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A Case of Seronegative Autoimmune Hepatitis

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Abstract: *Autoimmune hepatitis occurs when own immune system attacks hepatocytes of an individual causing inflammation with symptoms of fatigue or muscle aches or signs of acute liver inflammation like fever,jaundice & right upper quadrant abdominal pain.The disease most often diagnosed in females between age 40-50yrs.Usually a number of specific antibodies found in blood like antinuclear antibody (ANA),anti-smooth muscle antibody (ASMA),liver/kidney microsomal antibody (LKM),anti soluble liver antigen and liver-pancrease antigen(SLA/LP).But here we found a case otherwise consistent with Autoimmune hepatitis without the presence of autoantibodies.*

Patient was started on Prednisolone and Azathioprine with other supportive treatment and showed clinical improvement.Liver biopsy repeated after 6 months and 2 yrs showed histological regression of HAI (3/18).Patient is on maintenance dose of Azathioprine (50mg) and is on regular follow up for last 3 yrs.

Key words: *Autoimmune hepatitis,Antinuclear antibody,Anti-smooth muscle antibody,Liver kidney Microsomal Antibody,Anti-soluble liver antigen & liver pancrease antigen.*

• Introduction

Autoimmune hepatitis is a chronic autoimmune disease of the Liver that occurs when the body's immune system attacks liver cells causing the liver to be inflammed .Common initial symptoms include fatigue or muscle aches or rashes or signs of acute liver inflammation including fever,jaundice&right upper quadrant abdominal pain.Individuals with autoimmune hepatitis often have no initial symptoms & the disease is detected by abnormal liver function test.(1)Abnormal immune response results in inflammation of the liver,which can lead to further symptoms and complications which ranges from fatigue to Cirrhosis.(2)The disease may occur in any ethinic group and at any age ,but is most often diagnosed in patients between age 40-50.(3)Autoimmune hepatitis is more common in females.(4)A number of specific antibodies found in the blood like ANA,ASMA,LKM-1,LKM(LKM-1,LKM-2,LKM-3),SLA/LP and AMA are of use.There is also increased level of IgG.However the diagnosis of autoimmune hepatitis always requires a liver biopsy.(5)Liver biopsy shows presence of interface hepatitis.(6)

Autoimmune hepatitis may have diverse clinical presentation leading to difficult treatment decision and therapeutic judgment especially when lack typical auto antibodies.A case was encountered who had features of autoimmune hepatitis without the presence of autoantibodies which needed appropriate therapeutic judgement before starting immunosupressants.

• Case Report

A 40yrs old female presented with Dysphagia,Fever and Fatigue with urinary symptoms for 2 weeks.She was treated elsewhere in the line of urinary tract infection with Ofloxacin according to culture sensitive report.She developed rashes on extremities and trunk following treatment outside.Sonography of abdomen revealed retroperitoneal lymphnodes.She was non diabetic and non hypertensive.On admission she was found to be febrile,had maculo-papular,pruritic rashes and was lethargic.However abdomen and other systemic examination were normal.Routine investigations revealed abnormal liver biochemical test i.e T.bil:1.5mg/dl,AST-Aspartateaminotransferase(SGOT):565u/l.

ALT(SGPT):298u/l,SAP:186u/l,GGTP:133u/l,T.Protein:6.2g/dl,Alb:2.9g/dl,Globulin:3.3g/dl,A:G (Albumin:Globulin)ratio 0.9.PT/INR was normal (13.6/13.sec,1.01).CBC,KFT,Urine routine were normal.Viral markers for hepatitis A,B,C and E were all negative.USG abdomen revealed hepatomegally with GB wall thickening and edema with periportal and portocaval subcentric lympnodes.Upper GI Endoscopy revealed Esophageal candidiasis without any evidence of Portal hypertension.

ANA(IgG,IIFA,Indirect)ASMA,LKM1 along with SLA and p-ANCA were all negative.Liver biopsy was done revealed Interface hepatitis(+3/4),Confluent necrosis(+1/6),Focal lytic necrosis and Inflammation(+2/4) and portal inflammation(+3/4) (Knodell HAI).In the mean time patient started deteriorating clinically and increase in lethargy and drowsiness was found.

Patient was started on Prednisolone and Azathioprine with continuation of other supportive treatment.Within 3 days,she showed signs of clinical improvement and was discharged after 7 days.On follow up,she was fine.Liver biopsy was repeated after 6 months and 2 yrs showed significant histologic regression of HAI-Histological Activity Index (3/18).ANA,ASMA and LKM were repeated after 1 yr and found to be negative.She developed hypertension and other features of Glucocorticoid intolerance,hence is on maintenance dose of Azathioprine(50mg) and is on regular follow up for last 3 yrs.

• Discussion

The aim of pharmacotherapy are to reduce morbidity and to prevent complications.Treatment of autoimmune hepatitis includes high dose Prednisolone monotherapy or combination of Prednisolone and Azathioprine.(7,8) Corticosteroid therapy may decrease the disease progression to either fulminant hepatic failure or cirrhosis.It may decrease inflammation by reversing increase capillary permeability and supressing polymorphonuclear neutrophil activity.It stabilizes lysosomal membranes and also suppresses lymphocyte and antibody production.Azathioprine antagonizes purine metabolism and inhibits synthesis of DNA,RNA & Proteins.It may decrease proliferation of immune cells which results in lower immune activity.

The above case is suggestive of Autoimmune hepatitis with abnormal liver enzymes and interface hepatitis on liverbiopsy but absence of autoantibodies.So proper therapeutic judgement was done to start with Prednisolone and Azathioprine with other supportive treatment which responded well within 3 days.And the patient is on regular follow up for 3 yrs and is on maintenance dose of Azathioprine (50mg) due to developement of hypertension and features of Glucocorticoid intolerance.

• Conclusion:

Seronegative autoimmune hepatitis has similar demographic,biochemical and histological features of autoimmune hepatitis.They may be treated effectively with Corticosteroids and Azathioprine.Early recognition and judgement needed for treatment of seronegative autoimmune hepatitis to prevent progression to end stage liver disease.

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