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Case Report

A rare case of systemic lupus erythromatosus with acalculous cholecystitis as its first presenting feature

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ABSTRACT

SLE is an autoimmune disease of multisystem involvement primarily involving skin, kidney and joints. GI involvement is not a common feature of SLE, though acalculous cholecystitis (AAC) has been reported in patient with SLE. Most of the previous cases of AAC (with SLE) reported had occurred in diagnosed cases of SLE. We are hereby reporting a case of AAC diagnosed simultaneously with SLE. The patient responded to treatment with immunosuppresion with cyclophospamide.

Keywords: Systemic lupus erythromatosus, Anti-nuclear antibody, Acalculous cholecystitis

INTRODUCTION

Acalculous cholecystitis (AAC) is an acute necroinflammatory disease of the gallbladder with a multifactorial pathogenesis. It accounts for approximately 10 percent of all cases of acute cholecystitis and is associated with high morbidity and mortality rates.[1]

Most cases of acute acalculous cholecystitis occur in the setting of prolonged fasting, immobility and hemodynamic instability,[2] however autoimmune diseases like systemic lupus erythromatosus (SLE) has also been proposed as aetiology of AAC. Most of the cases of AAC associated with SLE has occurred in previously diagnosed cases of SLE[3] but here we are reporting a case of AAC which occured as a first manifestation of SLE

CASE REPORT

A 31 years old female Hindu patient presented to emergency with chief complaints of nausea with occasional vomiting, anorexia, bloating sensation, generalized weakness, abdominal pain, intermittent fever, discoloration of skin-initially on face and then involved hands, feet, both upper limb, both lower limb with large area on the back, alopecia & pedal edema from 3 months .

There was no history of photosensitivity, joint pain, facial puffiness, breathlessness, diarrhea, seizures, loss of consciousness or neural symptom.

Obstetrics history - G1 P1 A0 L1. No significant past or family history.

On examination; temperature was normal, pulse 90/min, blood pressure - 124/70 mmHg, respiratory rate - 18/min, mild pallor. General examination reveals malar rash, discoid rash, cutaneous erythema, oral ulceration, reticular telengiectesia erythema on palms and cheek. Similar lesions seen on neck and large area of back. White nails and ragged cuticle were seen along with diffuse hair loss and coarse, dry and fragile hair

especially on frontal border (lupus hair). Systemic examination reveals normal respiratory system and central nervous system, cardiovascular system shows mid systolic click at apex, with mild hepatomegaly and splenomegaly, fundus examination shows cotton wool spots and retinal vasculopathy.

Investigation shows haemoglobin (Hb) - 10.1gm%, Total Count (TC) - 2350/cmm, Differential Count (DC) - 70/26/2/2, Platelet Count (PC) - 1.17 lakhs, Peripheral Smear (PS) - few schistocytes, anisopoikilocytosis, Erythrocyte Sedimentation Rate (ESR) – 212 mm/hour, Random Blood Sugar (RBS) – 106 mg%, alanine transaminase (ALT) - 45 IU/l, blood urea - 16mg%, aspartate transaminase(AST) -38 IU/l, serum creatinine - 0.8 mg%, alkaline phosphatase (ALP) - 1950, serum sodium (Na) - 143 meq/l, serum potassium (K) - 4.3 meq/I, PT INR - 0.8, activated partial thromboplastin time (aPTT) - 29 sec, serum bilirubin - 0.84 mg%, serum calcium- 9.8 mg%, urine - albumin 1+. Serum HIV - negative, S. HBsAg - negative, S. HCV - negative, total protein - 7.5 gm%, serum albumin - 2 gm%, Serum globulin - 5.5 gm%, X-ray chest - normal, X-ray both hands - normal, electrocardiogram (ECG) - NSR/WNL, ANA screening: ANA by IF - positive 1:40, +++ intensity, ANA profile**:** Anti Sm ++, Anti SS-A naïve/Anti RO 52 +, Anti ribosomal P protein ++, anti-histone+ APLA - negative, rheumatoid arthritis factor (RA) - negative, antistreptolysin titre (ASO) - negative, C-reactive protein (CRP) - not raised, T3-0.91, TSH - 6.42 (raised), T4-7.07, 24 hour urinary protein – 333 mg/24 hours, S. LDH - 300.65 U/L, direct Coomb's test - positive, reticulocyte count - 1%, bone marrow examination - normocellular marrow, mildly raised myeloid: Erythroid ratio, not involved by any malignancy. Punch Biopsy from skin: Focal atrophy with keratotic plugging in the epidermis basal cell liquefaction, degeneration with colloid bodies, patchy mononuclear infiltrates in the dermis, superadded vasculitis.

Diagnosis of SLE is made - Following 6 American College of Rheumatology (ACR) criteria fulfiled: Malar rash, discoid rash, oral ulceration, Hematological manifestation, presence of anti sm ab ANA+,

On day 2 - Her condition worsened rapidly, she developed acute abdominal pain in right hypochondrium, severe colicky, radiating to back. Guarding/rigidity present, positive Murphy's sign, X-ray abdominal standing was normal. Urgent Ultrasound Abdomen shows fatty liver, mild hepatosplenomegaly, lammelated gall bladder. Contrast computed tomography abdomen showed- increased gall bladder (GB) wall thickness with peri GB fluid collection and there was no evidence of GB calculi so diagnosis of acute acalculous cholecystitis was made. Patient was advised to manage conservatively by surgeon. Ryle's tube was inserted and was made continuous. She was kept nil by mouth. Antibiotics and IV fluids were continued. Injection methyl prednisolone 1 gm daily for 3 days started. Patient did not respond to steroids so injection cyclophosphamide 900 mg (750 mg/m^2 with mesna was given.

Patient responded dramatically to treatment within 3-4 days. Acalculous cholecystitis resolved, skin rash started to fade gradually leaving behind hyper pigmentation, renal vasculitis showed improvement, constitutional symptoms resolved. Fresh blood investigation shows Hb - 10.6, TC-6130, DC - 80/18/1/1, PS - anisopoikilocytosis, Urine albumin-trace , renal function test- normal, serum bilirubin - 1.2, ALT - 291, AST - 190, ALP - 3023.

DISCUSSION

Acute acalculous cholecystitis is an uncommon but very serious illness.[4] The disease may go unrecognised due to complex medical and surgical conditions associated with it. It is mainly caused by decreased blood supply to gallbladder, sepsis and ischemia.[5]

SLE is an autoimmune disease of unknown aetiology primary affecting joint, skin and kidneys.[6] GI involvement is not so common in SLE, however vasculitis of mesenteric vessel is associated with some of gi disorders caused by SLE and AAC is one of such disorder caused by vasculitis of mesenteric vessels.[7]

AAC has poorer prognosis as compared to calculous cholecystitis and it is generally managed by cholecystectomy.[8] However in our case report patient was managed conservatively by immunosuppression with cyclophospamide showing good outcome.

Summary

We herein reported a rare case of SLE patient with AAC as an initial presentation who was treated successfully with high dose cyclophospamide pulse therapy and immunosuppressive agent. Awareness of this finding would be valuable in the early diagnosis and adequate management of AAC in patients with SLE.

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