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*Case Report*

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# A therapeutic dilemma on lupus enteritis with bowel ischemia in a filipino female

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## Abstract

Lupus enteritis is rare and life-threatening. It is a differential among patients with Systemic Lupus Erythematosus (SLE) with severe abdominal pain. To our knowledge, this is the first locally reported case to date. A high index of suspicion and immediate treatment are necessary due to its high mortality rate and poor prognosis.

A 57-year-old Filipino female with a history of recurrent pregnancy loss, chronic left leg swelling, and a significant family history of thrombotic illnesses presented with a one-month history of abdominal pain, bloating, and vomiting. Abdominal pain was not proportionate to the physical exam. Workups showed thrombocytopenia, distal jejunum wall thickening, portal vein thrombosis, and gallbladder fossa and peripancreatic varices. She was diagnosed with partial small intestinal obstruction secondary to lupus enteritis, bowel ischemia, and portal vein thrombosis, with probable Antiphospholipid Syndrome (APS). She underwent segmental resection and jejunal anastomosis. Postoperatively, her lupus flare was treated with hydrocortisone despite the risks of anastomotic failure. Mycophenolate mofetil, hydroxychloroquine, and trimethoprim-sulfamethoxazole were added. Upon stabilization of platelet counts, fondaparinux was started and overlapped with warfarin. The patient eventually stabilized and was discharged.

Lupus enteritis affects up to 9.7% of patients with SLE and up to 65% of those presenting with an acute abdomen. Our patient was in severe organ-threatening disease with severe thrombocytopenia, needing steroids to control the flare and bowel resection to prevent necrosis. Corticosteroids are the cornerstone of therapy and are preferred over platelet transfusion in patients with SLE-associated thrombocytopenia. Oral vitamin K antagonists are the gold standard for APS with thrombosis. The co-existence of SLE with APS portends a poorer prognosis.

Due to the scarcity of data, weighing interventions' clinical risks and benefits was paramount in determining this patient's treatment course.

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## Introduction

Systemic lupus erythematosus, otherwise known as SLE, is a chronic disease in which autoantibodies and immune complexes damage multiple organs and cells. Treatment involves high-dose glucocorticoids. In the Philippines, SLE occurs in 30 to 50 per 100,000 of the population and commonly affects women of childbearing age [1,2]. In approximately 30% of patients, SLE is associated with Antiphospholipid Syndrome (APS). Treatment involves long-term anticoagulation [3].

Gastrointestinal symptoms are common in SLE and are often related to medication side effects or infections. However, 9.7% of patients with SLE and 65% of those with an acute abdomen are due to lupus enteritis. Lupus enteritis is the bowel wall inflammation caused by SLE, commonly involving the mesentery [4]. When damage is severe and occlusive, it may progress to bowel ischemia, obstruction, necrosis, perforation, and hemorrhage. This life-threatening complication of SLE is known as lupus mesenteric vasculitis and has a 50% mortality rate [5]. Mesenteric ischemia can also occur with APS, but data on the prevalence of SLE with secondary APS is lacking.

When complications arise, treatment involves aggressive immunotherapy with high-dose glucocorticoids and surgery. Postoperatively, balancing the relative increased risk for anastomotic leakage and controlling the flare among patients on steroids are essential. In addition, it is equally prudent to weigh the risk for bleeding and prevent thrombosis among patients with SLE in flare and APS on anticoagulants. This case report is an example of a multidisciplinary therapeutic dilemma and is the first such local report.

## Case report

A 57-year-old Filipino female with a history of chronic left leg swelling was admitted due to progressive diffuse, crampy abdominal pain, bloatedness, and vomiting for one month. She has a history of stillbirth at six months gestational age and blighted ovum at three months gestational age. Family history includes stroke, aneurysm, pulmonary embolism, intestinal vasculitis, Deep Vein Thrombosis (DVT), SLE, APS, and postpartum hemorrhage.

On admission, abdominal pain was not proportionate to physical findings of non-distention, hypoactive bowel sounds, tympanitic sound, direct tenderness at epigastric and left hemiabdomen, and absence of rigidity, guarding, or rebound tenderness. For the consideration of mesenteric ischemia, a whole abdomen Computed Tomography (CT) scan with triple contrast showed wall thickening in the distal jejunum (Figures 1A and 1B). CT angiography of the abdominal aorta revealed portal vein thrombosis (Figure 2A) with gallbladder fossa and peripancreatic varices (Figures 2B and 2C).

During this time, partial small intestinal obstruction from lupus enteritis with mesenteric ischemia was considered. Workups revealed platelets 39x10$^9$/L, positive Antinuclear Antibody (ANA), low Complement 3 (C3), borderline positive anti-double stranded DNA (anti-dsDNA) antibody, microscopic hematuria, and left leg DVT. Lupus anticoagulant diluted Russell Viper Venom Time (dRVVT), anticardiolipin, and beta-2 glycoprotein I antibodies were negative. However, due to high suspicion of APS, the patient was advised to repeat the tests after twelve weeks.

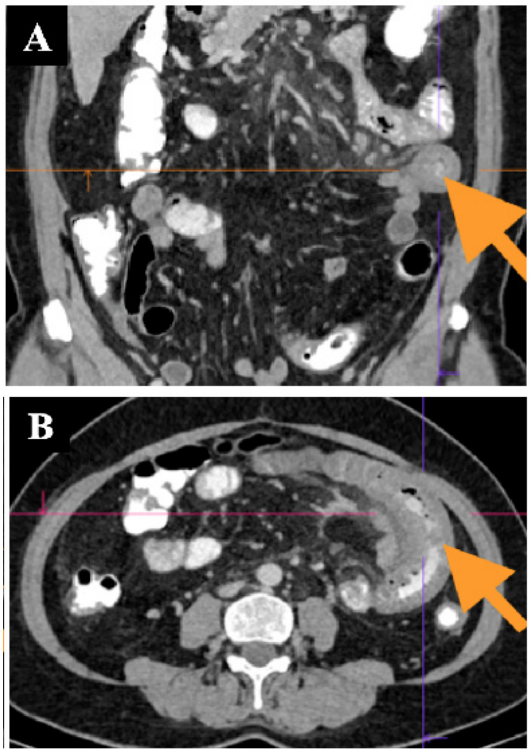


**Figure 1:** Whole abdomen CT scan with triple contrast showing persistent circumferential enhancing mucosal wall thickening in the distal jejunum **(A** and **B)**.

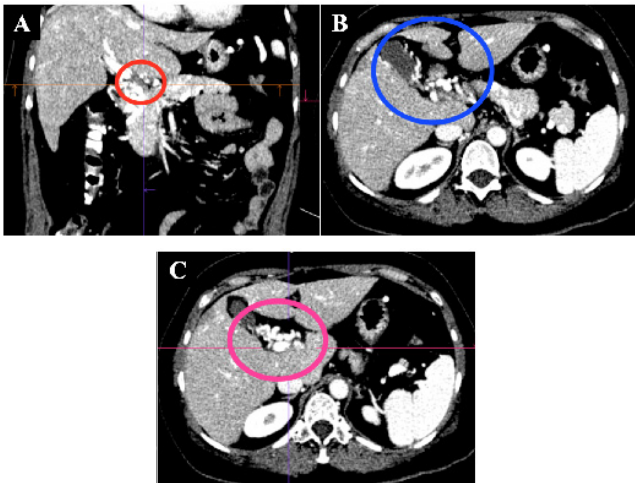


**Figure 2:** Abdominal aortogram and venogram showing persistent intraluminal filling defect in the main portal vein suggestive of portal vein thrombosis **(A)** with gallbladder fossa and peripancreatic varices **(B** and **C)**.

Worsening symptoms of intestinal obstruction prompted segmental resection and jejunal anastomosis, where histopathology showed inflammation. Lupus flare was postoperatively confirmed with positive ANA at 1:160 (homogenous pattern), and Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2K) of 25 points consisting of vasculitis, hematuria, proteinuria, malar rash, low C3, positive anti-dsDNA antibody, 38.6°C body temperature, platelets 19x10$^9$/L, and leukocytes 2.79x10$^9$/L. Hydrocortisone 1 mg/kg/day was started despite anastomosis failure risk. Mycophenolate mofetil 500 mg/tablet twice daily, hydroxychloroquine 200 mg/tablet once daily, and

trimethoprim-sulfamethoxazole 800/160 mg/tablet triweekly were added. Symptoms resolved, and platelet count stabilized. To prevent the progression of bowel ischemia, fondaparinux 2.5 mg once daily was started and was overlapped with warfarin 2.5 mg once daily to target an International Normalized Ratio (INR) of 2.0-3.0. Hydrocortisone was shifted to prednisone 20 mg/tablet twice daily and was tapered. Further workups revealed elevated lipid profile and fasting blood glucose. In addition to diet modification, atorvastatin 10 mg/tablet once daily, ezetimibe 10 mg/tablet once daily, fish oil capsule once daily, and linagliptin 5 mg/tablet half tablet once daily were started. The disease activity was controlled, and diet progression was tolerated. The patient was discharged stable.

On follow-up one month after discharge, the patient had no new symptoms. Workups showed platelets $120x10^9$/L, positive ANA at 1:160 (homogenous pattern), negative dRVVT and anticardiolipin antibody, microscopic hematuria, and trace proteinuria. Hemoglobin, leukocytes, INR, creatinine, and anti-dsDNA antibody were normal. Warfarin, hydroxychloroquine, mycophenolate mofetil, and trimethoprim-sulfamethoxazole were continued. Prednisone was tapered until discontinued.

## Discussion

We presented a rare case of lupus enteritis as the first clinical manifestation of SLE. It is unusual for the gastrointestinal tract to be involved without other lupus symptoms since other organs are generally affected first. In the Philippines, data on the incidence of lupus enteritis as a manifestation of SLE is lacking. Lupus enteritis usually affects females, and symptoms are nonspecific, including abdominal pain, ascites, nausea, vomiting, and diarrhea. In 80 to 85% of cases, the superior mesenteric artery is involved, which affects the ileum and jejunum [6].

Our patient was in severe organ-threatening disease with severe thrombocytopenia, needing steroids to control the flare and bowel resection to prevent necrosis. To prevent new flares and achieve remission or low disease activity, the 2019 European Alliance of Associations for Rheumatology (EULAR) recommends methylprednisolone pulses 250-1000 mg/day for 1 to 3 days, followed by prednisone 0.5-0.7 mg/kg/day with gradual tapering for severe or organ threatening disease [7]. Due to its availability, hydrocortisone was contemplated in our patient to control the flare.

Anastomotic leakage is a serious postsurgical complication associated with increased morbidity and mortality. Studies show that glucocorticoid use exhibited an increased relative risk of anastomotic leakage after colorectal surgery than in small intestinal surgery [8]. In our patient, hydrocortisone was discontinued preoperatively due to the risk of anastomotic leakage, given the proximity of the resection to the ligament of treitz and the risk for small bowel syndrome. Hydrocortisone was later resumed due to its benefits during postoperative SLE flare.

Thrombocytopenia is reported in 20% to 40% of patients with SLE. Corticosteroids are the cornerstone of therapy and are preferred over transfusion [9]. Thrombocytopenia is a relative contraindication to surgery due to bleeding risk [10]. Current guidelines regarding prophylactic platelet transfusion and thrombopoietin receptor agonists to prevent bleeding are mainly based on expert opinion rather than sound evidence [11]. Prophylactic transfusion was not yet indicated on our patient because platelets were above $10x10^9$/L, and there was no active bleeding.

Autoimmune diseases with APS are associated with an increased thrombotic risk [12]. The gold standard treatment for thrombosis is oral vitamin K antagonist to target an INR of 2.0–3.0. Recurrence rates without anticoagulation are high; thus, lifelong anticoagulation is generally accepted [13]. Low Molecular Weight Heparin (LWMH) is recommended if thrombocytopenia is present. Fondaparinux is considered when warfarin and LMWH have failed. Dual anticoagulants should be avoided due to recent evidence of increased risk for arterial events [1]. Although our patient was negative for APS workup, treatment was pursued postoperatively to prevent thrombosis due to high suspicion of APS. Fondaparinux was initially chosen over enoxaparin for mesenteric ischemia due to the fear of heparin-induced thrombocytopenia. It was later shifted to warfarin. The need to balance the risk of bleeding and recurrent thrombosis must be decided carefully. An individualized approach was made for our case.

Studies show that trimethoprim-sulfamethoxazole prophylaxis for systemic lupus erythematosus on immunosuppression prevents major infections [14] and was thus started on our patient.

Because patients with SLE are at increased risk for hyperlipidemia and diabetes, workup was done for our patient and had elevated results. Along with diet modification, atorvastatin, ezetimibe, fish oil, and linagliptin were started. Conventional arterial risk factors such as being overweight, smoking, hypertension, hyperlipidemia, and poor glycemic control should be addressed to minimize the risk of recurrent arterial thrombosis [1].

The researchers recommend a detailed clinical history, physical examination, and maximal utilization of the diagnostic modalities as it can help unravel the diagnoses and guide the managing team in the treatment options.

## Conclusion

Among patients with lupus enteritis, balancing the risks and benefits of an intervention is crucial. A multidisciplinary and individualized approach is paramount in saving the patient.

**Abbreviations:** ANA: Antinuclear Antibody; APS: Antiphospholipid Syndrome; Anti-dsDNA: Anti-double Stranded DNA; C3: Complement 3; CT: Computed Tomography; dRVVT: Diluted Russell Viper Venom Time; DVT: Deep Vein Thrombosis; EULAR: European Alliance of Associations for Rheumatology; INR: International Normalized Ratio; LMWH: Low Molecular Weight Heparin; SLE: Systemic Lupus Erythematosus; SLEDAI-2K: Systemic Lupus Erythematosus Disease Activity Index 2000.

## Declarations

**Conflicts of interest:** The authors declare no conflict of interest.

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