Abdominal cocoon as a presenting feature of systemic lupus erythematosus

A rare presentation

Rinkesh K. Bansal, MBBS, MD, Ashish Kumar, MD, DM, Naresh Bansal, MD, DNB, Anil Arora, MD, DM

ABSTRACT

Systemic lupus erythematosus (SLE) is a common autoimmune disorder characterized by multisystem involvements, including the involvement of any part of the gastrointestinal tract (GIT), although often subtle and difficult to interpret. Sclerosing encapsulating peritonitis (SEP) is a rare event, defined as a chronic inflammatory condition that transforms the peritoneal membrane into a dense fibrous sheet. With progression of the fibrosing and sclerosing process, the bowel is encased partially or completely into a thick grayish-white fibrotic membrane forming “cocoon,” at which point the term “sclerosing encapsulating peritonitis” is given. The SEP can be classified as idiopathic, or secondary. The idiopathic form classically described in young adolescent females from the tropical and subtropical countries, but also seen in adult from non-tropical areas. The SEP is rarely reported in SLE as case reports in literature. We hereby present a case of abdominal cocoon with repeated vomiting and subacute intestinal obstruction (SAIO) as a presenting feature of SLE in a young girl.

Case Report. A 14-year-old girl presented with a history of low-grade intermittent fever with generalized weakness and joint pain for the last 4 months. She was investigated by a local practitioner and was found to have hemoglobin (10.2 g/dL), total leucocyte count (TLC, 5410/µl), and erythrocyte sedimentation rate (ESR, 54) at the end of first hour. Based on urine culture and sensitivity reports, she was given 10 days injectable antibiotic but without any benefit. Meanwhile, she developed vomiting, painful abdomen,
and abdominal distention. One month ago, with these complaints she was admitted in a medical college hospital. She was found to have ascites, and her ascitic fluid analysis revealed protein of 4.3 g/dL, cell count of 270/µl with 70% lymphocytes. Serum-ascites albumin gradient (SAAG) could not be evaluated because of lack of ascitic albumin. She had features of intestinal obstruction, and hence laparotomy was carried out, which revealed gross peritoneum thickening encasing most of the small intestine in a cocoon. Omental biopsy was obtained, which showed vascular fibro-fatty tissue without any evidence of granuloma. She was started on anti-tubercular treatment (ATT) on a presumptive diagnosis of abdominal tuberculosis, but due to repeated vomiting, ATT was stopped after 10 days. Subsequently, she was admitted to our hospital.

On general examination she had pallor, pitting edema, cachexia, and erythematous ulcer over hard and soft palate. On abdominal examination, there was free fluid, and vague palpable lump. The chest, CVS and CNS examination was normal. On routine investigation hemoglobin was 10.4 g/dL, TLC - 9800/µl, and platelets - 134×10^3/µl. Renal function test revealed BUN - 30 mg/dL, creatinine -1.2 mg/dL, uric acid - 14.56 mg/dL, calcium - 7.46 mg/dL, phosphorus - 2.2 mg/dL, sodium - 128 mEq/L, and potassium - 3.7 mEq/L. Liver function test revealed bilirubin (0.5 mg/dL), aspartate aminotransferase (186 IU/L), alanine aminotransferase (70 IU/L), alkaline phosphatase (117 IU/L), gamma glutamyl transferase (111 IU/L), protein (7 g/dL), and albumin (2.9 g/dL). Her erythrocyte sedimentation rate was 5 mm/hr, and Mantoux negative. The thyroid-stimulating hormone was 7.89 mIU/L. Approximately 2.3 L ascitic fluid was aspirated, in which cell count was 350 cell/µl, lymphocyte (80%), protein (3.2 g/dl), albumin (2 g/dL), SAAG (0.9), sterile culture, adenosine deaminase (8.4 U/L), and no malignant cells. The contrast enhanced CT chest and abdomen revealed heterogeneously enhancing mediastinal and left axillary lymph nodes, hepatomegaly, edematous small intestine, and colon with clumping of small bowel loops forming an abdominal cocoon, mild ascites, and bilateral mild pleural effusion (Figure 1). Endoscopic ultrasound guided fine needle aspiration cytology (FNAC) of subcarinal lymph node revealed no granuloma, and no evidence of malignancy. The ultrasound sonography test guided axillary lymph node biopsy showed reactive lymph nodes. Routine urine examination revealed nephritic range proteinuria (4+ albumin), pus cells (20/HPF), red blood cell (70-80), and culture revealed growth of *Escherichia coli*. A 24-hour urinary protein was 999.2 mg/dL. Bone marrow aspiration and biopsy revealed cellular bone marrow. Colonoscopy examination showed normal colonic mucosa. Antinuclear antibodies (ANA) was positive and anti-dsDNA was highly positive (551 IU/mL). The complement factor (C)-3 and C-4 were low. Renal biopsy revealed focal proliferative glomerulonephritis that is, World Health Organization stage III (Figure 2). Hence, using the diagnostic criteria, presence of oral ulcer, serositis, renal disorder, ANA and anti-dsDNA, a final diagnosis of SLE was established. She initially did not tolerate even a liquid diet, and a naso-jejunal tube was inserted and feeding was started. After intravenous methylprednisolone initiation and discontinuation of ATT, she had significant improvement in symptoms; her vomiting decreased, her appetite improved, and the ascites disappeared. Subsequently, the naso-jejunal tube was removed, as she resumed a normal diet. She was then discharged on oral steroids, mycophenolic acid, and hydroxychloroquine. She was doing well on follow-up.

**Discussion.** Systemic lupus erythematosus is a common autoimmune disorder characterized by multisystem involvements, including any part of the...
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GIT. Patients with SLE present with various GIT symptoms like nausea, vomiting, abdominal pain, gastroesophageal reflux, dysphagia, constipation, diarrhea, fecal incontinence, intestinal pseudo-obstruction, perforation, and hemorrhage. The incidence of GIT manifestations varies with the method of studies. In a study by Xu et al,22% had SLE-related gastrointestinal manifestations, and in 30.8% of these patients, it presented as initial symptoms. Out of these, 64.1% had abdominal pain, 56.4% had nausea and vomiting, 30.8% had diarrhea, and 7.7% had gastrointestinal hemorrhage.4

Sclerosing encapsulating peritonitis is a rare cause of bowel obstruction. It is characterized by a thick grayish-white fibrotic membrane, partially or totally encasing the small bowel, and can extend to involve other organs, such as the large intestine, liver, and stomach.5 The SEP can be classified as idiopathic, or secondary. The idiopathic form is classically described in young adolescent females from the tropical and subtropical countries, but is also seen in adults from non-tropical areas. The secondary form of SEP has been reported in association with continuous ambulatory chronic peritoneal dialysis.6 Other rare causes of secondary form of SEP include prior abdominal surgery, recurrent peritonitis, beta-blocker treatment (practolol), peritoneovenous shunting, ventriculoperitoneal shunting, and more rarely, abdominal tuberculosis, sarcoidosis, intraperitoneal chemotherapy, liver cirrhosis, liver transplantation, gastrointestinal malignancy, and fibrogenic foreign material.7 The SLE is also reported as a rare cause of SEP in the literature.3,7

For the diagnosis of SEP, a high index of clinical suspicion is required preoperatively. Most cases are diagnosed incidentally at laparotomy, although a preoperative diagnosis can be established by imaging which plays an important role in management. Plain abdominal x-ray may show air-fluid levels and ultrasonography may show dilated intestinal segments, free fluid and peritoneal membrane, if it is thick enough. A barium follow-through may show concertina pattern or cauliflower sign and delayed transit of contrast medium, CT is considered as the gold standard imaging modality for abdominal cocoon. Classic CT findings include ascites with small bowel loops congregated in a single area, or the concentration of the small bowel to midline encased by a soft-tissue density mantle.8,9 Lupus enteritis and abdominal cocoon have similar treatments. The conditions are generally reversible in response to treatment with corticosteroids, but the relapse is usual. In relapse, multiple drugs are used. Tacrolimus10 and mycophenolate mofetil11 are used recently in few cases.

Our patient has predominant presentation of abdominal cocoon with distention and vomiting. In Indian scenario, these patients are empirically treated with ATT, and may even be subjected to unnecessary laparotomy, as in our case. Presence of oral ulcer and significant proteinuria in this young female with history of joint pain, a suspicion of collagen vascular disease was considered, which was later confirmed by ds-DNA. Thus, this young girl was a case of abdominal cocoon with repeated vomiting and SAIO as a presenting feature of SLE.

In conclusion, we suggest that all patients with features of intestinal obstruction, especially with multisystem involvement should be screened for collagen vascular disease, so that a timely intervention after a correct diagnosis can be carried out, and patients can avoid unnecessary exposure to ATT, or laparotomy.

References

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