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CASE REPORT

Systemic Lupus Erythematosus in a Male Patient With Autoimmune Hepatitis

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ABSTRACT

Overlap syndrome involving autoimmune hepatitis (AIH) and systemic lupus erythematosus (SLE) is a rare condition. The difference between the hepatic involvement of SLE and autoimmune hepatitis has not been clearly defined due to the presence of clinical and laboratory common features. We report a 42-year-old male patient with autoimmune hepatitis who presented with intermittent fever, jaundice, and dyspnea. CT scan showed pleural and pericardial effusion, high titer of antinuclear antibodies and positive anti-DNA antibody were found. Our patient fulfilled both the international criteria of SLE and AIH. Clinical symptoms and laboratory findings improved with treatment by corticosteroids.

Key words: Cirrhosis; Autoimmune hepatitis; Lupus; Overlap

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INTRODUCTION

Autoimmune hepatitis is a chronic inflammatory liver disease that affects mainly women in early adulthood. The usual presentation includes non-specific symptoms such as fatigue, general ill health, right upper quadrant pain, nausea, pruritus, and fluctuating jaundice[1]. Immunosuppression, generally results in clinical and laboratory improvement[2]. The disease, if untreated, often leads to cirrhosis and liver failure. Other autoimmune diseases may be associated with AIH such as primary biliary cholangitis (PBC), celiac disease, autoimmune thyroiditis and SLE. SLE is a systemic autoimmune disease, with multisystemic involvement and several presentations[3]. The term overlap syndrome is usually used in the context of overlap of AIH with PBC. Overlapping case of SLE and AIH has been occasionally diagnosed and reported. In this report, we present a patient with an overlap syndrome involving autoimmune hepatitis and SLE.

CASE REPORT

A 42-year-old male without a notable personal history, and without a family history of autoimmune diseases, presented in November 2019 with liver dysfunction and ascites. Laboratory data showed: Serum alanine aminotransferase (ALT) 230 IU/L (N: 5-40 IU/L), aspartate aminotransferase (AST) 390 IU/L (N: 5-40 IU/L), gamma-glutamyl transpeptidase (GGT): 50 IU/L (N: 5-40 IU/L), alkaline phosphatase 80 IU/L (N: 35-140 IU/L), total bilirubin 44 µmol/L (N: 3-17 µmol/L), conjugated bilirubin 39mg/L (N:1-3 µmol/L), and a low prothrombin activity at 37%. Hemoglobin rate was 14 g/dL White blood cells: 4030 elements/mm^3 Platelets: 140 000 elements/mm^3.

Positive results for serum antibodies against nuclear antigen (ANA): 1/800 IU/L and anti-double-stranded DNA. Anti-soluble liver antigens/liver-pancreas (SLA/LP) were also positive. Gamma globulin levels were very high at 45 g/L (N: 8-13.5 g/L). Viral serology was negative for hepatitis B and C virus.

The patient was diagnosed to have probable AIH according to diagnostic criteria for AIH.

A liver biopsy was not performed due to the presence of ascites and

coagulopathy. Prednisolone therapy was not used because the patient had an advanced decompensated cirrhosis at the time of diagnosis.

In July 2020, the patient was admitted for fever with cough and dyspnea. On physical examination he had: Fever at 38.3°C, Pulse: 74 bpm, jaundice and large volume ascites. The laboratory findings showed: White blood cells: 7400 E/mm^3 Hemoglobin: 10.7 g/dL, platelets: 134 000 /mm^3; CRP: 90 mg/L; AST: 57 IU/L, ALT: 21 IU/L, GGT: GGT: 14 IU/L, ALP: 76 IU/L, Total bilirubin: 73 µmol/L Direct bilirubin: 17 µmol/L; a positive direct coombs test, C3: 0.25 g/L (VN: 0.9-1.8 g/L) C4: 0.05 (VN: 0.1 - 0.4 g/L) positive ANA: 1/1600 UI /L positive native anti DNA bodies.

Chest X-ray (Figure 1) and computed tomography showed pericardial and pleural effusion. These clinic-biological data allow us to make the diagnosis of SLE according to SLICC criteria and EULAR/ ACR 2019 criteria.

The Patient was treated by corticosteroid: prednisolone 0.5 mg/kg with good outcome: disappearance of fever, regression of biological inflammatory syndrome, marked decrease in bilirubin and decrease in pleural effusion.

DISCUSSION

Systemic lupus erythematous (SLE) is a multisystem autoimmune disorder involving various organs including the liver, skin, kidney, and joints. Increased serum levels of liver enzymes are common in SLE[4]. This can reflect liver disease as a component of SLE or a liver disorder associated with another immune disease such as AIH.

Autoimmune hepatitis is a chronic progressive inflammatory liver disorder of unknown etiology with fluctuating course and potentially progressing to cirrhosis.

HAI and SLE overlap syndrome has previously been reported in some cases but it is considered to be rare however its exact frequency is not clear.

Our patient, was diagnosed with SLE according to the criteria of EULAR/ACR 2019 and to the SLICC criteria[5,6].

The criteria for the diagnosis of AIH in adult patients have been established by the International Autoimmune Hepatitis Group (IAIHG). Diagnostic criteria are based on elevation of Immunglobulin G (IgG), demonstration characteristic autoantibodies, histological features of hepatitis and the absence of viral disease[7] (Table 1). AIH score in our patient was 6 suggesting probable AIH.

Liver biopsy plays an important role in the diagnostic regimen. It would have been of major diagnostic and prognostic interest[8].

Histological findings in AIH are periportal piecemeal necrosis and hepatocyte rosette formation abundant plasma cell and lymphocyte infiltrate. Percutaneous Liver Biopsy was contraindicated for our patient due to the presence of ascites and coagulopathy[9].

Transjugular liver Biopsy (TJLB) is performed in patients when liver biopsy is essential for the diagnosis and management but percutaneous biopsy is contraindicated due to deranged bleeding parameters or ascites[10]. Nevertheless, TJLB is not available in our hospital.

The difference between the hepatic involvement in SLE and AIH has not been clearly defined both have features of an autoimmune disorder, such as the presence of hypergammaglobulinemia and positive tests for ANA. It is important to distinguish between these two entities because the prognosis and the treatment are different. Differential diagnosis between SLE hepatitis and AIH must be made in order to avoid potential complications associated with inadequate therapy.

We believe that our patient presents an overlap syndrome (AIH and SLE) because of the advanced liver disease at diagnosis:

Table 1 Simplified criteria for diagnosis of autoimmune hepatitis[7].

Feature /Parameter	Discriminator	Score
ANA ou SMA +	≥1 :40	1
ANA ou SMA +	≥1 :80	2
Or LKM1 +	≥1 :40	2
Or SLA/LP+		2
IgG ou Gamma globulins levels	> Upper limit of normal	1
	> 1.1 Upper limit of normal	2
Liver biopsy	Typical of HAI	2
	Compatible with AIH	1
Absence of viral hepatitis	Yes	2

Definite autoimmune hepatitis: ≥ 7; Probable autoimmune hepatitis: ≥ 6.

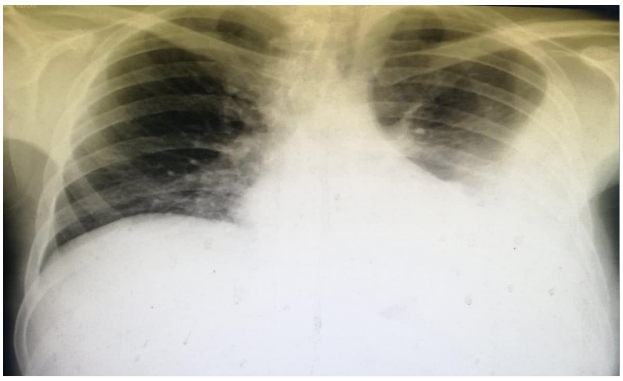

Figure1 Chest X-ray showing pleural effusion.

decompensated cirrhosis classified Child-Pugh C 10, the negativity of Anti- ribosomal P antibody, which are strongly correlated with SLE associated hepatitis[11], and the presence of specific markers for AIH, which usually do not occur in SLE: soluble liver antigen (SLA)[12].

Overlap syndrome (SLE and HAI) has been reported to respond rapidly to corticosteroid therapy[13], the prognosis is generally good[14].

AIH patients with decompensated cirrhosis constitute a subgroup with a poor prognosis and a high mortality rate[15].

For our patient the response to corticosteroid was good: The patient's condition improved rapidly with a medium dose of prednisolone. However, the prognosis is considered to be poor since the diagnosis was made at an advanced stage of chronic liver failure[15].

CONCLUSION

AIH and SLE with hepatic involvement are two autoimmune diseases that share several clinical and biological similarities. The association of these two diseases in one patient is rare. Autoimmune hepatitis is associated with a poor prognosis, higher risk of progression to cirrhosis. It's needs to be considered in SLE patients with elevated liver enzymes.

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