What Do These Eosinophils Mean? Treatment Naive Systemic Lupus Erythematous Presenting With Eosinophilic Ascites, Eosinophilic Pleural Fluid, and Enteritis of Unclear Etiology

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INTRODUCTION

- Eosinophilic ascites (EA), lupus enteritis (LE), and eosinophilic enteritis (EE) are rare systemic lupus erythematosus (SLE) complications. When they do occur, overlapping diagnostic criteria causes problems in distinguishing between a SLE-related etiology or EE. For example, intestinal eosinophil infiltration associated with EE is also consistently found in disseminated SLE autopsy case reports[1]. Although both conditions are treated with steroids, therapies after treatment failure are different. We present a rare case of a SLE patient with this diagnostic dilemma with suboptimal response to SLE therapies, questioning whether an EE treatment algorithm may be more appropriate.

CASE PRESENTATION

History of Present Illness

- 55 year old female who presented with complaints of abdominal distention, abdominal pain, bloating, and constipation.
- 3 year history of SLE that was untreated due to socioeconomic barriers. Course complicated by chronic pain managed with opioids and recurrent pulmonary embolism managed with apixiban. Condition associated with unintentional weight loss of 30lbs over 2 years.
- 2 weeks prior, presented to another hospital where her constipation had minimal response to Miralax, Colase, Doculax suppositories, & 2 doses of Resistor. Denied baseline constipation. Complaint was attributed to acute idiopathic pancreatitis diagnosed by imaging and abdominal pain with a normal lipase.

Physical Exam

- Intermittent tachypnea and tachycardia but had otherwise normal vital signs. No anasarca.
- Soft, distended, mildly diffusely tender abdomen - worst in the epigastrium.

Laboratory Workup

- Normal electrolytes & hepatobilliary enzymes except for low albumin (3.1)
- No peripheral eosinophilia

C-Reactive Protein	<1 mg/dL
ESR	5 mm/h
Antinuclear Ab	Positive (1:160)
Anti-DS DNA	Positive
Anti-SSa/Ro	Positive
Anti-SmDp-s	Positive
Anti-uRNP1	Positive
Anti-ENA	Positive
ANCA Panel	Negative
C3	37mg/dL ↓
C4	<4 ↓↓
Stool Studies	Negative for infections or parasites

Ascitic Protein	3.9 g/dL ↑
Ascitic WBC	513 ↑
Ascitic Eosinophils	**71%.** ↑
Ascitic Lipase	13 U/L
SAAG	0.9 g/dL ↓
Fluid Classification	Exudative
Cytology	No malignancy or infection

Pleural WBC	292
Pleural Eosinophils	**51%.** ↑
Fluid Classification	Exudative ↓
Cytology	No malignancy or infection

Imaging

- **CT abdomen/pelvis with IV contrast:** moderate ascites, continuous small bowel thickening, ileum, and bilateral pulmonary infiltrates with pleural effusions. Similar findings were found on MRI enterography.

Endoscopy

- **Esophagoduodenoscopy (EGD):** erythematous antrum with pathology indicating mild chronic inactive gastritis. No H.pylori.
- **Colonoscopy:** Moderately hyperemic friable mucosa with loss of vasculature in patches involving the right colon (*figure 1*), a normal terminal ileum, and areas of mild hyperemia in the left colon. Pathology was non-specific for either transient colitis or early ischemic colitis. No eosinophils were present. Overall findings suspicious for lupus vasculitis.

Hospital Course

- Rheumatology diagnosed her with SLE serositis with peritoneal and pleural involvement complicated by lupus enteritis. However, they were unable to rule out serosal eosinophilic enteritis due to her being a poor candidate for full thickness small bowel biopsy. [2,3,4,5,6]
- Started on intravenous (IV) solumedrol (methylprednisolone) at 1mg/kg/day with slow improvement over 10 days.
- She was discharged on oral prednisone and steady decline outpatient with her re-presenting to the hospital within 2 weeks of discharge.
- After prolonged hospitalization, she was discharged on a regimen of hydroxychloroquine and oral prednisone. She re-presented to the hospital within 1 week of starting a steroid taper.
- During her most recent hospitalization, she was transitioned to a regimen of hydroxychloroquine, mycophenolate mofetil, and oral prednisone.

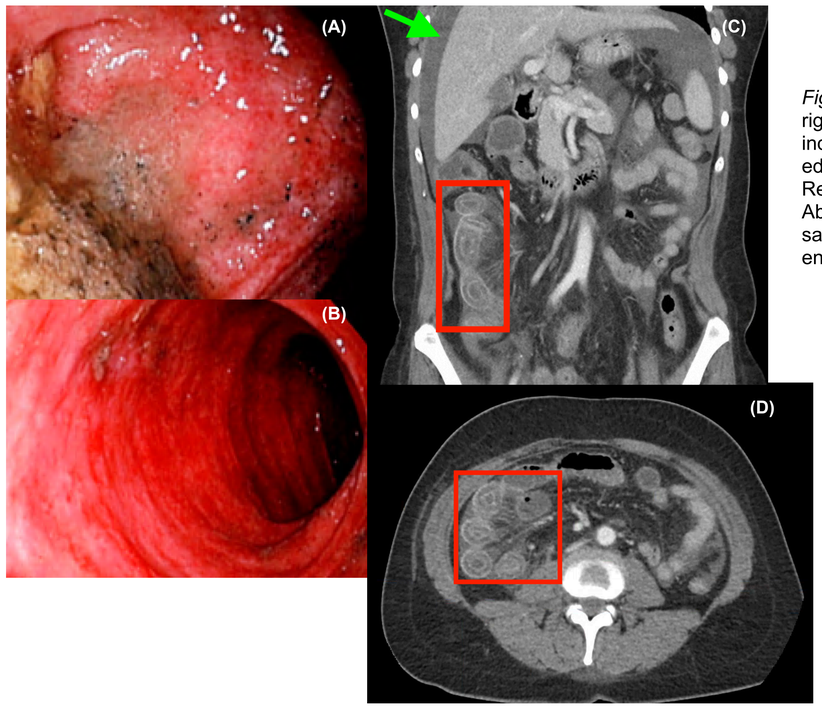

Figure 1. **(A & B)** Endoscopically visualized moderately hyperemic right colon with patchy loss of vasculature **(C)** Green arrow indicating ascites and red box highlighting characteristic bowel wall edema (target sign) on CT Abdomen/Pelvis with IV Contrast. **(D)** Re-demonstrating the "target sign" on transverse view of CT Abdomen/Pelvis with IV Contrast. Presence of a"Target Sign" satisfies the radiologic evidence criteria for diagnosis of lupus enteritis.[2,5]

DISCUSSION

- The main characteristics of serosal EE presenting as eosinophilic ascites are high ascitic eosinophil percentage (up to 90%) and a robust response to corticosteroids.[4,6]
- Typical biopsies on endoscopy are rarely diagnostic for serosal EE and absence of peripheral eosinophilia does not exclude the diagnosis.[4,6]
- Both lupus enteritis and serosal EE are poorly characterized and differentiated in the existing literature. Although they have overlapping features, the diseases have minimal overlap in their treatment algorithms.[5,6]
- It is unclear whether our patient's sub-optimal therapy response is due to SLE or EE. However, with her symptoms are attributed to serositis, her therapeutic strategy has been to escalate SLE immunosuppressive therapy rather than re-examine the diagnosis.

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