CASE REPORT

A case of lupus hepatitis

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SUMMARY

Systemic lupus erythematosus (SLE) and systemic sclerosis (SSc) are autoimmune diseases with a multisystem involvement. Hepatic manifestations do occur in these patients. There are various aetiologies for an abnormal liver function tests in this group of patients. Among the differentials include primary SLE-related liver disease called lupus hepatitis. We report a case of a young female who presented with cutaneous and hepatic manifestations of an SLE and scleroderma overlap disease. After ruling out other aetiologies, the patient was then diagnosed as lupus hepatitis. The patient responded well to steroids and is currently on regular follow-ups.

INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disease which can affect the skin, joints and kidneys.[1] Liver involvement has been shown to be common in SLE despite not being included in classification criteria.[2] There are various aetiologies for an abnormal liver function in this group of patients. Common causes are drug reaction, viral hepatitis and fatty liver disease.[3] After excluding other causes of abnormal liver function test, the patient was then diagnosed as lupus hepatitis. Lupus hepatitis a non-specific reactive liver disease characterised by asymptomatic elevated alanine transaminases level.[1] The prevalence of it in SLE patients has been reported to be around 3–23%.[3] We report a case of lupus hepatitis in a young female who presented with cutaneous and hepatic manifestation of an SLE and scleroderma overlap disease.

CASE PRESENTATION

A 26-year-old female was admitted with generalised maculo-papular rash involving the face, trunk and limbs (Figure 1). The rashes started 3 days prior to admission and were described as itchy and painless. It was associated with fever and polyarthralgia for 2 weeks. There was no oral ulcer or vasculitic skin lesion seen. She denies a history of allergies, illicit drug or traditional medication consumption. The patient also had incidental acute hepatitis whereby her alanine aminotransferase (ALT) test was almost > 10x ULN. An ultrasound of the abdomen revealed she had hepatosplenomegaly with no biliary duct obstruction. Her antinuclear antibody was positive with titre 1:320, speckled pattern. Her anti-dsDNA level and extranuclear antibody (ENA) were negative. She had a low C4 complement at the level of 0.11 g/L with normal complement C3 level, high anti-Cardiolipin Ig G level at 36.8U/ml and other workout for antiphospholipid syndrome were all negative. Hepatitis B, hepatitis C screening and AIH/PBC (autoimmune hepatitis, primary biliary cholangitis) workout were negative. Therapeutic drug monitoring for paracetamol was sent randomly and reported a level of 0.6mcg/ml, which was not significant. She was then counselled for a liver and skin biopsy. She strongly refused a liver biopsy. A skin biopsy was then done over the left back, revealing lymphohistiocytic cells infiltrates at the perivascular and periadnexal area, correlating to the diagnosis of acute cutaneous lupus. At the same time, dense thickened collagen bundle was seen in the dermal area, which is suggestive of scleroderma (Figure 2). She fulfilled 2019 EULAR/ACR classification criteria for SLE. She was diagnosed with lupus hepatitis and acute cutaneous lupus with underlying SLE scleroderma overlap syndrome after correlating with her skin biopsy result. A tapering dose of oral prednisolone together with topical hydrocortisone 1% cream was initiated, and she responded well to it. Her skin rashes had fairly subsided, and her liver function test has almost normalised. Her serial liver function is shown below (Table I).

DISCUSSION

SLE and systemic sclerosis (SSc) can have a multisystem involvement involving the skin, kidneys and the central nervous system. Liver involvement is not part of its criteria but can affect at least 60% of its patients. The most common causes of involvement in these patients would include drug-induced hepatitis, viral hepatitis such as hepatitis B or hepatitis C and autoimmune hepatitis which is characterised by elevated serum immunoglobulin levels and presence of autoantibodies. In this patient, however, the results were all negative. Hence, the patient was diagnosed as lupus hepatitis in the setting of an SLE and SSc overlap disease. Bessone et al.[4] described lupus hepatitis as entity with asymptomatic elevated transaminitis in the setting of an active SLE flare.[4] Recent studies into hepatic manifestations in SLE patients have found that the presence of ribosomal P autoantibodies to be involved. 70% of SLE patients with lupus hepatitis are known to have this autoantibody.[5] It is an antilymphocyte antibody that specifically reacts with activated T cells.[5] The most common hepatic manifestation with SSc would include primary biliary cholangitis (PBC). PBC is characterised by the presence of a positive antimitochondrial antibody or presence of a florid duct lesion based on liver biopsy. However, this patient refused for liver biopsy during the current admission. As the patient was initially thought to

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Table I: Serial liver function test of the patient showed resolving transaminitis after started on steroid

	Day 1	Day 4 Started steroid	Day 5 1 week on steroid	Day 10 2 weeks on steroid
TB	13.5	41.3	14.1	12.9
ALT	981	1026	362	82
AST	529	387	61	23
LDH	1489	1281	266	/
HB	14.4	14.3	13.1	13.8
TWC	13.63	11.25	13.7	16.80
EOS Abs	0.13	0.13	0.04	0.01
EOS %	1.0	1.2	0.3	0.1
PLT	259	170	346	416

TB: total bilirubin umol/L, ALT: alanine aminotransferase U/L, AST: aspartate aminotransferase U/L, LDH: lactate dehydrogenase U/L, HB: haemoglobin 10^9/L, TWC: total white count 10^9/L, EOS Abs: eosinophil absolute count 10^9/L, EOS: eosinophil percentage %, PLT: platelet 10^9/L.

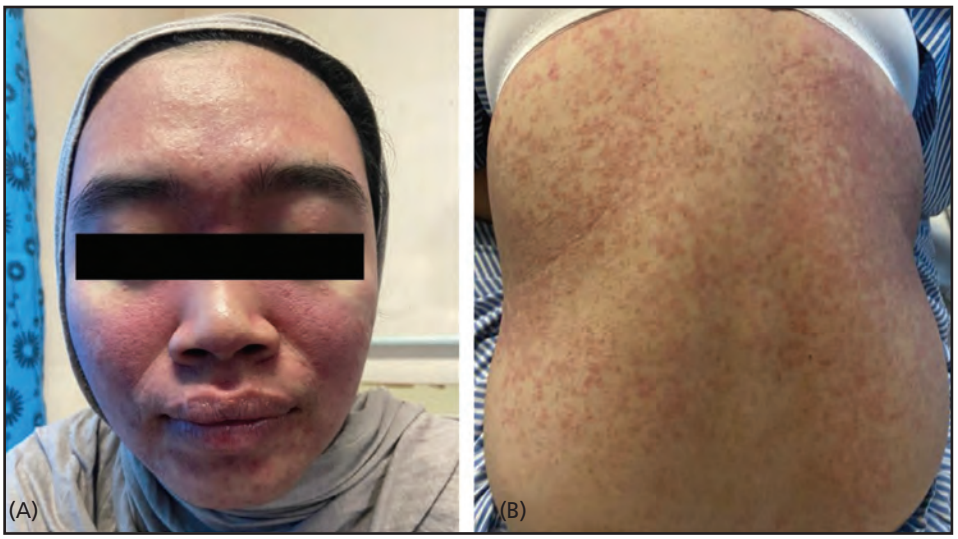

Fig. 1: (A) Facial erythema (B) Back of trunk with maculo-papular rashes.

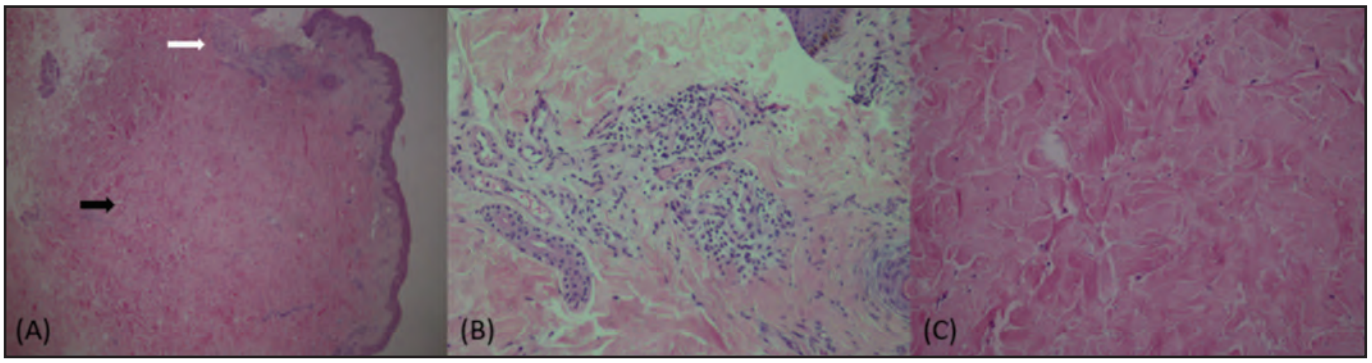

Fig. 2: Skin biopsy (A) White arrow: Mild lymphohistiocytic cells infiltrate at perivascular and periadnexal area. Black arrow: Dense thickened collagen bundle in the dermal area with a displacement of adnexal structures, 10×. (B) lymphohistiocytic cells infiltrate at perivascular and periadnexal area, 40×. (C) Dense thickened collagen bundle, 40×.

have a lupus flare, she was initiated with a tapering course of corticosteroids. She began improving for both her skin lesion and liver function. She is currently on the low prednisolone dose, 5 mg daily and her quality of life has improved tremendously. She does not exhibit other manifestations of SSc at the time being such as Raynaud's phenomenon or 'CREST' syndrome (Syndrome of calcinosis, Raynaud phenomenon, oesophageal dysmotility, sclerodactyly and telangiectasia).

CONCLUSION

Lupus hepatitis, though uncommon, has to be considered in any patients with SLE and SSC overlap syndrome presenting with a deranged liver function. Liver biopsy would be beneficial to clinch the diagnosis. However, in this case, the patient refused. Prompt investigations to exclude other causes and early treatment initiation would be beneficial.

CONFLICT OF INTERESTS

The authors declare that there is no conflict of interests regarding the publication of this paper.

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PATIENT CONSENT

Patient reviewed the photos and agreed for the publications of her history and photos.

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