

Case report

The Liver In Systemic Lupus Erythematosus: A case report

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Summary

A case of systemic lupus erythematosus presented sequentially over ten years as, pancytopenia, pericarditis with pericardial effusion, recurrent psychosis, proteinuria with active sediment, and a positive ANA and Anti-dsDNA. The patient developed liver failure. The spectrum of hepatic manifestation of systemic lupus erythematosus is reviewed.

Introduction

Multi-organ involvement is the hallmark of systemic lupus erythematosus (SLE), but there is no consistent pattern of hepatic involvement. Hepatic disease is not a significant cause of morbidity and mortality, but subclinical liver involvement is common. A wide spectrum of hepatic disorders have been associated with SLE.(1). A correlation between aminotransferase elevation and lupus activity suggests that subclinical liver disease may be a manifestation of SLE. (2) However severe and even fatal disease may occur in SLE. (3).

Case report

A.K. a 50 years old male married Iraqi patient, presented with three months history of cholestatic Jaundice, abdominal distension, malaise, anorexia and weight loss associated with hematuria, loin pain with recurrent attacks of abnormal behaviour labelled as psychosis..

His past medical history is remarkable in that he developed the following problems:-

- 1- Insulin dependent diabetes mellitus (IDDM) in 1990.
- 2- Atelectasis of the right lung with paralysis of the right hemidiaphragm in 1995 , bronchoscopy was normal that time .
- 3- Sensory neuropathy mainly of lower limbs

proved by nerve conduction study in 1996 .

- 4- In 1997 splenectomy was done because of the presence of huge spleen with normal liver function and evidence of hypersplenism, (pancytopenia with high ESR (130 mm\hr) with normal bone marrow aspirate).
- 5- In 1999 pericardiectomy was done for Pericardial effusion, and histopathology showed chronic inflammatory cell infiltrate with no granuloma and no mesothelial cells.
- 6- Allergic rhinitis in 2000 treated with local and systemic medication.

No history of arthritis, red eye , skin rash, orogenital ulceration or hair fall. Physical examination showed jaundice, pallor, ascites, sensory neuropathy mainly of lower limbs.

Abdominal ultrasound showed a small liver with echogenic texture, irregular outline, moderate ascites.

Upper GI Endoscopy showed grade 2 oesophageal varices with fundal varices with portal hypertensive gastropathy and active duodenal ulcer.

Investigation showed total serum bilirubin 3.0 mg/dl, direct 2.4 mg/dl, Aspartate aminotransferase 12 iu/l, Alanine aminotransferase 10 :iu/l , alkaline phosphatase 17 KAU, prolonged prothrombin time (INR 2.2).

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Serum protien electrophoreses showed low total protien, albumin (2 gm/dl) ,alpha 2 globulin (0.4 gm/dl), marked increase in the gamma globulins (3.8 gm/dl).

The following investigations were normal liver virology screen, serum iron, TIBC, S.Ferritin, Wilson screen ,blood urea and s.creatinine .

Urine examenation showed + red blood cell with few RBC cast, albumin ++ , sugar ++++ pus cells +.

Immune screen showed a postive ANA by Immunofluorescence, homogenous pattern, titer 1/320 anti ds DNA + ve , concentrntion 180 u / L (N < 20 u/L) for which the patient diagnosed as acase of SLE..

AMA, ASMA , AntiRo/SSA , AntiLa/SSB, anti U1 RNP and anti centromer all were nagative.

In spite of treatement with prednisolone 40mg and azathioprine 100 mg daily, the patient developed rapid deterioration of liver function and died two months after diagnosis because of liver failure.

Discussion

Systemic lupus erythematosus (SLE) is an immunologically mediated multisystem disease characterized by exacerbation and remission. The presentation of SLE is varied with skin, musculoskeletal system, cardiovascular, renal, pulmonary, central nervous system, hematologic, and gastrointestinal tract involvement (4). Strict criteria that reflect multi-organ involvement have been established by the American College of Rheumatology in order to uniformly diagnose SLE and distinguish it from other connective tissue diseases; patients are required to have 4 of 11 criteria before a diagnosis of SLE may be established. These criteria need not to be present at one time, but may

criteria before a diagnosis of SLE may be established. These criteria need not to be present at one time, but may develop sequentially over many years. Abnormalities of liver function are not included in these diagnostic criteria and the liver is generally not a major target for end organ damage in patient with SLE. (5). The diagnosis of liver disease preceded the diagnosis of SLE by up to 5 years in 27% in one series. In 45% the diagnosis of liver disease occurred within 1 year of diagnosis of SLE and in 28% it was made 1 or more years after the diagnosis of SLE. Patients who had SLE and liver disease differed from those without liver disease by having a higher number of mucosal ulcers (73 versus 18%), more thyroid gland involvement (24 versus 4%) and less arthritis (30 versus 90%). Runyon and associates reviewed 238 patient, who met the criteria for SLE, 21% had abnormal liver enzyme at some point during their illness, the first liver enzyme abnormalities were noted during an exacerbation of SLE. Elevation of serum aminotransferase and alkaline phosphatase activaties were usually mild, less than four fold the upper limit of normal. However more sever liver disease occurred. Abnormalities included cirrhosis, chronic active hepatitis, granulomatous hepatitis, chronic persistent hepatitis and steatosis. Three patient died of liver failure. In contrast, Gibson and Myers found no significant liver disease in their series of 81 patient.(6). Matsumoto and his colleagues reviewed the Japanese lupus registry data found that the incidence of chronic hepatitis was 2.4%, cirrhosis 1.1% and liver fibrosis 0.8%.None of these patients died as a result of complication of liver disease. (Table)(7).

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Finding	number of Patients (N = 52)
Congestion	40
Fatty liver	38
Arteritis	11
Cholestasis	9
Peliosis hepatis	6
Chronic persistent hepatitis	6
Nonspecific hepatitis	5
Cholangitis	4
Nodular regenerative hyperplasia	3
Hemangioma	3

Matsumoto T, Yoshimine T, Shimouchi K, etal. The liver in systemic lupus erythmatosus: Pathologic analysis of 52 cases and review of Japnese autopsy registry. Human pathol 1992; 23: 1151 - 1158.

Secondary histologic findings that have been described have included microabscesses from bacterial infection, hemosiderosis from blood transfusion, primary biliary cirrhosis and drug toxicity. The liver may be involved in neonatal SLE. Interpretation of any studies of the incidence of liver disease in patients with SLE is complicated by comorbid condition and the potential for drug toxicities that may mimic chronic liver disease, most patients in those studies have been on varying doses of prednisone, which made them more likely to develop fatty infiltration of the liver. Hepatic injury from salicylates may also be a factor in producing some of the liver dysfunction associated with SLE. There is a correlation between blood salicylate level and the serum ALT activity. Discontinuation of aspirin results in prompt improvement of liver enzymes with no chronic sequelae. The antiphospholipid syndrome (APS) is increasingly recognized as a cause of hypercoagulable state that is associated with SLE. Vascular disease associated with the presence of lupus anticoagulant includes Budd-Chiari syndrome, hepatic venoocclusive disease, and hepatic arteritis.(8). There is no direct link between autoimmune chronic active hepatitis and SLE. Although both diseases are characterized by the presence of circulating antinuclear antibodies, the specification of these antibodies differs. The immunogenetic and pathogenetic mechanism of CAH & SLE are also dissimilar. Patients in whom both entities are present probably represent coexistent disease rather than an overlap syndrome.(9). There are case reports of primary sclerosing cholangitis and malacoplakia occurring in patients with SLE.(10,11), while criteria for treatment and treatment options are currently not well defined for those patients found to have SLE related liver disease, the majority of patients treated with steroids in one series had an improvement in liver biochemistry.

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