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HEPATIC GRANULOMATOSIS AND LYMPHADENOPATHY AS AN UNUSUAL MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS - A RARE CASE REPORT

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ABSTRACT

Systemic Lupus Erythematosus (SLE) is a multisystem autoimmune disorder with diverse clinical manifestations. Hepatic granulomatosis with lymphadenopathy is an unusual presentation in SLE. We report a 15-year-old female with prolonged fever, rash, lymphadenopathy, and hepatosplenomegaly. Laboratory evaluation revealed pancytopenia, elevated liver enzymes, positive ANA, anti-dsDNA, and hypocomplementemia. Imaging showed hepatic granulomas, and renal biopsy confirmed lupus nephritis (Class II). Infectious and malignant causes were excluded. The patient was diagnosed with SLE and responded well to corticosteroids and hydroxychloroquine. This case emphasizes that hepatic granulomatosis, though rare, may represent an extrapulmonary manifestation of SLE and should be considered in differential diagnoses.

KEYWORDS

Systemic Lupus Erythematosus (SLE); Hepatic Granulomatosis; Lymphadenopathy; Autoimmune Hepatitis; Lupus Nephritis; Adolescent; Case Report

INTRODUCTION

Systemic Lupus Erythematosus (SLE) is a multisystem autoimmune disorder with a broad spectrum of clinical presentations encompassing almost all organs and tissues. Incidence is common in age group of 15-35 years, females more common than males. It is uncommon in children and young adolescents, with 0.5 0.6 per 1,00,000 per year. All patients classified as having SLE must have a serum titre of ANA of atleast 1:80 dilution. Only a few case reports have described hepatic granulomas in association with lymphadenopathy in SLE patients. Hepatic granulomatosis in SLE is exceedingly rare and may mimic infectious, malignant, or sarcoid etiologies. The presence of hepatic granulomas with generalized lymphadenopathy in a young female with lupus features poses a diagnostic challenge. This case highlights the importance of considering SLE in patients with unexplained granulomatous hepatitis once other common causes such as tuberculosis, sarcoidosis, and lymphoma are excluded. Here we report a case of adolescent female presented with fever, rash, lymphadenopathy with hepatic granulomas with a final diagnosis of SLE.

CASE REPORT

A 15-year-old female patient presented with moderate grade fever for 4 months, progressive abdominal distension for 3 months, generalized rash for 2 weeks. There was no history of cough, weight loss, joint pain, pain abdomen, jaundice or any other significant illness in the past. There was no past history of tuberculosis in the patient or in any other family member. General examination revealed moderate pallor and bilateral cervical axillary lymph nodes, with the largest being 1x1 cm. Rash was generalized and maculopapular with hyperpigmentation involving face, torso and extremities. Systemic examination revealed palpable spleen 4cm below the coastal margin.

Investigations revealed:

Hemogram was suggestive of pancytopenia.
Haemoglobin – 9.2 g/dL
WBC count – 2,800/µL
Platelet count – 90,000/µL
ESR – 58 mm/hr
AST – 128 IU/L, ALT – 102 IU/L, ALP – 340 IU/L
ANA – strongly positive (homogeneous pattern)
Anti-dsDNA – positive (high titre)
C3 and C4 – markedly reduced
Viral markers (HBsAg, anti-HCV, HIV) – negative
Serum ACE – normal
Mantoux test – negative
Direct coombs test: negative
Mantoux test: negative
CXR: B/L mild pleural effusion
CUE: 2+ proteinuria; UPCR: 0.62
24hr Urinary Protein: 496mg/day

Cervical LN biopsy: Shows Reactive lymphoid hyperplasia.

BM aspiration: Proliferative marrow with hypercellularity.

Ultrasound and CECT abdomen revealed hepatosplenomegaly with multiple small hypoechoic nodules in the liver and prominent abdominal lymph nodes. A fine-needle aspiration cytology from the liver showed epithelioid cell granulomas without caseation, suggestive of granulomatous hepatitis. Ziehl–Neelsen stain and PCR for mycobacteria were negative. Renal biopsy revealed mesangial proliferation consistent with lupus nephritis (ISN/RPS Class II).

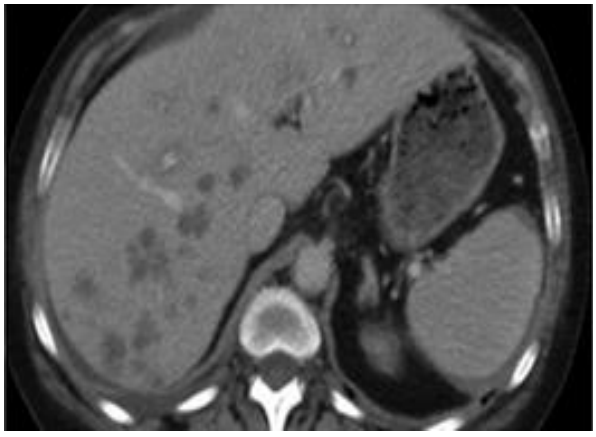
The patient fulfilled the 2019 EULAR/ACR classification criteria for SLE. A diagnosis of Systemic Lupus Erythematosus with hepatic granulomatosis and lymphadenopathy was established.

Treatment and outcome:

She was initiated on intravenous methylprednisolone pulses (1 g/day for 3 days) followed by oral prednisolone (1 mg/kg/day) and hydroxychloroquine 200 mg daily. Supportive therapy included calcium, vitamin D supplementation, and PPI coverage. Over 4 weeks, she showed marked clinical improvement with normalization of liver enzymes and regression of lymphadenopathy on follow-up imaging. She is currently under regular follow-up with gradual tapering of steroids.

DISCUSSION:

Hepatic involvement in systemic lupus erythematosus (SLE) is relatively frequent biochemically but rarely manifests as clinically significant liver disease. Elevated aminotransferases are common and may result from systemic inflammation, drug toxicity, or overlap with autoimmune hepatitis. However, hepatic granulomatosis remains an exceptionally uncommon presentation, reported in less than 2% of patients with SLE in major series [1].



Granulomatous hepatitis in lupus represents a diagnostic dilemma. The hepatic granuloma-composed of epithelioid histiocytes, lymphocytes, and occasional multinucleated giant cells-reflects a non-specific immune reaction pattern rather than a disease-specific lesion. It may arise secondary to infections (notably tuberculosis and histoplasmosis), sarcoidosis, drug hypersensitivity, or neoplastic infiltration. In endemic countries like India, tuberculosis is the most frequent consideration; however, sterile granulomas with strong lupus serologies, as in this case, point to an autoimmune etiology.

The pathogenesis of granulomatous hepatitis in SLE is incompletely understood. Immunopathological studies suggest deposition of immune complexes within hepatic sinusoids and periportal areas leading to complement activation and recruitment of macrophages. These activated macrophages release cytokines such as TNF-α and IL-6, inducing epithelioid transformation and granuloma formation [2]. Lymphadenopathy accompanying hepatic granulomatosis reflects systemic immune activation and parallels disease activity.

Lupus-related granulomatous hepatitis must be distinguished from "lupus hepatitis" and "autoimmune hepatitis overlap" [3,7]. Lupus hepatitis typically presents with mild enzyme elevation and lobular necro inflammatory activity without granulomas, while autoimmune hepatitis overlap shows plasma-cell-rich interface hepatitis and elevated IgG [3]. Histologic demonstration of non-caseating granulomas in the absence of infectious organisms or drug exposure strongly favours lupus-related granulomatous hepatitis.

Treatment is largely empirical and guided by disease activity. Corticosteroids remain the cornerstone, producing rapid biochemical and symptomatic improvement in most cases[4]. Adjunctive immunosuppressants such as azathioprine or mycophenolate may be required in refractory disease or when lupus nephritis coexists[5]. Hydroxychloroquine provides additional benefit in controlling systemic manifestations and reducing flare frequency. Our patient responded dramatically to corticosteroid therapy alone, consistent with prior reports describing excellent prognosis with early immunosuppression [4,6].

A review by Kobayashi et al. (2021) and Pires et al. (2018) noted that hepatic granulomatosis may precede or accompany other organ involvement, underscoring its value as a potential marker of systemic activity. Hence, recognition of this entity is clinically important not only for targeted treatment but also to prevent unnecessary anti-tubercular or oncologic therapy.

CONCLUSION:

Granulomatous hepatitis is a rare but important manifestation of systemic lupus erythematosus that can easily masquerade as infectious or neoplastic liver disease. In young women presenting with hepatosplenomegaly and lymphadenopathy, particularly in tuberculosis-endemic areas, awareness of lupus-related hepatic granulomatosis is crucial for accurate diagnosis. Histopathological confirmation combined with positive immunological markers (ANA, anti-dsDNA, hypocomplementemia) establishes the diagnosis once infections and malignancies are excluded.

The prognosis is favourable when recognized early and treated with corticosteroids or other immunosuppressants. Multidisciplinary collaboration between hepatologists, rheumatologists, and pathologists facilitates timely identification and prevents overtreatment. This case reinforces the principle that hepatic granulomas do not always signify infection—sometimes, they are the liver's cryptic whisper of lupus activity.

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