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Case Report

A rare presentation of systemic lupus erythematosus with lupus hepatitis

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ABSTRACT

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by multisystem inflammation and the presence of circulating autoantibodies directed against self-antigens. Prevalence of SLE in children and adolescents is 1-6 per 100,000 population. Liver dysfunction occurs in approximately in 50-60% of the patients with SLE. And the incidence of lupus hepatitis in diagnosed SLE patients is 9.3%. We are reporting a case of 11-year-old female child who presented with fever, skin rashes, oral ulcers, irritability and positive antinuclear antibody (ANA) with deranged liver function test (LFT) in the form of elevated transaminases, bilirubin level and altered coagulation profile. Hepatitis serology was negative, with low C3 levels, diagnosed as SLE with lupus hepatitis with lupus nephritis stage IV with psychosis. Clinical improvement along with improvement in terms of laboratory findings was seen on corticosteroids therapy. It is important to differentiate lupus hepatitis from autoimmune hepatitis (AIH) as AIH presents similar to lupus hepatitis, has poor prognosis. While lupus hepatitis being rare cause for mortality in patients with SLE has good response with early intervention with corticosteroids therapy.

Keywords: SLE, Lupus hepatitis, AIH

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by multisystem inflammation and the presence of circulating autoantibodies directed against self-antigens with involvement of the kidney, the joints, the skin and central nervous system rarely liver is also affected in this disease.[1] Hepatic involvement in patients with SLE is well documented, but considered to be rare with incidence being 9.3%.[4] Hepatic dysfunction secondary to parenchymal injury when it is associated with SLE is referred to as lupus hepatitis, hepatic dysfunction can also occur secondary to hepatotoxic drugs, viral infection or following steroid administration (non-alcoholic fatty liver disease) or it can be due to presence of other autoimmune disorder as in overlap syndrome.[6] In patient with SLE, who presented with abnormal LFTs one should consider lupus hepatitis and primary liver diseases such as AIH as differential diagnosis and detailed clinical evaluation, appropriate serological tests should be done to diagnose lupus hepatitis. Early diagnosis and early intervention with corticosteroids are important to prevent mortality. Histopathological features which help in diagnosis of AIH does not excludes SLE.[7] Clinical presentation of AIH is similar to lupus hepatitis but it has poor prognosis while, lupus hepatitis being rare cause for mortality in patients with SLE who has good response with corticosteroid therapy.[2,5]

CASE REPORT

A 11-year-old female child 1st born to non-consanguineously married couple presented with complaints of fever with rash for 3 months and irritability for 1 week. Fever was intermittent in nature not relieved

on medication associated with skin rashes initially started on the face and gradually progressed to involve axillary region and back. No significant past history, family history, developmentally appropriate for age, immunized according to national immunization schedule. On examination she had icterus, submandibular and posterior cervical lymphadenopathy, patchy alopecia, malar rash, palatal ulcer, vasculitic rash in the toes (Figure 1 and 2), oral ulcers (Figure 3) and distended abdomen with mild hepatomegaly (4 cm below right costal margin). Complete blood count with peripheral smear showed macrocytic, microcytic hypochromic anemia with few schistocytes. Coombs's test was weekly positive. LFT showed elevated bilirubin level (total/direct bilirubin=4.7/4.18 mg/dl), and transaminases (AST/ ALT=1966/154 IU/dl) with deranged coagulation profile. Urine routine showed proteinuria with urine spot protein creatinine ratio of 1.17 mg/g of creatinine with normal renal function test. Provisionally diagnosed to have SLE with psychosis based on EULAR criteria, with involvement of kidney, liver, skin, CNS.

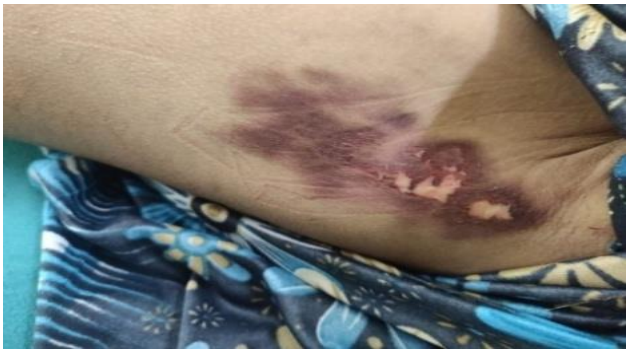

Figure 1: Vasculitic rashes in flank.

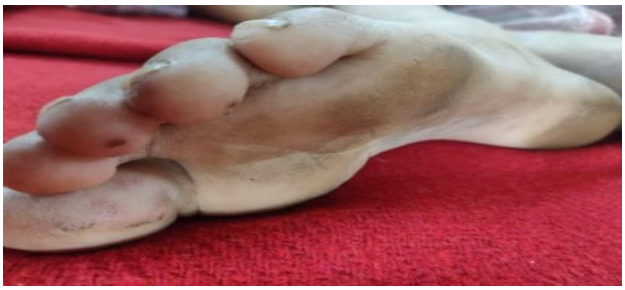

Figure 2: Vasculitic rashes in toes.

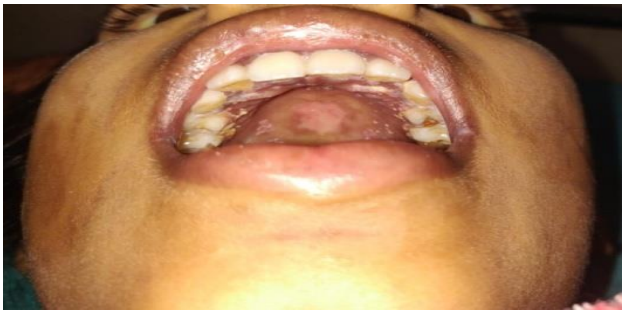

Figure 3: Ulcers in oral mucosa.

Child developed abdominal distension with hypokalemia, USG showed mild ascites with altered liver echo pattern. Child was shifted to PICU and hypokalemia correction has been done under ECG monitoring. ANA profile was strongly positive for ribosomal P protein, SS-A, SS-B, Smith, RNP antibodies and Ro-52 was borderline positive with low complement (C3) levels and hepatitis serology was negative.

Child was started on IV methylprednisolone pulse therapy 30 mg/kg/day for 3 days along with respiridone (for psychosis). For deranged coagulation profile vitamin K was given along with fresh frozen plasma was transfused. Followed by methylprednisolone child was started on tablet prednisolone 1 mg/kg/day with Hydroxychloroquine 50 mg. Child had persistent proteinuria and increased spot protein creatinine ratio with active disease in the form of circulating autoantibodies, hence child was diagnosed as lupus nephritis stage IV and started on cyclophosphamide 500mg/m^2 and tab enalapril 2.5 mg after taking nephrologist opinion. Since child had deranged INR with deranged LFT. Kidney biopsy and liver biopsy were planned on follow-up.

Gradual improvement noticed clinically and in liver functions in the form of normalization of liver enzymes, coagulation profile and renal function in the form of improvement in proteinuria. Child was given 14 days of antibiotics for enterococcus sepsis and discharged with tablet prednisolone and hydroxychloroquine and resperidone.

DISCUSSION

SLE is multisystem involved autoimmune disorder most commonly affecting adolescent female with reported 2-5:1 ratio before puberty, 9:1 ratio during reproductive years.[1] SLE commonly affects skin, joints, kidney, blood vessels and CNS but involvement of liver is rare with the prevalence of 11.8% in active SLE patients.[4] In SLE, there is no specific pattern of liver injury. However, when there is hepatic dysfunction in patients with SLE, auto antibodies have role in triggering hepatic dysfunction and causing subclinical hepatopathy which is referred to as "lupus hepatitis" and it is described as an asymptomatic elevation of transaminases which declines to normal values after corticosteroid therapy in patients with SLE. In lupus hepatitis development of hepatitis is due to expression of proinflammatory cytokines upregulated by anti-ribosomal P antibodies, hence anti-ribosomal P antibodies can be used as diagnostic marker. Other markers like anti dsDNA, low levels of complement 3, histopathological examination showing nonspecific inflammation, fatty degeneration are key to diagnose lupus hepatitis.[6]

AIH has similar clinical manifestation and liver dysfunction in terms of elevated transaminases. Hepatomegaly is important and distinct feature of AIH.

To diagnose AIH autoantibodies like ANA, anti-smooth muscle antibody (ASMA), anti-liver kidney microsome antibodies must be present in a titer of at least 1:20 in children and serum gamma globulins level should be greater than one and half times the upper limit, and liver biopsy findings showing piecemeal necrosis, bridging necrosis, lymphoplasmacytic infiltrate must be present.[8,9]

In patient with SLE and AIH there is increased levels of IgG and antinuclear antibodies, which is characteristic of both diseases. Presence of anti-double stranded DNA antibodies are frequently associated with SLE but it is present in patients with AIH also. Sensitivity for the diagnosis of SLE with anti-double stranded DNA range from 25-85%.[10] Autoantibodies to 3 ribosomal proteins P0, P1, P2 are specific in diagnosis of lupus hepatitis, which are found in 10- 40% of patients with SLE.[11] Overall sensitivity and specificity for anti-ribosomal P antibody in diagnosis of lupus hepatitis was 23.1% and 99% repectively.[12]

In this case report patient fulfills the criteria for SLE, since other causes for liver dysfunctions like viral hepatitis, drug induced hepatitis and auto immune hepatitis are ruled out, with positive ANA, decreased complement 3 levels, and positive anti-ribosomal P antibody (useful serologic marker to differentiate between lupus hepatitis and AIH) patient was diagnosed to have lupus hepatitis. An accurate diagnosis and early intervention are important as lupus hepatitis is a benign condition, and AIH is associated with poor prognosis and progression of which requires aggressive therapy.[2,3]

CONCLUSION

Presentation of patients with SLE and AIH in terms of clinical features, laboratory features are often similar it is important to differentiate between lupus hepatitis from AIH in suspected SLE patients with elevated liver enzymes as AIH has poor prognosis which requires aggressive treatment. In resource limited settings where liver biopsy is not feasible, we can use serological tests like positive ANA and anti-ribosomal P antibodies as a useful diagnostic marker in diagnosing lupus hepatitis for early intervention and to prevent mortality.

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