J MEDICINE 2022; 23: 162-164

Case Reports

A Case of Systemic Lupus Erythematosus with Severe Jaundice

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Abstract:

Abnormal liver tests are common (60%) in some point of Systemic Lupus Erythematosus (SLE) illness. In rare cases, severe cholestasis may invite diagnostic dilemma. Here we reported a 48-years-old female SLE patient with severe cholestasis (serum bilirubin 37mg/dl) and her management in a financial constrain situation.

Key words: *Systemic lupus erythematosus (SLE),Autoimmune hepatitis (AIH), Severe jaundice, Cholestatic jaundice, Ursodeoxycholic acid (UDCA).*

DOI: https://doi.org/10.3329/jom.v23i2.60634

Received: *11 September 2021;* ***Accepted:*** *10 May 2022*

Introduction

SLE is a multifaceted autoimmune disorder with various organ involvement including musculoskeletal, kidney and central nervous system[1]. It is most common in female and the female to male ratio is 9:1[2]. Liver involvement is around 60% in some point of this disease but rare as a part of its disease activity. Nonimmune hepatopathy e.g. hepatotoxic drugs, coincident viral hepatitis and non-alcoholic fatty liver disease are some common possibilities of abnormal liver function test in SLE. But rarely lupus hepatitis or overlap syndrome of autoimmune hepatitis (AIH) may complicate this disease[3]. One study reported 9.3% incidence of lupus hepatitis[1]. However, the prevalence is widely variable 3-23%[4]. On the other hand, AIH is a chronic inflammatory disease of the liver of unknown etiology with female to male ratio 4:1[5]. Diagnosis based on elevated IgG, specific autoantibodies, characteristic histology in absence of viral hepatitis. Patients may present with nausea, anorexia, abdominal discomfort and jaundice and sometimes with acute fulminant hepatic failure[6]. Arthralgia is a common feature in both SLE and AIH. Anti-nuclear antibodies (ANA) and raised IgG may found in both conditions but anti-double stranded DNA (anti-dsDNA) is highly associated with SLE. Soluble liver antigen (SLA), anti-liver kidney microsomal antibody (anti-LKM) and anti-smooth muscle antibody (ASMA) are the specific marker for AIH[3]. Liver histology may differentiate AIH from lupus hepatitis in diagnostic dilemma[3]. To date, there is scarce study on lupus hepatitis. Here we reported a case on severe cholestatic hepatitis in a patient with SLE.

Case presentation:

A 48-years-old female hypertensive, CKD stage 2, SLE patient was in remission with HCQ and prednisolone for 7 years. One year back while on the treatment of UTI she developed jaundice, anorexia, nausea, epigastric pain, pale stool, generalized itching along with hematemesis and melaena. On examination she was deeply icteric, anemic, BP-90/60 mmHg, Pulse 110/min associated with epigastric tenderness, mild hepatomegaly without ascites. Her Hb% was 5.7 gm/dl with normal RBC indices, TC 8000/cumm, neutrophil 88%, lymphocytes 5%, platelet 59,000/cumm, reticulocyte 4% and

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Table 1: *Liver function tests*

Date	Direct Bilirubin (mg/dl)	Indirect Bilirubin (mg/dl)	Total Bilirubin (mg/dl)	ALT (U/L)	AST (U/L)	ALP (U/L)	S. Albumin (gm/L)	Prothrombin Time (INR)
08.06.20			8.7	201				
10.06.20			9.9	122		1053		
14.06.20			12.2		211	917		
15.06.20			13.3		285	980		16.7 (1.5)
16.06.20						761	20	38.9 (3.17)
19.06.20			17.98					
20.06.20			18.74	47	222	1504		
22.06.20			22					
24.06.20							23	12.9 (1.08)
26.06.20			28.5					
29.06.20			31.41					
01.07.20	19.40	12.12						
05.07.20	21	16	37	837		1929		
07.07.20								15 (1.22)
08.07.20	22.11	10.99	33.10					
11.07.20			18.9					
12.07.20				198		622		
21.07.20				104		224		
27.07.20			3.3					

nonspecific PBF. Hb Electrophoresis and Coombs' test were normal. S. creatinine 2.2 mg/dl, CRP 46.6 mg/dl and normal Blood and Urine C/S. Baseline bilirubin was 8.7 mg/dl and in around 2 weeks' time increased to 37.0 mg/dl. Her GGT was 443 U/L and other liver function tests in different times have shown in table 1. Viral markers HAV, HEV, HBV, HCV, CMV, EBV, and HSV were negative. Acute hepatitis, mild hepatomegaly, no biliary obstruction, no ascites and evidence of bilateral renal parenchymal disease was observed in ultrasonogram. The ANA, anti-dsDNA and anti-smooth muscle Ab tests were positive. The anti-LKM-1 and anti-mitochondrial antibodies (AMA) were negative. Upper GIT endoscopy showed antral erosions and congestive gastropathy. Colonoscopy was normal. Wilson's disease panel, MRCP and liver biopsy were not done for financial constrain. She was diagnosed as a case of lupus hepatitis with severe cholestasis and treated with hydroxychloroquine (HCQ), blood transfusion, IV Ceftriaxone, IV Dexamthasone 5mg daily, IV omeprazole, IV albumin and inj Vit K. For cholestatic hepatitis she was treated with Ursodeoxycholic acid 300 mg BID and cholestyramine 8gm daily. After 1 month, S. bilirubin was 5.0 mg/dl, ALT 104 U/L, AST 92 U/L, ALP 224 U/L, and GGT 84 U/L. During subsequent and at one year visit her liver functions were within normal range, no flare of SLE with HCQ and 5 mg prednisolone.

Discussion

Liver involvement in SLE is common but the prevalence of lupus hepatitis is rather a wide range. Prevalence of lupus hepatitis is more common in active disease than inactive SLE (11.8 vs 3.2%)[1]. Absence of viral hepatitis, NAFLD and use of hepatotoxic drugs raised the possibility of diagnosis of lupus hepatitis in this patient[3]. According to Simplified diagnostic criteria for the diagnosis of AIH, score of this patient is only 3 (positive ASMA for +1 and absence of viral hepatitis +3)[7]. According to serology, AIH is further subdivided into 2 types: type 1 positive for anti-nuclear antibody (ANA) and/or anti-smooth muscle antibody (ASMA), while AIH-type 2 is positive for anti-liver kidney microsomal antibody type 1 (anti-LKM1) and/or anti-liver cytosol type 1 (anti-LC1)[8]. ANA and ASMA both also can positive in SLE[3]. The more specific anti-LKM-1 for AIH was negative and anti-ds-DNA which is more specific for SLE

was positive here. The similar clinical and biochemical features of lupus hepatitis and AIH make these conditions difficult to differentiate, however their treatment and prognosis differ[3]. AIH has more aggressive histological features and a poor prognosis than lupus hepatitis. Untreated AIH has poor outcome with 5 years survival rate is 50%[9]. Liver histology is important for differentiation between lupus hepatitis and AIH. AIH have characteristics histopathologic features like interface hepatitis, lymphocyte infiltration, resetting of hepatocytes, empaeriopolesis and fibrosis to cirrhosis[10]. However, liver histology in SLE patients shows nonspecific changes like fatty degeneration or hydropic hepatocytes or features of drug toxicity[3]. In this subject, it was not possible to do other autoantibodies test and the most important liver biopsy due to financial constrain. High dose prednisolone (1-2mg/kg daily) and azathioprine is the mainstay of treatment of AIH[11]. About 85% patients required azathioprine as a steroid sparing agent[12]. The treatment of SLE is individualized and depends on organ involvement, disease activity, disease severity and previous response of treatment[13]. This patient was respond well with only i.v dexamethasone 5mg equivalent to 34 mg prednisolone. Severe cholestasis is rare in the both conditions though well managed here with ursodeoxycholic acid and cholestyramine. Again, AMA negativity might exclude the possibility of primary biliary cirrhosis in this case.

Conclusions

Liver biopsy is crucial for the diagnosis of lupus hepatitis and AIH. In a financial problem, high clinical suspicion for diagnosis and appropriate treatment can improve the disease outcome in a patient with SLE with abnormal liver functions tests.

Ethical statement

The patient provided all of the clinical and laboratory information and agreed to the case publication.

Consent to publish

Written informed consent was obtained from the patient for publication of this case report

Author contributions

NF and JG drafted the initial manuscript, designed the study, interpreted the data, screened the literature, and approved the final manuscript for submission. NI provided recommendations for this manuscript. NI conceptualized the study, reviewed and revised the manuscript, and approved the final manuscript for submission. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

Acknowledgements

We sincerely thank the patient and her husband for providing all of the clinical and laboratory information.

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