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A therapeutic dilemma on lupus enteritis with bowel ischemia presenting as small intestinal obstruction in a Filipino female: A case report

Maderazo, PAL[1]

Hererra, CY[1]

Vaje, CJV[1]

Obrado-Nabablit, GMM[2]

Chan, RC[3]

Amante, EJB[4]

[1]*Department of Internal Medicine*
[2]*Section of Cardiology, Department of Internal Medicine*
[3]*Section of Gastroenterology, Department of Internal Medicine*
[4]*Department of Medicine*

[1, 2, 3] *Manila Doctors Hospital*
United Nations Avenue, Ermita
Manila 1000, Philippines

[4] *Philippine General Hospital*
Taft Avenue, Ermita
Manila 1000, Philippines

Correspondence:
Princess Aurea L. Maderazo, MD
maderazo.princess@yahoo.com

Abstract

Significance: Lupus enteritis is rare and life-threatening. Lupus enteritis, as the main manifestation of SLE, is underreported in the Philippines. To our knowledge, this study is the first local report of this case. A high index of suspicion and immediate treatment are necessary due to its high mortality rate and poor prognosis.

Clinical Presentation: A 57-year-old Filipino female from Leyte, Philippines, with a history of recurrent pregnancy loss, chronic left leg swelling, and significant family history of thrombosis presented with a one-month history of diffuse, crampy, intermittent, and progressive abdominal pain, graded 3/10, non-radiating, aggravated by food intake, and associated with bloatedness and vomiting. The severity of abdominal pain did not correlate with the abdominal physical examination. Workups showed thrombocytopenia, positive antinuclear antibody (ANA), low complement component 3 (C3), borderline positive anti-double-stranded deoxyribonucleic acid (anti-dsDNA) antibody, and negative anti-Smith antibody. Whole abdomen computed tomography (CT) scan with triple contrast revealed distal jejunum wall thickening. CT angiography of the abdominal aorta showed portal vein thrombosis. Venous duplex study revealed left leg deep-venous thrombosis (DVT). Antiphospholipid syndrome (APS) was highly suspected despite having negative lupus anticoagulant diluted Russell viper venom time (dRVVT), anticardiolipin antibodies (ACA) IgG and IgM, and beta-2 glycoprotein I (β2GPI) antibodies IgG and IgM. The patient was advised to repeat dRVVT, ACA, β2GPI antibodies, and kaolin clotting time (KCT) after 12 weeks, for further evaluation. She was diagnosed with partial small intestinal obstruction secondary to lupus enteritis, bowel ischemia, and portal vein thrombosis, with probable APS.

Management: Worsening symptoms of intestinal obstruction prompted exploratory laparotomy, segmental resection, and anastomosis of the jejunum under general endotracheal anesthesia. Lupus flare was postoperatively confirmed. Hydrocortisone was started despite the risks of anastomotic failure. Mycophenolate mofetil, hydroxychloroquine, and trimethoprim-sulfamethoxazole were added. After the platelet counts stabilized, fondaparinux was started and later overlapped with warfarin. The patient was discharged, improved and stable. Due to the distance of her province and the limitations imposed by the pandemic, most of the patient's follow-up was conducted online. On follow-up one month after discharge, the patient remained well and stable. Workups were unremarkable. The patient was unable to undergo the follow-up tests for APS (dRVVT, ACA, β2GPI antibodies, and KCT) due to their limited availability in her province. Warfarin, hydroxychloroquine, mycophenolate mofetil, atorvastatin, ezetimibe, fish oil, and linagliptin were continued. Prednisone was tapered until discontinued.

Recommendations: Management of lupus enteritis involves a multidisciplinary and individualized approach. A comprehensive history and physical examination, along with careful assessment of an intervention's risks and benefits, are crucial.

Keywords: *Antiphospholipid syndrome, Case report, Lupus enteritis, Mesenteric ischemia, Philippines, Small intestinal obstruction, Systemic lupus erythematosus*

Introduction

Systemic lupus erythematosus (SLE) is a chronic disease in which autoantibodies and immune complexes damage multiple organs and cells. Treatment involves 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 30% of patients, SLE is associated with antiphospholipid syndrome (APS). Treatment involves long-term anticoagulation.[3]

In 9.7% of patients with SLE, 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] Lupus enteritis, as the main manifestation of SLE, is underreported in the Philippines. Lupus mesenteric vasculitis (LMV) involves inflammation of the vessels of the gastrointestinal tract, hence the need for steroids. If unresponsive to steroids, surgery is considered to prevent bowel ischemia, perforation and hemorrhage.[5] Postoperatively, it is important to monitor for anastomotic leakage, bleeding and thrombosis. LMV has a 50% mortality rate. LMV most likely occurs in SLE with high Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2K) score.[5]

All published literature until May 2025 regarding lupus enteritis in the Philippines from the following databases were reviewed: PubMed, Cochrane, Google scholar, and Embase. The following Mesh terms were used: “antiphospholipid syndrome”, “case report”, “Filipino”, “lupus enteritis”, “mesenteric ischemia”, “Philippines”, “small intestinal obstruction”, and “systemic lupus erythematosus”. To our knowledge, this study was the first local report of this case.

The researchers aim to discuss the approach to treatment, understand the importance of anticoagulation, and analyze the possible complications in patients with lupus flare, bowel ischemia, portal vein thrombosis, probable APS, and severe thrombocytopenia.

Clinical Presentation

A 57-year-old Filipino female from Leyte, Philippines, was admitted at a hospital in Manila, Philippines due to a one-month history of diffuse, crampy, intermittent, and progressive abdominal pain, graded 3/10, non-radiating, aggravated by food intake, and associated with bloatedness and vomiting. Past medical history included chronic left leg swelling, stillbirth at six months gestational age, and blighted ovum at three months gestational age. Family history included stroke, aneurysm, pulmonary embolism, intestinal vasculitis, deep-venous thrombosis (DVT), SLE, APS, and postpartum hemorrhage.

On admission, she felt severe abdominal pain that did not correlate with the abdominal physical examination: non-distention, hypoactive bowel sounds, tympany, and direct tenderness in the epigastric and left hemiabdomen areas; no abdominal rigidity, guarding, or rebound tenderness. This led to the consideration of mesenteric ischemia.

For the impression of intestinal obstruction from lupus enteritis with mesenteric ischemia, workups revealed platelets $39x10^9$/L, positive antinuclear antibody (ANA) screening, low complement component 3 (C3), borderline positive anti-double-stranded deoxyribonucleic acid (anti-dsDNA) antibody, negative anti-Smith antibody and microscopic hematuria. Urine albumin-creatinine ratio was elevated at 55.7 mg/g. Serum creatinine was 68 umol/L, with an estimated glomerular filtration rate of 85.8 mL/min/1.73^2. Whole abdomen computed tomography (CT) scan with triple contrast showed distal jejunum wall thickening (Figures 1A and 1B). CT angiography of the abdominal aorta revealed portal vein thrombosis (Figure 2A) with gallbladder fossa varices (Figure 2B) and peripancreatic varices (Figure 2C). Carcinoembryonic antigen (CEA) test was normal.

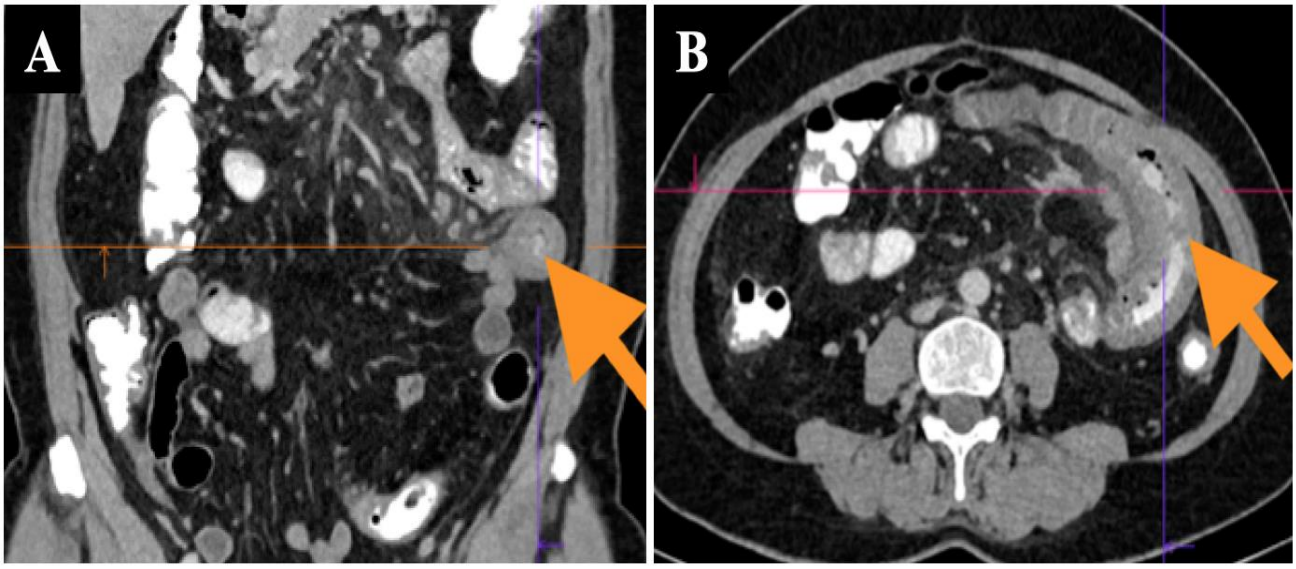

Figure 1. Whole abdomen CT scan with triple contrast showing persistent circumferential enhancing mucosal wall thickening in the distal jejunum (A and B, orange arrow) with associated mesenteric lymphadenopathies and mesenteric reactive changes

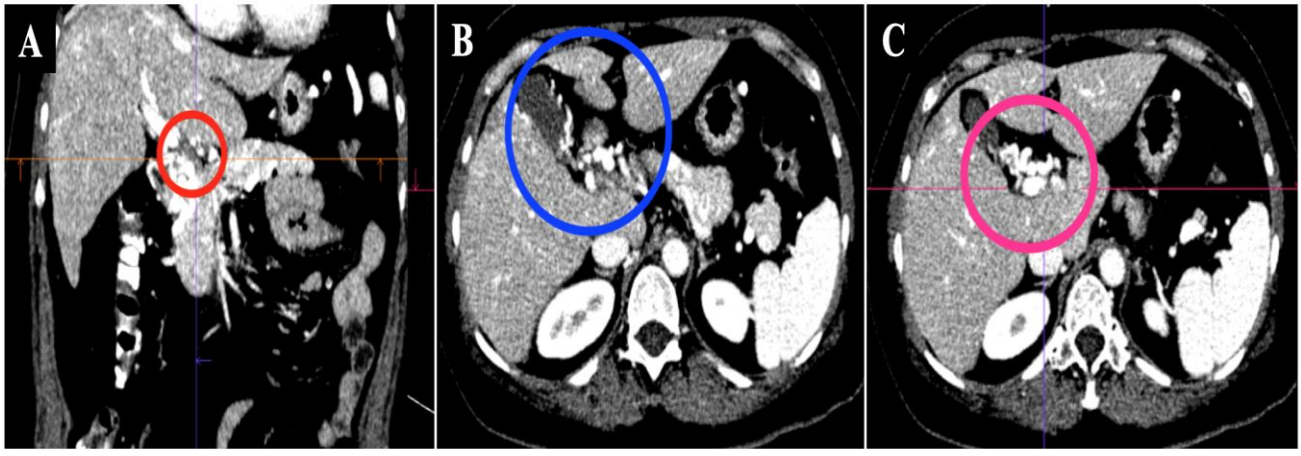

Figure 2. Abdominal aortogram and venogram showing persistent intraluminal filling defect in the main portal vein prior to the bifurcation of the left and right portal vein suggestive of portal vein thrombosis (A, red circle) with gallbladder fossa varices (B, blue circle) and peripancreatic varices (C, pink circle). No evidence of aneurysmal dilatation, dissection, thrombus formation or stenosis along the entirety of the abdominal aorta and its branches as well as the inferior vena cava and right major tributaries

Workups for hypercoagulable state were done due to a strong family history of thrombosis. D-dimer was elevated. Prothrombin time and activated partial thromboplastin time were normal. Peripheral blood smear showed marked thrombocytopenia with few large platelets. Venous duplex study revealed left leg DVT. Lupus anticoagulant diluted Russell viper venom time (dRVVT), anticardiolipin antibodies (ACA) IgG and IgM, and beta-2 glycoprotein I (β2GPI) antibodies IgG and IgM were negative. The patient was referred to hematology, vascular, rheumatology and surgery services. The patient refused referral to renal service for possible kidney biopsy due to financial constraints. Supportive care and trial feeding were done and tolerated. Fondaparinux 5 mg once daily was started subcutaneously. Due to high suspicion of APS, the patient was advised to repeat dRVVT, ACA IgG, ACA IgM, β2GPI antibody IgG, β2GPI antibody IgM, and kaolin clotting time (KCT) after 12 weeks.

On the 4th hospital day, the patient did not tolerate the trial feeding. Patient was further observed but at this time, surgery was already contemplated; hence, hydrocortisone was temporarily deferred to prevent the risk of impaired wound healing, anastomotic leakage, and subsequent morbidity.

On the 9th hospital day, worsening symptoms of intestinal obstruction prompted exploratory laparotomy, segmental resection, and anastomosis of the jejunum under general endotracheal anesthesia, where histopathology showed inflammation. The patient was stable post-procedure. Steroids were avoided to prevent potential disruption of the surgical anastomosis' healing. The postoperative platelet count decreased; hence, fondaparinux was put on hold.

On the 14th hospital day, the patient was seen with ecchymosis at the postoperative site, and upper and lower extremities. Lupus flare was postoperatively confirmed with positive ANA at 1:160 (homogeneous pattern), and SLEDAI-2K score 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. As agreed upon by all services, hydrocortisone 1 mg/kg/day was started for the lupus flare. Hydroxychloroquine 200 mg/tablet once daily was added to reduce the disease flare and increase the sustainability effect of steroids. Mycophenolate mofetil 500 mg/tablet twice daily was added to improve platelet counts. Trimethoprim-sulfamethoxazole 800/160 mg/tablet triweekly was started for prophylaxis against major infections while on immunosuppressants.[6] Symptoms resolved, and platelets increased to $56x10^9$/L. Upon further platelet stabilization, fondaparinux 5 mg once daily was started subcutaneously and was later overlapped with warfarin 2.5 mg/tablet 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.

Because patients with SLE were at increased risk for hyperlipidemia and diabetes, workups were done, and revealed elevated results of low-density lipoprotein, alanine transaminase, aspartate transaminase, and fasting blood sugar. Along with diet modification, atorvastatin 10 mg/tablet, ezetimibe 10 mg/tablet, fish oil capsule, and linagliptin 2.5 mg/tablet were started once daily to minimize the risk for recurrent arterial thrombosis.[1] The diet progression was tolerated. The disease activity improved, as indicated by a decrease in the SLEDAI-2K score to 14 points, which included vasculitis, positive anti-dsDNA, and proteinuria. The platelets increased to $134x10^9$/L.

After 33 hospital days, the patient was discharged, was stable with a diagnosis of partial small intestinal obstruction from lupus enteritis with bowel ischemia, portal vein thrombosis and probable APS.

Due to the distance of Leyte province from Metro Manila and the limitations imposed by the pandemic, most of the patient's follow-up was conducted online. On follow-up one month after discharge, she was well and had no fever, pain, vomiting, and bleeding. She had good compliance and tolerability. She was able to return to her daily activities. Workups showed platelets $120x10^9$/L, positive ANA at 1:160 (homogeneous pattern), microscopic hematuria, and trace proteinuria. Hemoglobin, leukocytes, INR, creatinine, and anti-dsDNA antibody were normal. The patient was unable to undergo the follow-up tests for APS (dRVVT, ACA, β2GPI antibodies, and KCT) due to their limited availability in her province. Warfarin, hydroxychloroquine, mycophenolate mofetil, atorvastatin, ezetimibe, fish oil and linagliptin were continued. Prednisone was tapered until discontinued.

Discussion

We present a rare case of lupus enteritis as the first clinical manifestation of SLE. Lupus generally affects other organs first before the gastrointestinal tract. Lupus enteritis usually affects females. Symptoms include abdominal pain, ascites, nausea, vomiting, and diarrhea. The ileum and jejunum are commonly involved.[7]

APS occurs in 30% of patients with SLE. Diagnosis requires at least one clinical criterion (vascular thrombosis or pregnancy morbidity), and one laboratory criterion (intermediate or high titers of lupus anticoagulant, ACA, and/or β2GPI antibodies on two occasions 12 weeks apart).[1] Despite negative initial laboratory results, our patient fulfills the clinical criteria for APS: vascular thrombosis (deep-venous thrombosis and portal vein thrombosis), and pregnancy morbidity (stillbirth at six months gestational age, and blighted ovum at three months gestational age). In addition, our patient is female, has thrombocytopenia, and a significant family history of thrombotic events.

Our patient had severe thrombocytopenia and was at risk for bleeding. She also had bowel inflammation and ischemia, and was at risk for necrosis. For severe life-threatening conditions like these in patients with lupus, the 2019 European Alliance of Associations for Rheumatology recommended methylprednisolone pulses 250 to 1000 mg/day for 1 to 3 days, followed by prednisone 0.5 to 0.7 mg/kg/day with gradual tapering.[8] However, due to resource limitation, our patient was instead treated with hydrocortisone 1 mg/kg/day and bowel resection with good response.

Anastomotic leakage is a serious postsurgical complication associated with increased morbidity and mortality. A prospective study by Jina and Singh (2019) involving patients who underwent intestinal anastomosis shows that patients with peritonitis, sepsis or bowel obstruction pre-operatively or during admission, developed more anastomotic leaks. There is also a considerably higher rate of anastomotic leakage in patients on corticosteroids.[9]

In our patient, hydrocortisone was avoided pre-operatively to prevent the risk of impaired wound healing, anastomotic leakage and subsequent morbidity. Hydrocortisone was started at 1 mg/kg/day (equivalent to 75 mg prednisone) postoperatively due to its benefits for lupus flare. In patients undergoing major colorectal surgery, Kane and Berry (2023) recommended withholding steroids at least 4 weeks pre-operatively, and resuming it on the day postoperatively. Absolute recommendation could not be made for patients receiving more than 5 mg prednisone because of unclear suppression of hypothalamic–pituitary-adrenal axis.[10]

Thrombocytopenia was reported in 20% to 40% of patients with SLE and was rarely severe.[11] Glucocorticoids were the first-line treatment and were preferred over transfusion.[11,12] Immunosuppressive agents were second-line.[11] Glucocorticoids were recommended for platelets <$30x10^9$/L in asymptomatic adults with new immune thrombocytopenia (ITP).[13] In ITP and APS, thrombocytopenia might paradoxically increase thrombosis compared to bleeding. When our patient's platelets reached $19x10^9$/L, hydrocortisone, hydroxychloroquine and mycophenolate mofetil were started to decrease thrombosis recurrence. Prophylactic transfusion was unwarranted because platelets were above $10x10^9$/L, and there was no active bleeding.[14]

Although our patient had negative initial APS work up (dRVVT, ACA IgG, ACA IgM, β2GPI antibody IgG and β2GPI antibody IgM), treatment for thrombosis prevention was pursued due to high clinical APS suspicion. Oral vitamin K antagonist was the standard treatment in thrombotic APS; however, it was not recommended if platelets were <$50x10^9$/L.[11,15] Enoxaparin was initially considered for our patient as anticoagulant for mesenteric ischemia and probable APS. However, fondaparinux was chosen due to the severity of thrombocytopenia and to avoid heparin-induced thrombocytopenia. Upon platelet count recovery, fondaparinux was shifted to warfarin to target an INR of 2.0 to 3.0 for venous thrombosis.[1] Lifelong anticoagulation was recommended because of high thrombosis recurrence rates without anticoagulation.[16] However, lifetime anticoagulation was associated with increased bleeding risk. A balance between the bleeding risk and anticoagulation benefits

was crucial. A multidisciplinary and individualized approach for each patient was necessary.

Other relevant differential diagnoses for our patient included infectious enteritis, diverticulitis, and colon cancer. Infectious enteritis was considered because it is a common cause of enteritis, but was ruled out due to the absence of fever, hematochezia, loss of appetite, similar symptoms at home, recent travel, or intake of contaminated water. Additionally, stool exam did not show fecal leukocytes, blood, parasites, or ova. Diverticulitis was considered because of the patient's age, but was ruled out due to the absence of fever, constipation, or diarrhea. Moreover, CBC did not show leukocytosis, and whole abdomen CT scan with triple contrast did not reveal pericolic fat stranding, colonic wall thickening, fistula, phlegmon, abscesses or peritonitis. Colon cancer was considered because of the patient's age, but was ruled out due to the absence of diarrhea, constipation, rectal bleeding, weight loss, or family history. Furthermore, CBC did not reveal anemia, whole abdomen CT scan with triple contrast did not show any mass in the colon and CEA test was negative.

The limitations of the initial diagnostic workup were the inability to perform the serum complement 4 and KCT due to limited availability. In addition, because the patient had an elevated urine albumin-creatinine ratio of 55.7 mg/g, a kidney biopsy could have been performed for diagnostic and prognostic purposes.

The patient was followed up closely after discharge to monitor the disease activity of her SLE, and the tapering of her warfarin and prednisone dose. Due to the distance of Leyte province from Metro Manila and the limitations imposed by the pandemic, most of the patient's follow-up were conducted online. The patient was also unable to repeat the APS laboratories (dRVVT, ACA, β2GPI antibodies, and KCT) after 12 weeks due to their limited availability in her province.

The researchers recommend further studies on association of anastomotic leakage and high dose glucocorticoids, safety of glucocorticoids for patients with lupus undergoing small intestinal resection, and severe thrombocytopenia management in APS.

Conclusion

Lupus enteritis is a rare, life-threatening condition primarily treated with steroids and, if warranted, with surgery. Lifelong anticoagulation is recommended if with APS; however, it increases the bleeding risk, especially if with severe thrombocytopenia. Balancing the risks and benefits of anastomotic leakage, flare control, bleeding tendency, and thrombosis prevention is crucial to prevent unwarranted complications and subsequent morbidities. A multidisciplinary and individualized approach is necessary.

Ethics and Integrity Policy

Financial or funding statement:

This case report received no sources of funding.

Conflict of interest disclosure:

The authors declared no actual or potential financial, consultant, institutional, or other conflict of interest.

Ethics approval statement:

This study was conducted in accordance with the Declaration of Helsinki, and the protocol was approved by the Manila Doctors Hospital (MDH) Institutional Review Board (IRB).

Patient consent statement:

Before participating in our study, the patient read and signed an informed consent form for the presentation and publication of this case. The informed consent form used had received institutional legal approval. The patient possessed a copy of the form and understood that all personal identification would remain confidential.

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