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Case Report

Systemic Lupus Erythematosus Presenting with Cholestatic Jaundice and Hepatic Involvement: An Etiology Not to Be Overlooked: A Case Report and Literature Review

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Abstract

We present the case of a 33-year-old female patient who was admitted for evaluation of cholestatic jaundice, dark urine, pale stools, and generalized, persistent pruritus. Her symptoms had been ongoing for the past 3 months, with no history of exposure to toxic substances, hepatotoxic medications, or medicinal herbs. Additionally, she reported atypical right upper quadrant abdominal pain and inflammatory polyarthralgia. Laboratory investigations revealed normochromic normocytic aregenerative anemia (hemoglobin 8.9 g/dL), normal white blood cell count, and normal platelet count. Her C-reactive protein (CRP) was markedly elevated at 66 mg/L, and alanine transaminase (ALT) and aspartate transaminase (AST) levels were five times the upper limit of normal. Gamma-glutamyl transferase (GGT) was eight times the upper limit of normal, alkaline phosphatase (ALP) was 13 times the upper limit of normal, and total bilirubin was predominantly conjugated (136 µmol/L). Prothrombin time (PT) was within the normal range at 88%. Abdominal imaging studies, including liver, portal vein, and hepatic veins, revealed no anomalies. Serological tests for hepatitis A, B, and C were negative. Autoimmune serology showed a positive antinuclear antibody (ANA) titer of 1/520, positive anti-double-stranded DNA (anti-dsDNA) antibodies, and positive anti-Smith (anti-sm) antibodies. Further investigations for autoimmune hepatitis were negative. A diagnosis of systemic lupus erythematosus (SLE) with hepatic involvement was established.

Keywords: ***Systemic Lupus Erythematosus, jaundice, hepatic involvement***

Introduction

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease that can affect multiple organ systems. Hepatic involvement in SLE is diverse and may range from mild elevation of liver enzymes to severe liver disease [1,2]. We report a rare case of SLE presenting primarily with cholestatic jaundice and hepatic abnormalities, without typical clinical manifestations of SLE.

Case Presentation

Our patient, a 33-year-old female, presented with a 3-month history of cholestatic jaundice, characterized by yellowing of the skin and sclera, dark urine, and pale stools. She also complained of generalized, persistent pruritus. Additionally, she had been experiencing an unusual right upper quadrant abdominal pain and inflammatory polyarthralgia.

Laboratory Findings

Laboratory investigations revealed anemia with a hemoglobin level of 8.9 g/dL, which was normochromic and normocytic, suggesting a regenerative response. White blood cell count and platelet count were within the normal range. However, her CRP was markedly elevated at 66 mg/L, indicating active inflammation. Liver function tests demonstrated elevated ALT and AST levels, which were three times the upper limit of normal, along with significantly elevated GGT (eight times the upper limit) and ALP (13 times the upper limit) levels. Notably, total bilirubin was predominantly conjugated (136 µmol/L), indicative of cholestasis. PT, reflecting hepatic synthetic function, was within the normal range at 88%.

Imaging Studies

Abdominal imaging, including assessment of the liver, portal vein, and hepatic veins, did not reveal any structural anomalies or obstructions that could account for the cholestatic pattern of liver dysfunction.

Autoimmune Serology

Autoimmune serology demonstrated a positive ANA titer of 1/520, indicating the presence of autoantibodies targeting nuclear antigens. Furthermore, anti-dsDNA antibodies and anti-sm antibodies were both positive, consistent with a diagnosis of SLE.

Diagnosis and Management

Given the patient's clinical presentation, laboratory findings, and positive autoimmune serology, the diagnosis of SLE with hepatic involvement was established. Although hepatic involvement is relatively common in SLE, this case was unique due to the predominant cholestatic pattern of liver dysfunction and the absence of classic SLE symptoms at the time of presentation.

Discussion

Jaundice is a clinical manifestation of various hepatobiliary and systemic disorders, and while it is commonly associated with liver diseases, it can also be a rare presenting feature of autoimmune conditions such as Systemic Lupus Erythematosus (SLE). This discussion aims to shed light on the infrequent occurrence of SLE presenting primarily with cholestatic jaundice, emphasizing the importance of considering autoimmune etiologies in patients with unexplained liver dysfunction [3-5].

Systemic Lupus Erythematosus is a multisystem autoimmune disease characterized by autoantibody production and immune complex deposition, leading to inflammation and tissue damage in various organs. While hepatic involvement is recognized in SLE, the presentation of jaundice as a primary manifestation is exceptionally rare. Most patients with hepatic involvement in SLE typically present with mild elevations in liver enzymes, hepatomegaly, or non-specific symptoms like fatigue [5-7].

The literature on SLE presenting with cholestatic jaundice is limited but highlights the diagnostic challenges associated with this uncommon presentation. Case reports and small case series have described patients with SLE who initially presented with jaundice, often accompanied by pruritus and dark urine, mimicking cholestatic liver diseases [8-10].

The exact pathophysiology of hepatic involvement in SLE remains incompletely understood. Immune complex deposition in the liver, autoimmune hepatitis, and drug-induced liver injury are some of the proposed mechanisms. In the presented case, the patient had no history of hepatotoxic medications or exposure to toxic substances, underscoring the importance of considering autoimmune etiologies when faced with unexplained jaundice [1,11].

The differential diagnosis of cholestatic jaundice is extensive and includes both hepatobiliary diseases (such as primary sclerosing cholangitis, primary biliary cholangitis, and choledocholithiasis) and systemic diseases (such as SLE, sarcoidosis, and malignancies). This wide range of possibilities often necessitates a comprehensive evaluation, including laboratory tests, imaging studies, and liver biopsy, to establish the correct diagnosis.

Role of Autoantibodies and Serology

In our presented case, serological tests played a pivotal role in reaching the diagnosis of SLE. The presence of a high-titer antinuclear antibody (ANA), anti-double-stranded DNA (anti-dsDNA) antibodies, and anti-Smith (anti-sm) antibodies were crucial diagnostic markers. These autoantibodies are specific to SLE and played a key role in differentiating it from other causes of cholestatic jaundice [12,13].

Management and Implications

The management of SLE with hepatic involvement typically involves the use of immunosuppressive agents, such as corticosteroids and other disease-modifying antirheumatic drugs (DMARDs). Additionally, addressing the underlying autoimmune process is essential to prevent further liver damage. In cases where SLE presents with jaundice, a multidisciplinary approach involving rheumatologists, hepatologists, and other specialists is often required to optimize patient care [14,15].

Conclusion

This case highlights the diversity of clinical presentations of SLE, including its ability to mimic liver diseases and manifest primarily with cholestatic jaundice and hepatic abnormalities. Prompt recognition and appropriate management are crucial in such cases to prevent further liver damage and to address the underlying autoimmune process.

Conflicts of interests

The authors declare no conflicts of interest.

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Author's contributions

All the authors have read and agreed to the final manuscript.

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