S.Albumin	1.9 mg/dl	3.5-5 mg/dl
USG of HBS	Coarse hepatic parenchyma with splenomegaly	
HbsAg	Negative	
Anti HCV	Negative	
S.Ceruloplasmin	319 mg/L	200-600 mg/L
Eye evaluation for KF ring	No K-F ring or Sun-flower cataract	
S. IgG	Not done	
ANA	96.1 AU/ml	>40 AU/ml Positive
Anti LKM1 antibody	Negetive	
Anti Ds DNA	15.20 IU/ml	> 25 IU/ml Positive
S.TSH	10.4 mIU/L	Children (2-12 yrs):0.72-4.4 mIU/L
FT4	1.82 ng/dl	0.78-2.19 ng/dl
Endoscopy of UGIT	Oesophageal varices	

Histopathology of Liver Tissue	Hepatocytes had mild feathery degeneration, the portal tract shows evidence of bile duct injury, infiltration of lymphocytes, plasma cells, some polymorphs and marked fibrosis including bridging fibrosis, the fibrous bands have encircled the hepatic plates forming cirrhotic nodules, mild piecemeal necrosis, interface hepatitis and emperipoiesis of hepatocytes by lymphocytes are seen, no dysplasia or malignancy are seen.	

Conclusion:
AIH is relatively a rare disease; it remains a diagnostic& therapeutic challenge, especially among the children and adolescents. All unexplained liver disease should screen for AIH, which will make early diagnosis and will ensure good outcome.

Ethical issue: Inform written consent was taken from parents.

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