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RESEARCH ARTICLE

INTUSSUSCEPTION DUE TO MESENTERIC VASCULITIS A RARE MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

Brajagopal Jana, Indranil Sen, Triparna Bhadra and Spandan Bhadury

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Abstract

Mesenteric vasculitis is one of the rare but deadliest complication of systemic lupus erythematosus (SLE) with a vast diverse spectrum of vasculopathy developing hemorrhage, ulceration, perforation, bowel necrosis, thrombosis. Intussusception due to underlying mesenteric vasculitis in background of SLE has been reported previously as a rare association in various literatures [1, 2]. Here we report a case of SLE with low SLEDAI where initial presentation was mesenteric vasculitis but it progressed to intussusception which is a rare presentation.

Introduction:-

Gastrointestinal manifestations are very common in patients with SLE with variable incidence (15% to 75 %) from different studies and registries. [1]. Clinical picture varies from minor local symptoms like oral ulcer, dysphagia, nausea, vomiting to life threatening conditions notably pancreatitis, hepatitis, mesenteric vasculitis. Among gastrointestinal presentations mesenteric vasculitis is uncommon(2.2% to 9.7%)[1][5] but with grave outcome unless prompt treatment is administered .Gastrointestinal vasculitis results from bowel ischemia due to diffuse deposition of circulating immunocomplexes and clinically manifested with diffuse abdominal pain, vomiting, fever. [4]

Case report:

A 35 years old married lady, home maker, mother of two children without any history of pregnancy loss, normotensive, non diabetic, without any history of addiction developed gradually increased fatigability, anorexia and swelling of abdomen over last one month followed by low grade intermittent fever for next 15 days without any associated chills and rigor, headache, convulsion, cough, rash, dysuria, myalgia, arthralgia. Then she suddenly experienced severe colicky, diffuse, periumbilical pain which was gradually progressive reaching peak within 8 hours, spreading to whole abdomen without any radiation or aggravation in particular posture or in change of posture, movement, sneezing, coughing, except aggravation on intake of solid food. Pain abdomen was associated with nausea, vomiting and inadequate relief from oral medications including PPI, antispasmodic, compelling her for hospitalization. There was no history of diarrhoea, upper GI bleeding, and constipation, bleeding PR/PV, strangury, weight loss, and abdominal trauma. Her LMP was 6 days prior to development of pain abdomen. General examination revealed moderate pallor and on gastrointestinal examination, flanks were full, there was diffuse tenderness on palpation, shifting dullness on percussion, and sluggish IPS on auscultation, there was no organomegaly.

Laboratory investigations showed pancytopenia with microcytic hypochromic RBC (Hb- 6.5 g/dl, TLC- 2250/µL, Platelet 70,000/µL).Biochemical and microbiological investigations are shown on table 1 to 5. Digital chest x-ray (PA view) and 12 leads ECG were normal. Echocardiography showed mild pericardial effusion. USG of whole

Corresponding Author:- Brajagopal Jana

abdomen demonstrated moderate ascites with 21× 24 mm size hypoechoic lesion in VIth segment of liver and gut within gut shadow suggestive of Intussusception. Sputum for AFB and CBNAAT were negative and sputum culture yields no growth. CECT of whole abdomen showed moderate ascites, inflamed mesenteric fat, edematous gut, Target sign and Comb sign and 24×26 mm ring enhancing cystic SOL in VIth segment of liver [Fig 3]. ANA in hep 2 cell line was 4+, coarse speckled pattern and ANA profile positive (Anti-dsDNA 1+, RNP/Sm 3+, Sm 3+ [Fig 2]. Bone marrow study for pancytopenia revealed hypoplastic bone marrow with presence of haemophagocytes, plenty of macrophages, fibroblasts with relative increase in plasma cells[Fig 1].

In this case the patient didn't show any evidence of clinical SLE activity except polyserositis. Laboratory test showed raised pancreatic enzymes amylase, lipase with low C3, C4 complements level. Urine analysis showed no significant proteinuria. ANA profile was positive with SLEDAI score 4. Abdominal CT demonstrated ascites and diffuse jejunoileal thickening with Target sign and Comb sign, suggesting ongoing microvascular bowel ischemia and slow transit time in small bowel with distended handles suggesting intestinal obstruction due to intussuception. Patient was given IV antibiotics, analgesic, PPI and 2 unit PRBC transfusion. Methylprednisolone pulse 1gm for 3 days initiated due to severe progressive intense pain in background of subacute intestinal obstruction suggestive of mesenteric vasculitis and pain abdomen was little bit relieved. After establishing the diagnosis Cyclophosphamide therapy was started as per NIH protocol which yields significant symptomatic improvement. After completing 6 cycles of Cyclophosphamide over 6 months she is now on maintenance therapy with mycophenolate mofetil resulting in complete resolution of symptoms on follow up.

Discussion:-

Mesenteric vasculitis is an uncommon but extremely serious complication of SLE with very high case fatality rate (50%) [1]. Early diagnosis of mesenteric vasculitis and prompt initiation of therapy dramatically changes the prognosis of the disease with better outcome.

Characteristic histopathological findings in mesenteric vasculitis includes diffuse inflammation of vascular wall with fibrinoid necrosis due to intense deposition of complements, immune-complexes and fibrinogen, resulting in regional ischemia and dysfunction of intestinal musculature[1,3,5].

Mesenteric vasculitis commonly presents with abdominal pain due to colonic ischemia, distension and pseudo-obstruction [1, 2, 4, 8,9]. Laboratory investigation including biochemical, microbiological and serological parameters primarily shows underlying inflammatory activity. Radiological investigations including X-ray, USG and CECT of abdomen demonstrate bowel wall thickening, mesenteric fat necrosis, colonic ischemic changes and pseudo-blockage likewise in present case [1, 2, 4, and 6]. Abdominal CT accurately detects the mesenteric vasculitic changes:- Comb sign (engorgement of the mesenteric vessel with vascular dilatation, tortuosity with spacing of the vasa recta and prominence of surrounding mesenteric fat resembling a comb), and Target sign/ Doughnut sign/ Bull's eye sign (thickening of intestinal wall in which a lower attenuated middle layer is surrounded by layer of higher attenuation/ a round soft tissue mass containing radioluscent ring of fat within-represents intussusception)[1,3,5,7-9]. Presence of comb sign suggests an active vascular inflammation, confirming diagnosis of mesenteric vasculitis in an initial phase of the disease. Vasculitis can be demonstrated by mesenteric vasculature arteriography, by detecting irregular vascular branches mainly in the region of superior mesenteric artery. Angioresonance and gallium scintigraphy are other methods which can be used to detect mesenteric vasculitis. Abdominal pathologies like intestinal obstruction perforated peptic ulcer, peritonitis, pyelonephritis, tuberculous colitis, pancreatitis, acute appendicitis, acute cholecystitis to be excluded while evaluating for mesenteric vasculitis. Causes of intestinal vascular ischemia like atherosclerosis, embolism, APS, must need to be excluded. Various literatures affirmed that mesenteric vasculitis as a result of lupus activity in multiorgans generally found with SLEDAI more than 8[1]. Quick effective response is achieved in treatment of mesenteric vasculitis with Methylprednisolone Pulse therapy followed by immunosuppressive therapy particularly Cyclophosphamide, MMF, Rituximab though there is no randomized control trial proving superiority regarding selection of immunosuppressive agents[1,5,6].

Conclusion:-

Our case report describes an unusual presentation of Intussusception due to mesenteric vasculitis in a systemic lupus erythematous patient with low SLEDAI score. Prompt and aggressive immunosuppression is required to manage such life-threatening complication before going for surgical procedure.

Figures and Tables:

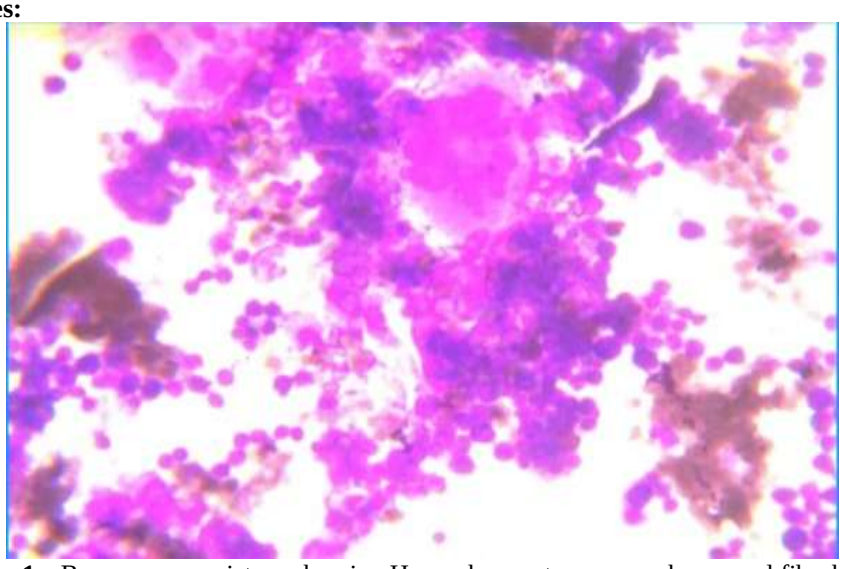

Figure 1:- Bone marrow picture showing Hemophagocytes, macrophages and fibroblasts.

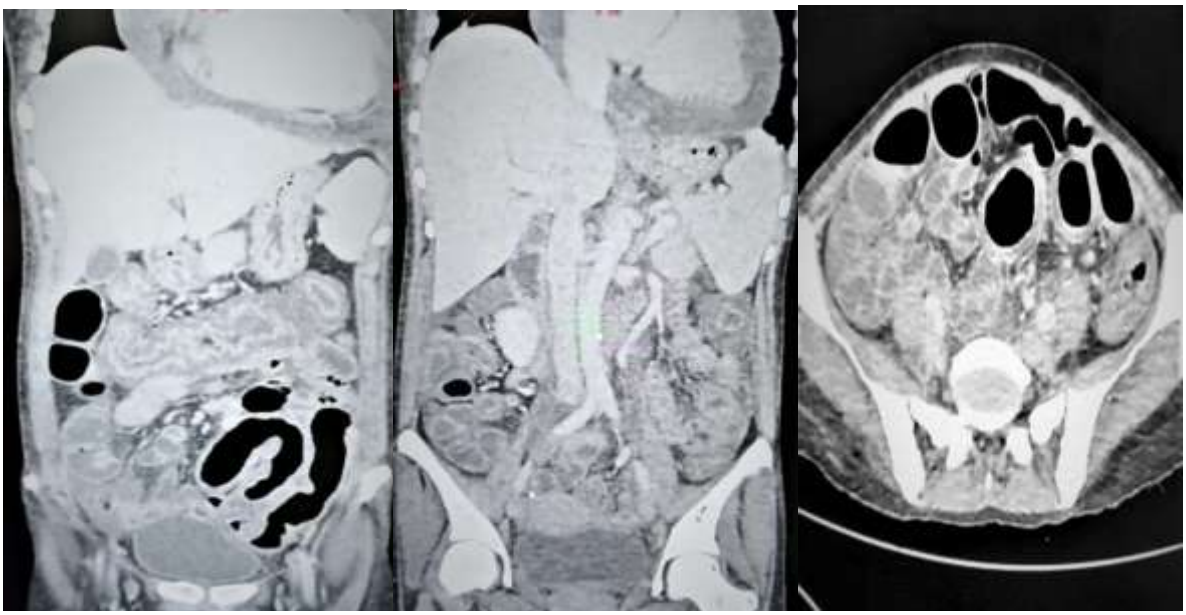

Figure 2:- CECT abdomen showing target sign and coomb sign.

Table 1:- Metabolic parameters.

CRP	7.3 mg/dL	Amylase/Lipase	126/127 U/L
ESR	26 mm 1st hour	Ferritin	1001 µ/L
Ur/Cr	29/0.7 mg/dL	Serum LDH	239 U/L
Na/K	136/4.1 mEq/ L	Corrected calcium	9.4 mg/dL
Total bilirubin	0.3 mg/dL	DCT	Negative
Direct bilirubin	0.1 mg/dL	Fastinglipid profile	Normal
SGOT/SGPT	30/26 U/L	Serum C3	62.81 mg/dL
Total protein/ Albumin	6.1/2.6 gm/dL	Serum C4	10.22 mg/dL

Table 2:- Ascitic fluid analysis.

Parameters	Results
Colour	Turbid
Cell count	100/cmm
Cell type	N60L40
Glucose	9.8
Protein/ Albumin	3.6/1.8
M cell	Nill
SAAG	Low

Table 4:- Urine analysis.

Parameters	Results
Protein	Trace
Sediment	Present
Cast	Granular
Pus cell	2-3/hpf
C/S	No growth
24 hours urinary protein	337.2 mg/d

Conflicts of interest:
None declared.

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