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Severe gastrointestinal manifestations in childhood-onset lupus: A single-center cohort and review of the literature

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Abstract

Objective: Systemic lupus erythematosus (SLE) is a complex autoimmune condition with diverse manifestations. Childhood-onset SLE (cSLE) presents more severely than adult-onset disease with a higher incidence of severe neurologic or renal manifestations and lower estimated survival rates. There is limited literature on the range of severe gastrointestinal manifestations at presentation in cSLE, and this lack of information results in underappreciation of these potentially life-threatening complications and little guidance regarding management of these patients.

Methods: We reviewed cases of patients with cSLE and severe GI manifestations at presentation diagnosed at our institution and additionally provide a comprehensive review of existing cases in the literature to discuss presenting symptoms, other clinical features, longitudinal course, treatment, and long-term outcomes.

Results: We identified six cases of cSLE with primary, severe GI symptoms at time of diagnosis at our institution and an additional 25 patients who presented similarly in the literature. Severe GI manifestations included protein-losing enteropathy, thoracoabdominal aortitis, severe pancreatitis, lupus enteritis or mesenteric vasculitis, and intestinal pseudo-obstruction. Delays in diagnosis affected several patients, and 26% of patients required surgical intervention. Many patients required intensive immunomodulatory treatment in addition to prolonged bowel rest and parenteral nutrition. Long-term outcomes of GI manifestations varied overall, however, most patients at our institution achieved clinical remission.

Conclusions: Understanding these rare cases of severe GI manifestations in cSLE can aid clinicians in prompt diagnosis and collaborative management, improving clinical outcomes for this vulnerable population.

Keywords

Protein-losing enteropathy, thoracoabdominal aortitis, pancreatitis, enteritis, intestinal pseudo-obstruction

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Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune condition with heterogeneous manifestations.[1] It is estimated that the incidence and prevalence of SLE in North America is 23/100,000 people per year and 241/100,000 people respectively with 10%–20% of cases being childhood-onset SLE (cSLE).[2,3] Despite similarities in laboratory findings, pathophysiology, and management, cSLE has a more severe disease presentation and course, with lower estimated survival rates and a two to three times higher incidence of death from sepsis or renal failure than adult-onset SLE.[4–7] Although severe hematologic, renal, cardiac, and neurologic manifestations at presentation of cSLE have been well documented, there is limited literature on gastrointestinal (GI) manifestations at presentation in this population; this rarity of data may delay diagnosis and treatment, leading to serious complications.

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GI manifestations in cSLE range from mild symptoms to severe life-threatening complications including lupus enteropathy, aortitis, bowel ischemia, intestinal perforation, and necrotizing or hemorrhagic pancreatitis. The pathophysiology of these complications is suspected to be related to small vessel vasculitis or vasculopathy, but large vessel vasculitis has also been described. Although approximately up to one-third of individuals develop GI symptoms at one point in their cSLE course, GI complaints are rarely the initial presenting symptom. Previous cohort studies and reviews have estimated that GI manifestations are the presenting symptom in 5–19% of cSLE cases.[8–12] These cases prove to be challenging diagnostic dilemmas given the extensive number of pediatric conditions that present with non-specific GI complaints and the limited literature describing the prevalence and manifestations of severe GI symptoms at presentation in cSLE.

Methods

With approval from our Institutional Review Board, a retrospective chart review of patients with a diagnosis of SLE as per SLICC 2012 and EULAR/ACR 2019 classification criteria whose disease presentation included severe GI disease was completed. Patient characteristics, clinical course, and outcomes were abstracted. A thorough review of the literature using Pubmed for case reviews or case series of cSLE with significant GI features at diagnosis was performed with the collected articles critically reviewed. Articles were appraised and included in the final analysis if the following criteria were met: (1) new diagnosis cSLE before age 18 and (2) severe GI complications at the time of diagnosis. Severe GI disease was defined as either life-threatening or impacting treatment decisions for induction immunomodulation therapy.

Our initial search identified 89 articles which were screened to ensure they met our inclusion criteria. Of the 89 initial articles, 18 met criteria for our case series. Articles or patients were excluded if patients were over the age of 18 years at the time of SLE diagnosis, GI symptoms were not present at initial presentation, GI symptoms were deemed to not be severe, or if there was insufficient information provided about SLE diagnosis or GI course.

Results

Institutional cohort

Out of a total of 714 children diagnosed with SLE from 2000 to 2023 at our institution, six patients (0.84%) presented with severe GI manifestations. Presenting features and induction treatment for each of the cases are described below. Table 1 summarizes patient characteristics, clinical course, and outcomes.

Protein-losing enteropathy

Patient 1 is a 6-year-old male with a history of progressively worsening non-painful abdominal distension for 2 years who presented with 1 month of acute-on-chronic abdominal distension associated with pain, emesis, diarrhea and fever. On admission, initial exam revealed anasarca with associated periarticular swelling, and initial evaluation was notable for profound hypoalbuminemia 1.1 g/dL, elevated stool alpha-1 antitrypsin >1.33 mg/g (normal <0.62 mg/g), and computed tomography of abdomen and pelvis (CTAP) with bilateral pleural effusions and significant ascites. Additionally, he was found to have hypocomplementemia with positive ANA, anti-dsDNA and anti-RNP. His urinalysis showed elevated protein and microscopic hematuria with kidney biopsy demonstrating Class IV/V lupus nephritis.

Given disease severity, he was treated with intravenous and oral corticosteroids and monthly cyclophosphamide infusions for induction. He required prolonged bowel rest and parenteral nutrition for 3 months along with frequent albumin replacement. His initial hospital course was further complicated by microscopic GI bleed (fecal occult blood test was positive although stool was never grossly bloody), and endoscopic biopsies ultimately revealed crypt abscesses in addition to CMV positivity. The patient was treated with a 14-day course of ganciclovir for CMV colitis. After discharge, his abdominal distension and hypoalbuminemia resolved after 5 months of continued immunomodulatory therapy.

Thoracoabdominal aortitis

Patient 2 is a 15-year-old female admitted with 1 week of emesis, diarrhea, and abdominal pain. Initial evaluation was remarkable for acute kidney injury (Cr 1.42 mg/dL) responsive to intravenous hydration. CTAP demonstrated diffuse small bowel wall thickening, stranding of mesentery, and small intraperitoneal ascites with trace pericardial effusion which was attributed to infectious inflammatory response. She received supportive care and was discharged after 24 hours with improved symptoms. Two weeks later, she was readmitted due to recurrence and worsening of her GI symptoms. A repeat CTAP at this time showed worsening small bowel wall edema, extensive hyperemia, mesenteric fat stranding, ascites, and anasarca with pericardial and pleural effusions. Further evaluation revealed hypocomplementemia, autoimmune hemolytic anemia, leukopenia, positive ANA and autoantibodies (dsDNA, SSA Ro52/60, SSB and anti-cardiolipin IgM). Magnetic resonance angiography (MRA) of her abdomen demonstrated diffuse thoracoabdominal aortitis without visceral artery stenosis, diffuse enterocolitis with extensive colonic submucosal edema, and persistent third-spacing.

Given significant large vessel vasculitis, her induction therapy consisted of both intravenous and oral corticosteroids

Table 1. Clinical summary of cSLE patients with severe GI manifestations (institutional cohort).

Age and sex at presentation (years)	Duration of illness prior to diagnosis	Duration of GI symptoms prior to diagnosis	SLICC criteria	SLEDAI at presentation	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
6 M	2 years	2 years	5/17: serositis, renal (Class IV/V lupus nephritis), ANA, dsDNA, low C3 and C4	13 points: fever, proteinuria, microscopic hematuria, low complement, elevated dsDNA ab	Protein-losing enteropathy (chronic abdominal distension, abdominal pain, vomiting, severe diarrhea)	Albumin 1.1 g/dL; stool alpha-1 antitrypsin >1.33 mg/g (normal 0-0.62 mg/g); abdominal CT: ascites; endoscopy: diffuse edematous boggy colonic mucosa and then with cryptogenic/crypt abscesses (1-2 ileal cells + CMV)	Corticosteroids, IV cyclophosphamide, mycophenolate mofetil	5 months (still with intermittent diarrhea, improved)	Remission on mycophenolate mofetil, flare of GI and renal disease 3 years after diagnosis while off treatment requiring reinitiation of immunomodulatory treatment
							Other: bowel rest and parenteral nutrition (3 months), albumin replacement, IVIG replacement, ganciclovir for CMV colitis		Duration of follow-up: 2 years
15 F	3-4 weeks	3-4 weeks	7/17: serositis, hemolytic anemia, leukopenia, ANA, dsDNA, low C3 and C4, anticardiolipin ab	9 points: rash, pericarditis, low complement, elevated dsDNA ab, leukopenia	Thoracoabdominal aortitis and mesenteric vasculitis (abdominal pain, nausea, vomiting, diarrhea, ascites)	Abdominal US: bowel wall edema; abdominal CT: worsening bowel wall edema from stomach to small bowel, hyperemia/ mesenteric fat stranding, worsening anasarca/ascites; MRA abdomen: diffuse thoracoabdominal aortitis without stenosis, diffuse enterocolitis with extensive colonic submucosal edema and evidence of third spacing	Corticosteroids, IV cyclophosphamide, rituximab, hydroxychloroquine, mycophenolate mofetil	2 months	Remission on hydroxyquloroquine (MRA abdomen changes resolved 1.3 years after diagnosis), no flare
							Other: bowel rest and parenteral nutrition (2 weeks), diuretics		Duration of follow-up: 5 years

(continued)

Table 1. (continued)

Age and sex at presentation (years)	Duration of illness prior to diagnosis	Duration of GI symptoms prior to diagnosis	SLICC criteria	SLEDAI at presentation	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
12 F	4-5 weeks	2-3 weeks	9/17: oral ulcers, serositis, renal (Class IV lupus nephritis), ANA, dsDNA, Sm, anticardiolipin ab, low complement, DAT	22 points: vasculitis, hematuria, proteinuria, oral ulcers, low complement, elevated dsDNA ab	Necrotizing, hemorrhagic pancreatitis complicated by pseudocyst (abdominal pain)	Lipase (max) > 10,000 U/L (normal 15-110 U/L); abdominal MRI: severe necrotizing pancreatitis, large volume ascites, bilateral pleural effusions, and diffuse body wall edema, thickened duodenum and submucosal edema likely reactive to pancreatitis; MRCP: redemonstration of necrotizing pancreatitis with interval development of a 17.7 cm acute necrotizing collection; RUQ US: newly identified discrete fluid within the pancreatic body/tail which measures about 3.1 × 1.3 cm transaxial, separate from the pancreas, there is a hypoechoic well-circumscribed ovoid lesion without internal vascularity which measures about 3.0 × 1.7 × 2.2 cm, favored evolving walled off pancreatic necrosis or pseudocyst	Corticosteroids, rituximab, IV cyclophosphamide, mycophenolate mofetil Other: bowel rest and parenteral nutrition (4 weeks)	N/A	Active symptoms (abdominal pain) from sequalae of necrotizing pancreatitis including chronic pancreatitis with pseudocyst and development of diabetes mellites requiring insulin treatment Duration of follow-up: 7 months

(continued)

Table 1. (continued)

Age and sex at presentation (years)	Duration of illness prior to diagnosis	Duration of GI symptoms prior to diagnosis	SLICC criteria	SLEDAI at presentation	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
10 F	3 months	2 months	6/17: acute cutaneous, oral ulcers, serositis, neurologic, ANA, low C3 and C4	30 points: psychosis, stroke, vasculitis, hematuria, low complement	Necrotizing, hemorrhagic pancreatitis complicated by pseudocyst (abdominal pain, severe ascites leading to competitive abdomen and respiratory failure)	Lipase (max) 8032 U/L (normal 15-110 U/L); Stool elastase: 49 mcg/g (<100 mcg/g suggestive of severe pancreatic insufficiency); abdominal CT: edematous pancreas, large amount peripancreatic fluid, large volume ascites; RUQ US: suggestion of non-occlusive portal vein thrombosis. findings compatible with known pancreatitis; abdominal CT (repeat): extensive thin-walled encapsulated peripancreatic fluid collection 20.6 × 5.4 × 8.4 cm with extension into the perihepatic, perisplenic, and bilateral paracolic spaces; MRI MRCP: decreased in size of the walled off necrotic collection in the pancreatic bed, which now measures 9.1 cm in maximal diameter, pancreatic atrophy, consistent with sequela from necrotizing pancreatitis	Corticosteroids, rituximab, anakinra for refractory serositis, cyclophosphamide, hydroxychloroquine, mycophenolate mofetil	13 months	Remission on mycophenolic acid and hydroxychloroquine, no flares

(continued)

Table 1. (continued)

Age and sex at presentation (years)	Duration of illness prior to diagnosis	Duration of GI symptoms prior to diagnosis	SLICC criteria	SLEDAI at presentation	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
							Other: bowel rest and parenteral nutrition (2 weeks), transluminal stent placement for pancreatic pseudocyst (removed after 2 months)		Duration of follow-up: 26 months
15 F	2 days	2 days	9/17: serositis, neurologic, renal (Class III lupus nephritis), ANA, dsDNA, Sm, low C3 and C4, anticardiolipin and LAC, DAT	22 points: stroke, hematuria, proteinuria, pleuritic chest pain, low complement, elevated dsDNA ab	Enteritis (abdominal pain, nausea, vomiting, diarrhea, dysphagia, ascites)	Abdominal US: bowel wall edema; CT abdomen: significant bowel thickening, particularly in the duodenum and jejunum, predominantly small bowel hyperenhancement, moderate amount of ascites; abdominal US: with significant dilation of common bile duct	Corticosteroids, rituximab, IV cyclophosphamide, hydroxychloroquine, mycophenolate mofetil	1 month	Remission on mycophenolic acid and hydroxychloroquine, no flares
							Other: Bowel rest and parenteral nutrition (3 weeks), anticoagulation, albumin replacement		Duration of follow-up: 16 months
16 F	1-2 months	1-2 weeks	11/17: joint disease, serositis, neurologic, thrombocytopenia, hemolytic anemia, renal (Class I lupus nephritis), ANA, dsDNA, Sm, anticardiolipin ab, low complement	10 points: fever, arthritis, low complement, elevated dsDNA ab, platelets	Intestinal pseudoobstruction with bowel perforation (abdominal pain, nausea, vomiting, re-feeding syndrome)	Abdominal CT: bowel wall thickening throughout entirety of jejunum/ileum w/ mechanical SBO/ ischemia; worsened anasarca: ascites, pleural effusions, body wall edema & pericardial effusion; retroperitoneal LAD (reactive to pyelonephritis)	Corticosteroids, IV cyclophosphamide, hydroxychloroquine, mycophenolic acid	5 months	Remission on mycophenolic acid and hydroxychloroquine, no flares
						Histopathology: small intestine: small bowel segment with patchy surface epithelial necrosis, marked submucosal edema and congestion, focal necrosis and inflammation of muscularis propria, and moderate serositis	Other: small bowel resection (30 cm of jejunum), bowel rest and parenteral nutrition (3 weeks), anticoagulation		Duration of follow-up: 12 months

IV = intravenous; CT = computed tomography; US = ultrasound; MRA = magnetic resonance angiography; MRI = magnetic resonance imaging; MRCP = magnetic resonance cholangiopancreatography; RUQ = right upper quadrant; EGD = esophagogastroduodenoscopy; ANA = antinuclear antibody; dsDNA = double strand deoxyribonucleic acid; Sm = smith; ab = antibodies; DAT = direct antiglobulin test; LAC = lupus anticoagulant; CSF: cerebrospinal fluid; CMV = cytomegalovirus; SBO = small bowel obstruction; LAD = lymphadenopathy; PLE = protein losing enteropathy; IVIG = intravenous immunoglobulin; PLEX = plasma exchange; ASA = aspirin; N/A = not available.

with rituximab and monthly cyclophosphamide. In addition, she required prolonged bowel rest and parenteral nutrition for 2 weeks. She had clinical remission of GI symptoms after 2 months of induction treatment. She was later transitioned to maintenance therapy with hydroxychloroquine and mycophenolate after completing a 6 month course of cyclophosphamide. MRA changes resolved after 1 year post-diagnosis. She is currently on hydroxychloroquine monotherapy without any reported flares of GI disease.

Severe pancreatitis

Patient 3 is a 12-year-old female with a history of gastritis (resolved and off medications) who presented to the hospital with 2 weeks of progressive, severe abdominal pain without weight loss, nausea, vomiting, or diarrhea. She also endorsed several weeks of fatigue, painful oral ulcers, left knee pain with swelling and a tender red rash on her palms. On admission, she was found to have evidence of severe necrotizing pancreatitis on magnetic resonance imaging (MRI) of the abdomen with lipase peak >10,000 U/L. Other imaging findings included large ascites, bilateral pleural effusions and diffuse body wall edema. She became critically ill with respiratory failure, pulmonary hemorrhage, and *Capnocytophagia* septicemia. She was found to have positive ANA and autoantibodies (DAT, anti-dsDNA, anti-Smith, anti-RNP, anti-SSA-Ro60, anti-Scl-70, anti-cardiolipin and anti-MPO) as well as hematuria and proteinuria with kidney biopsy confirming Class IV lupus nephritis.

Patient's induction treatment included intravenous and oral corticosteroids, rituximab, and intravenous cyclophosphamide. She was placed on bowel rest with parenteral nutrition for approximately 4 weeks. Her course was further complicated by large pseudocyst measuring 17.7 × 6.4 × 10.7 cm that was managed conservatively with close monitoring. Genetic testing was notable for a heterozygous pathogenic mutation in cystic fibrosis transmembrane conductance regulator (CFTR). Clinical remission was achieved, and patient was discharged after a 2 month hospitalization. Maintenance therapy with mycophenolate mofetil was then initiated, and she also continued on hydroxychloroquine. Six months after discharge, she was re-admitted for 2 days due to abdominal pain. Lipase was normal with CTAP showing improvement of pseudocyst to 6.1 × 3.1 × 4.8 cm. An incidental finding of Nutcracker syndrome was reported from CTAP. She continues to have mild, intermittent abdominal pain and nausea attributed to her chronic pancreatitis.

Patient 4 is a 10-year-old female with a similar course. She presented to an outside hospital for 3 months of weight loss, fever, fatigue, malar rash, oral ulcers, and arthralgia and was ultimately diagnosed with SLE given hypocomplementemia, hemolytic anemia, leukopenia, and +ANA. She had improvement with intravenous and oral corticosteroids, hydroxychloroquine, and azathioprine, however, was readmitted 4 weeks later for severe abdominal pain. She required intubation due to respiratory failure secondary to competitive abdomen and was transferred to our institution for further care. Evaluation revealed necrotizing, hemorrhagic pancreatitis with anasarca and ascites with peak lipase 8032 U/L (normal 15-110 U/L). Her hospital course was complicated by neuropsychiatric lupus, non-occlusive portal vein thrombosis, and development of a large pancreatic pseudocyst measuring 20.6 × 5.4 × 8.4 cm (Figure 1).

Induction therapy included intravenous and oral corticosteroids, anakinra for severe serositis, rituximab, and monthly intravenous cyclophosphamide. She required bowel rest and parenteral nutrition for 2 weeks. Her pancreatic pseudocyst persisted for up to a year after initial diagnosis, and an Axios stent was placed to allow for a

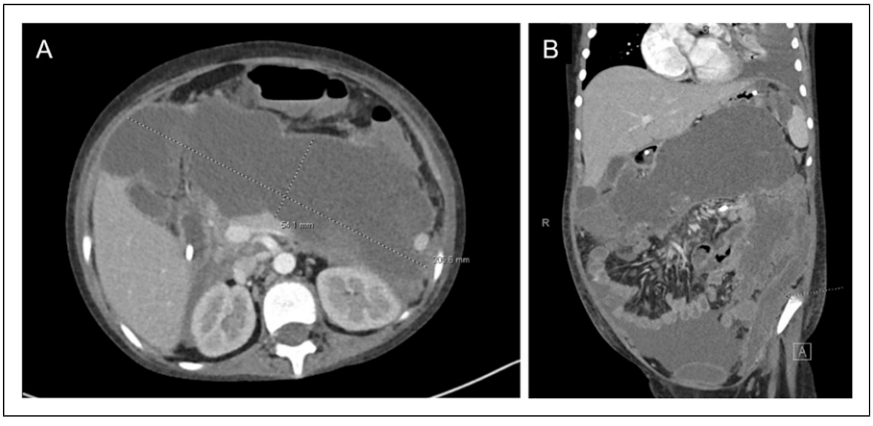

Figure 1. CT abdomen and pelvis. (A) Axial and (B) coronal views demonstrating large pancreatic pseudocyst (20.6 × 5.4 × 8.4 cm).

conduit from the pseudocyst to her GI tract. This stent was removed 2 months later without recurrence of pancreatitis or pseudocyst. She is currently in remission on hydroxychloroquine and mycophenolate as maintenance therapy without disease flare.

Lupus enteritis or mesenteric vasculitis

Patient 5 is a 14-year-old female who presented with 2 days of acute, severe, and diffuse abdominal pain with associated nausea, vomiting, and dysphagia. CTAP revealed significant bowel thickening, particularly affecting the duodenum and jejunum, and a moderate amount of ascites consistent with enteritis (Figure 2). Further evaluation revealed that she had hypocomplementemia with positive ANA and autoantibodies (DAT, anti-dsDNA, anti-Smith, and anti-phospholipid). Antibodies to RNP, SSA, Scl-70 as well as myeloperoxidase (MPO) were also positive. Patient had microscopic hematuria with significant proteinuria. Kidney biopsy later revealed Class III lupus nephritis. Her clinical course was further complicated by micro-ischemic strokes.

Due to her severity of disease, induction therapy was started with both intravenous and oral corticosteroids, rituximab, and monthly intravenous cyclophosphamide. She was placed on bowel rest and parenteral nutrition for 3 weeks with slow transition to enteral nutrition. Early advancement of diet was complicated by hematochezia. Bowel ultrasounds were used serially to assess for bowel wall edema and enhancement while enteral nutrition was advanced. She had complete resolution of her GI symptoms and imaging findings 1 month post-treatment. She remains on hydroxychloroquine and mycophenolate as maintenance with sustained clinical remission.

Intestinal pseudo-obstruction

Patient 6 is a 16-year-old female who developed 2 weeks of progressively worsening abdominal pain, emesis, and presenting in compensated shock. Her disease features were initially attributed to multi-drug resistant *E. coli* pyelonephritis. CTAP revealed pyelonephritis as well as areas of focal proximal small bowel thickening consistent with enteritis. Laboratory findings were notable for severe anemia and thrombocytopenia. She was placed on bowel rest and started on antibiotics with temporary improvement in symptoms. Following advancement of her diet, her abdominal pain and vomiting recurred with increased severity. Repeat CTAP revealed increased bowel wall thickening throughout entirety of jejunum and ileum with evidence of mechanical small bowel obstruction and ischemia. Imaging also showed ascites, pleural effusions, body wall edema, and a pericardial effusion (Figure 3). She underwent multiple exploratory laparotomies resulting in resection of 33 cm of jejunum due to ischemic injury (Figure 4).

Further review of the history revealed pallor, chronic arthralgia, behavioral and cognitive symptoms concerning for psychiatric manifestations. Additional testing verified diagnosis of cSLE, with proteinuria, microscopic hematuria, hypocomplementemia, ANA positivity and positive autoantibodies (anti-dsDNA, anti-Smith, and anti-phospholipid). Kidney biopsy showed Class I lupus nephritis. A diagnosis of neuropsychiatric lupus was made with imaging findings of micro-ischemic strokes and cerebrospinal fluid abnormalities.

Induction immunotherapy included intravenous and oral corticosteroid along with monthly intravenous cyclophosphamide. Bowel rest and parenteral nutrition was started for a total of 3 weeks. Early advancement of diet resulted in recurrent abdominal pain and diarrhea. Complete remission

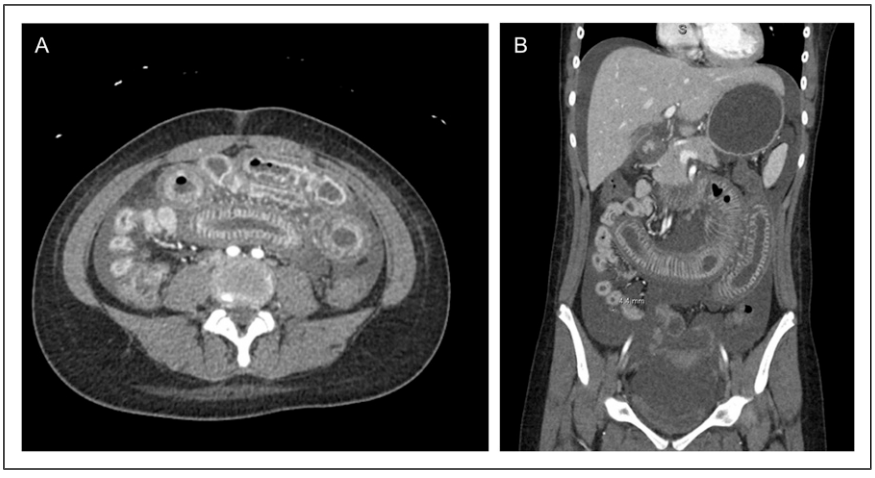

Figure 2. CT abdomen and pelvis. (A) Axial and (B) coronal views demonstrating extensive bowel wall thickening and edema primarily involving the proximal small bowel.

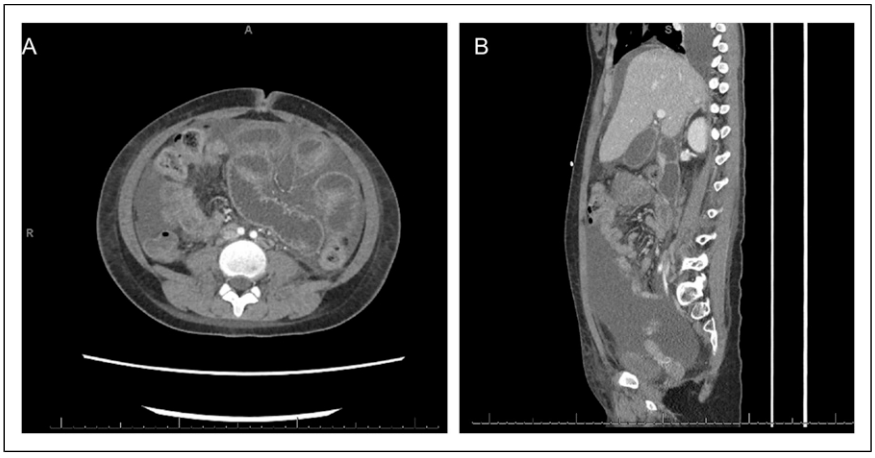

Figure 3. CT of Abdomen and Pelvis. (A) Axial and (B) sagittal views demonstrating severe bowel wall thickening throughout entirety of jejunum and ileum with a transition to decompressed right lower quadrant small bowel, demonstrative of mechanical small bowel obstruction with bowel wall thickening suggestive of ischemia.

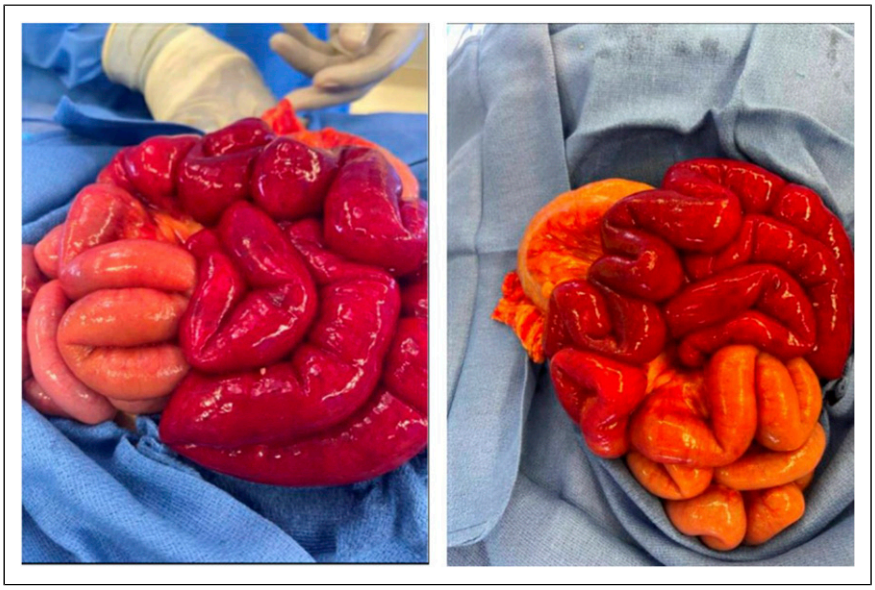

Figure 4. Stark contrast in ischemic versus perfused bowel during initial exploratory laparotomy where 33 cm of jejunum was resected.

was achieved after 5 months of therapy. She remains in remission with hydroxychloroquine and mycophenolic acid as her maintenance agents.

In summary, our institutional cohort was composed of primarily female patients (83%, 5/6) who presented at an average age of 12 years. Within our cohort, the most reported symptoms prior to cSLE diagnosis were abdominal pain and emesis (83%) while diarrhea was seen in half the patients. The duration of GI symptoms prior to cSLE diagnosis ranged from 2 days to 2 years. Surgical intervention was seen in two patients including Axios stent placement and exploratory laparotomy with bowel resection.

Literature review cohort

Our literature search identified 18 published articles with a total of 25 cases of cSLE in which severe GI symptoms where the predominant chief complaint. Table 2[13–32] summarizes patient characteristics, clinical course, and outcomes. Female patients made up 88% (22/25) of reported cases with an average age of 14 years at time of presentation. Abdominal pain (84%, 21/25) and emesis (56%, 14/25) were the most prevalent symptoms reported, followed by diarrhea (36%, 9/25) and weight loss (28%, 7/25). Approximately 24% (6/25) of patients underwent surgical

Table 2. Clinical summary of cSLE cases with severe GI manifestation reported in the literature (Literature Review Cohort).

Author of case series (year published)	Author of case (year published)	Age and sex at presentation (years)	Duration of GI symptoms prior to diagnosis	SLICC criteria	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
Jin et al. (2015)[13]	Munyard et al. (1997)[14]	15 F	Appx. 4 weeks	4/17: serositis, lymphopenia, thrombocytopenia, dsDNA	Intestinal pseudo-obstruction (abdominal pain, distension, vomiting, diarrhea, weight loss)	Abdominal ultrasound: fluid-filled loops of small bowel and feces in the large bowel; abdominal x-ray: pronounced dilation of small and large bowel	Corticosteroids, hydroxychloroquine	2 months	Remission, no flares
							Other: parenteral nutrition		Duration of follow-up: N/A
Jin et al. (2015)[13]	Mok et al. (2000)[15]	18 F	N/A	4/17: thrombocytopenia, ANA, dsDNA, low C3 and C4	Intestinal pseudo-obstruction	Abdominal CT: circumferential thickening of the small and large bowel and large amount of ascites; Sigmoidoscopy: mucosal edema and erythema	Corticosteroids, azathioprine	N/A	Remission, flare 8 months after diagnosis
									Duration of follow-up: N/A
Jin et al. (2015)[13]	Yamazaki-Nakashimada et al. (2009)[16]	10 M	1 week	6/17: serositis, neurologic (seizures), renal (proteinuria), ANA, low C3 and C4, anti-β2 glycoprotein and anticardiolipin ab	Intestinal pseudo-obstruction (abdominal pain, constipation, fecaloid vomiting)	Abdominal x-ray: distended intestinal loops with air-fluid levels and no air in the rectum	Corticosteroids, oral cyclophosphamide, IVIG, ASA	N/A	Remission, no flares
						Histopathology: appendix: mural infiltration of eosinophils with few plasma cells and neutrophils; small and large intestine: numerous eosinophils predominantly in the muscularis propria with diverse grades of fibrosis	Other: laparotomy, appendectomy		Duration of follow-up: 2 years

(continued)

Table 2. (continued)

Author of case series (year published)	Author of case (year published)	Age and sex at presentation (years)	Duration of GI symptoms prior to diagnosis	SLICC criteria	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
Jin et al. (2015)[13]	Yamazaki-Nakashimada et al. (2009)[16]	15 F	4 months	5/17: serositis, lymphopenia, renal (Class III lupus nephritis), ANA, Sm	Intestinal pseudo-obstruction (abdominal pain, diarrhea, vomiting, weight loss)	EGD: candidal esophagitis; upper GI series: mechanical obstruction in the lower portion of the duodenum	Corticosteroids, oral cyclophosphamide	N/A (prior to hospital discharge)	Remission, no flares
						Histopathology: Esophagus: ulcer with fibrin and numerous neutrophils and eosinophils, eosinophils of the muscularis propria; Small and large intestine: florid acute peritonitis and mild perivasculitis	Other: appendectomy, laparotomy, parenteral nutrition		Duration of follow-up: 1 year
	Hsu et al. (2020)[17]	7 F	weeks	6/17: serositis, lymphopenia, ANA, Sm, low C3 and C4, anticardiolipin ab	Intestinal pseudo-obstruction (abdominal pain, constipation, anorexia, vomiting, weight loss)	Abdominal x-ray: air fluid levels without mechanical obstruction	Corticosteroids, rituximab, IV cyclophosphamide, mycophenolate mofetil	78 days (prior to hospital discharge)	Remission, no flares
						Abdominal MRI/MRA: large stool burden in the colon and gas paucity	Other: PLEX, parenteral nutrition		Duration of follow-up: N/A
Naddei et al. (2019)[18]	Tsukahara et al. (1980)[19]	12 M	2 weeks	4/17: acute cutaneous, ANA, dsDNA, low C3 and C4	PLE (generalized edema, ascites, weight gain)	Albumin 1.32 g/dL	Corticosteroids	N/A	Flare 9 months after diagnosis, persistent hypocomplementemia
							Diuretics		Duration of follow-up: N/A

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Table 2. (continued)

Author of case series (year published)	Author of case (year published)	Age and sex at presentation (years)	Duration of GI symptoms prior to diagnosis	SLICC criteria	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
Naddei et al. (2019)[18]	Chase et al. (1982)[20]	12 F	2 weeks	4/17: serositis, renal (diffuse proliferative glomerulonephritis), ANA, dsDNA	PLE (periorbital and generalized edema)	Albumin 1.2 g/dL; upper GI series: small intestinal mucosal edema; mesenteric arteriogram and inferior venocavogram: general elevation in venous pressures; 51Cr-labeled albumin tracer: 13.04% GI protein losses in stool Histopathology: liver: minimal, non-specific portal inflammation; ileum: distal tips of villi were swollen and contained enlarged, dilated, prominent lymphatic channels	Corticosteroids, IV cyclophosphamide	N/A	N/A (patient deceased due to septic shock) Duration of follow-up: 4 years
Naddei et al. (2019)[18]	Molina et al. (1996)[21]	13 F	3 months	4/17: joint disease, thrombocytopenia, ANA, low C3 and C4	PLE (generalized edema, abdominal pain, diarrhea)	Albumin 0.7 g/dL; 24h Stool alpha-1 antitrypsin clearance 3125 mg/dL (normal <50 mg/dL); abdominal CT: ascites Histopathology: Small intestine: minimal villous blunting with edema and mild infiltration of lymphoid follicles and eosinophils, no evidence of vasculitis	Corticosteroids, azathioprine	N/A	Remission, no flares Duration of follow-up: N/A

(continued)

Table 2. (continued)

Author of case series (year published)	Author of case (year published)	Age and sex at presentation (years)	Duration of GI symptoms prior to diagnosis	SLICC criteria	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
Naddei et al. (2019)[18]	Park et al. (2004)[22]	11 F	4 months	9/17: oral ulcers, joint disease, serositis, thrombocytopenia, renal (Class IV lupus nephritis), ANA, dsDNA, low C3 and C4, anticardiolipin ab	PLE (periorbital edema, abdominal pain, diarrhea, weight loss)	Albumin 2.9 g/dL; 24h stool alpha-1 antitrypsin clearance 315 mg/dL (normal ≤54 mg/dL), 99Tc-human serum albumin scintigraphy: protein loss from distal ileum Histopathology: duodenum: mild, chronic inflammation	Corticosteroids, IV cyclophosphamide	5 months	Remission, no flares Duration of follow-up: 5 months
Naddei et al. (2019)[18]	Ranawaka et al. (2012)[23]	13 F	3 years	6/17: acute cutaneous, neurologic (multiple infarcts on brain MRI, pleocytosis of CSF), leukopenia, thrombocytopenia, ANA, low C3 and C4	PLE (periorbital and generalized edema, diarrhea)	Albumin 1.8 g/dL; EGD and colonoscopy: normal	Corticosteroids, azathioprine, mycophenolate mofetil	N/A	Remission, no flares Duration of follow-up: 3 months
Naddei et al. (2019)[18]	Sansinanea et al. (2016)[24]	17 F	3 years	8/17: acute cutaneous, oral ulcers, joint disease, serositis, thrombocytopenia, ANA, low C3 and C4, anticardiolipin ab and lupus anticoagulant	PLE (abdominal pain, diarrhea, ascites)	Albumin 1.7 g/dL; stool alpha-1 antitrypsin clearance 1040 mL/24h (normal <30 mL/24h), EGD: marked edema in the second segment of duodenum; colonoscopy: normal Histopathology: Duodenum: chronic nonspecific duodenitis and chorionic edema	Corticosteroids, IV cyclophosphamide, azathioprine, hydroxychloroquine, cyclosporine, mycophenolate mofetil, rituximab Other: parenteral nutrition	57 months	Remission, flares at 24 months, 36 months, and 51 months after diagnosis Duration of follow-up: N/A

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Table 2. (continued)

Author of case series (year published)	Author of case (year published)	Age and sex at presentation (years)	Duration of GI symptoms prior to diagnosis	SLICC criteria	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
	Tu et al., (2009)[25]	15 F	5 days	5/17: neurologic (seizures), renal (proteinuria), ANA, dsDNA ab, low C3 and C4	Mesenteric vasculitis (epigastric pain, vomiting, weight loss)	Abdominal CT: diffuse bowel wall edema with prominent celiac, superior mesenteric, and inferior mesenteric branches	Corticosteroids, IV cyclophosphamide	N/A	Remission, no flares Duration of follow-up: 2.5 years
	Tu et al., (2009)[25]	13 F	N/A	5/17: acute cutaneous, neurologic, ANA, dsDNA, low C3 and C4	Mesenteric vasculitis (abdominal pain, bilious vomiting, diarrhea)	Abdominal x-ray: diffuse bowel distension with an air-fluid level; abdominal CT: bowel wall thickening with enhancement of the small and large bowel, especially on the small mesenteric artery and inferior mesenteric artery	Corticosteroids, IV cyclophosphamide, azathioprine	N/A	Remission, no flares Duration of follow-up: N/A
	Tu et al., (2009)[25]	15 F	N/A	5/17: acute cutaneous, renal (proteinuria), ANA, dsDNA, low C3 and C4	Mesenteric vasculitis (epigastric pain, bilious vomiting)	Abdominal US: ascites and diffuse severe small bowel edema with hyperemic mesentery; abdominal CT: serositis and long-segment small bowel wall thickening	Corticosteroids	N/A	Remission, no flares Duration of follow-up: N/A
	Fotis et al. (2016)[26]	15 F	6-8 weeks	8/17: lymphopenia, hemolytic anemia, thrombocytopenia, renal (Class II lupus nephritis), ANA, dsDNA, low C3 and C4, anticardiolipin ab	Mesenteric vasculitis (abdominal pain, diarrhea, hematochezia, nausea, vomiting, weight loss)	Colonoscopy: normal	Corticosteroids, IV cyclophosphamide, ASA	N/A	Remission, no flares Duration of follow-up: N/A

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Table 2. (continued)

Author of case series (year published)	Author of case (year published)	Age and sex at presentation (years)	Duration of GI symptoms prior to diagnosis	SLICC criteria	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
	Liu et al. (2018)[27]	9 F	1 month	4/17: renal (proteinuria), ANA, dsDNA, low C3 and C4	Mesenteric vasculitis (abdominal pain, vomiting, diarrhea)	Abdominal US: ascites and enlarged mesenteric lymph nodes; abdominal CT: thickening of the intestinal wall; contrast test: decreased motility of upper GI tract; gastroscopy: superficial antral gastritis	Corticosteroids, IV cyclophosphamide	2 months	Remission, readmission 15 days after diagnosis Duration of follow-up: 8 months
	Liu et al. (2018)[27]	14 F	2 months	4/17: leukopenia, renal (proteinuria), ANA, low C3	Mesenteric vasculitis (abdominal pain, vomiting)	Abdominal US: segmental intestinal wall thickening on the left side, including the transverse colon, sigmoid colon and descending colon and ascites; abdominal CT scan: ascites and possible peritonitis, thickening of intestinal wall	Corticosteroids, IV cyclophosphamide	N/A	Remission, no flares Duration of follow-up: N/A
	Liu et al. (2018)[27]	12 F	2 weeks	6/17: hemolytic anemia, neurologic, renal (proteinuria), ANA, dsDNA, low C3 and C4	Mesenteric vasculitis (abdominal pain, vomiting)	Abdominal US: intestinal wall thickening at the upper and middle abdomen with a small amount of ascites and left hydronephrosis; abdominal CT: local enhancement at the intestinal mucosa	Corticosteroids, IV cyclophosphamide	N/A	Remission, no flares Duration of follow-up: 3 months

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Table 2. (continued)

Author of case series (year published)	Author of case (year published)	Age and sex at presentation (years)	Duration of GI symptoms prior to diagnosis	SLICC criteria	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
	Ogbu et al. (2021)[28]	14 F	2 weeks	4/17: serositis, ANA, Sm, low C3 and C4	Mesenteric vasculitis (abdominal pain, nausea, vomiting)	Abdominal US: multiple dilated small bowel loops; abdominal CT: diffuse dilatation of proximal to mid small intestine, gastric, duodenal, colon and rectal wall thickening, mesenteric hypervascularity, mesenteric edema, and extensive ascites	Corticosteroids, mycophenolate mofetil, rituximab	N/A	Remission, flare 24 months after diagnosis
							Other: Laparotomy		Duration of follow-up: 34 months
	Patra et al. (2021)[29]	8 F	3 days	9/17: acute cutaneous, alopecia, oral ulcers, leukopenia, thrombocytopenia, ANA, dsDNA, low C3 and C4, DAT	Enteritis and mesenteric vasculitis (abdominal pain, vomiting, diarrhea, ascites)	Albumin 1.5 g/L (normal >3.5 g/L); abdominal US: associated with diffuse bowel wall thickening; abdominal CT: dilated small bowel and variable degree of mural thickening involving stomach, small bowel, and colorectum; laparotomy: two gangrenous segments of small bowel, contaminated peritoneal fluid, and thickened mesentery with multiple mesenteric lymph nodes	Corticosteroids, azathioprine, hydroxychloroquine	N/A	Flare 3 months after diagnosis (patient deceased 20 days after flare due to bowel perforation and septic shock)
						Histopathology: Bowel: diffuse mucosal necrosis and focal ulceration and deposition of IgG in blood vessel walls on IFA suggestive of immune-complex mediated vasculitis	Other: Laparotomy, bowel resection (100 cm total)		Duration of follow-up: 3-4 months

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Table 2. (continued)

Author of case series (year published)	Author of case (year published)	Age and sex at presentation (years)	Duration of GI symptoms prior to diagnosis	SLICC criteria	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
	Lepore et al. (2022)[30]	10 F	N/A	4/17: renal (glomerulonephritis), ANA, dsDNA ab, low C3 and C4	Mesenteric vasculitis with bowel perforation (abdominal pain)	Abdominal x-ray: air-fluid levels in the bowels	Corticosteroids, IV and oral cyclophosphamide, azathioprine	3 weeks	Remission, no flares
						Histopathology: Small and large intestine: inflammatory edema of the intestinal wall with necrotic areas and extensive ulceration, hemorrhage, and arterial thrombosis	Other: Appendectomy, laparotomy, bowel resection		Duration of follow-up: 17 years
	Lepore et al. (2022)[30]	16 M	48 hours	5/17: acute cutaneous, ANA, dsDNA, low C3 and C4, anticardiolipin ab and lupus anticoagulant	Mesenteric vasculitis (abdominal pain, GI bleeding)	Histopathology: Intestine: edema and hemorrhagic infarction of submucosa and mesentery with diffuse vasculitis involving mainly the venules, small and medium vessels showed mixed inflammatory cell infiltration with polymorphonuclear leukocyte prevalence	Corticosteroids, IV cyclophosphamide	N/A	Remission, no flares
							Other: Appendectomy, laparotomy, bowel resection		Duration of follow-up: 3 months
	Ramanan et al. (2002)[31]	14 F	4 weeks	12/17: acute cutaneous, oral ulcers, joint disease, serositis, renal, leukopenia, hemolytic anemia, thrombocytopenia, ANA, dsDNA, low C3 and C4, anticardiolipin ab	Acute necrotizing pancreatitis and mesenteric vasculitis (abdominal pain, vomiting, diarrhea, weight loss)	Laparoscopy: pancreatitis with free peritoneal fluid and multiple areas of fat necrosis; laparotomy: two jejunal perforations	Corticosteroids	N/A	N/A (patient deceased due to multiorgan failure and infection)

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Table 2. (continued)

Author of case series (year published)	Author of case (year published)	Age and sex at presentation (years)	Duration of GI symptoms prior to diagnosis	SLICC criteria	GI manifestations at diagnosis	Pertinent GI studies and histopathology	Immunomodulatory and other treatment	Time to remission of GI manifestations	Outcomes of GI manifestations at last follow-up
						Histopathology: Jejunum: severe necrotizing vasculitis	Other: PLEX		Duration of follow-up: 1 month
	El Qadiry et al. (2018)[32]	14 F	5 days	7/17: acute cutaneous, joint disease, serositis, neurologic, renal, thrombocytopenia, Sm, low C3 and C4	Acute necrotizing pancreatitis (abdominal pain, ascites)	Lipase 810 IU/L; abdominal US: edematous pancreas and intrapancreatic necrosis	Corticosteroids, IV cyclophosphamide	N/A	N/A (patient deceased due to multiorgan failure)
									Duration of follow-up: 20 days
	El Qadiry et al. (2018)[32]	14 F	15 days	10/17: acute cutaneous, joint disease, serositis, neurologic, renal, thrombocytopenia, ANA, dsDNA, Sm, low C3	Acute necrotizing pancreatitis (abdominal pain, ascites)	Lipase 610 IU/L; abdominal CT: swollen form of pancreas	Corticosteroids, IV cyclophosphamide	N/A	N/A (patient deceased due to septic shock)
									Duration of follow-up: 1 month

IV = intravenous; CT = computed tomography; US = ultrasound; MRA = magnetic resonance angiography; MRI = magnetic resonance imaging; MRCP = magnetic resonance cholangiopancreatography; RUQ = right upper quadrant; EGD = esophagogastroduodenoscopy; ANA = antinuclear antibody; dsDNA = double strand deoxyribonucleic acid; Sm = smith; ab = antibodies; DAT = direct antiglobulin test; LAC = lupus anticoagulant; CSF: cerebrospinal fluid; CMV = cytomegalovirus; SBO = small bowel obstruction; LAD = lymphadenopathy; PLE = protein losing enteropathy; IVIG = intravenous immunoglobulin; PLEX = plasma exchange; ASA = aspirin; N/A = not available.

intervention including appendectomy in 4 patients, emergent exploratory laparotomies in 6 patients, and surgical resection of necrotic or perforated bowel in 3 patients. The duration of GI symptoms prior to cSLE diagnosis in this cohort ranged from 3 days to 3 years.

Discussion

Several cohort studies and reviews have explored GI manifestations in cSLE, both at the initial presentation and throughout the clinical course. Although literature within the pediatric population is limited, this research suggests 19%–27.5% of patients with cSLE develop GI system involvement overall.[9,10] Of these patients with GI involvement, between one- to two-thirds experience these symptoms at their initial presentation.[9–11] Most studies report abdominal pain as the most common GI symptom,[8,9,11] but specific data on the frequency and outcomes of severe GI manifestations in children with SLE is lacking. In a French multicenter cohort study describing the initial presentations of 155 patients with cSLE, Bader-Meunier et al. reported 8 severe GI events including episodes of pancreatitis and intestinal myositis with ileus.[8] Additionally, in a retrospective case–control study comparing cSLE patients with and without mesenteric vasculitis, Zhu et al. found that cSLE patients with mesenteric vasculitis had high SLEDAI-2 K scores at onset.[33] These children were also more likely to have damage to other organs including kidneys, serous cavity, and lungs and more likely to receive intensive immunomodulatory therapy with cyclophosphamide when compared to cSLE patients without mesenteric vasculitis.[33] Most of the patients with mesenteric vasculitis in this cohort achieved remission of their GI symptoms within 1 month of treatment with low relapse rates, however, their overall recovery was worse compared to cSLE patients without mesenteric vasculitis.[33]

At our institution, 6 cases of the 714 total cases of cSLE (0.84%) evaluated over 23 years presented with primary GI symptoms with severe complications. Through our literature search, we identified an additional 25 patients who presented similarly. Consistent with the literature, females made up most of our combined sample size with a female to male ratio of 6.75:1 and mean age of presentation of 13 years.[1] The most common symptom at time of presentation in both groups were abdominal pain, emesis, or diarrhea. Abdominal complaints are a common occurrence in pediatric patients with a reported total of 4.2 million encounters for abdominal pain across the country from 2004 to 2011 with 2 million of those being at emergency centers (EC).[34] While abdominal pain, emesis, and diarrhea in conjunction with additional finding such as dysuria, right lower quadrant pain, peritonitis or guarding help guide the diagnostic work up, alone they are non-specific symptoms that are more often attributable to other causes including viral processes or functional GI disorders. Unfortunately, misdiagnosis of abdominal complaints is not a rare occurrence as several studies have reported such cases. Naiditch et al. found that of the 816 pediatric patients who underwent appendectomy from 2007 to 2010, 4.8% of them were initially misdiagnosed with viral gastroenteritis or constipation, while Reynold et al. report that 39% of patients seen at their EC were diagnosed with "abdominal pain" and discharged home without a clear etiology.[35,36] Patients within our institutional cohort were diagnosed with acute gastroenteritis, appendicitis, hypothyroidism, and pyelonephritis prior to cSLE diagnosis contributing to delays in diagnosis. Time to diagnosis ranged from 2 days to 2 years in our institutional cohort, and 3 days to 3 years in cases identified from our literature search.

We also found that a significant portion of our combined cohort (26%, 8/31) underwent surgical intervention. Of these cases, 7 patients underwent emergent exploratory laparotomies with 4 requiring resections of bowel due to bowel necrosis or perforation, 4 undergoing appendectomy, and 1 requiring transluminal stent placement for a large pancreatic pseudocyst. Surgical interventions were pursued primarily due to concerns for acute abdomen as determined by physical exam. In a study completed by Shoeler et al., they found 1% of patients presenting to their EC with an abdominal complaint underwent a surgical procedure.[37] In comparison, patients within our cohort underwent surgical procedures at a much higher rate due to severity of their symptoms without a clear etiology. The severity of these presentations highlights the importance of making prompt diagnosis to expedite treatment of cSLE to prevent such invasive interventions and severe complications.

Although GI complaints were the most prominent symptom for these patients at the time of diagnosis, all patients had other features highly suspicious for cSLE including mucocutaneous, joint, cardiopulmonary, and hematologic manifestations. In our cohort, 62% of patients had renal and/or neurologic manifestations, two major organs which indicate a severe disease process, and this was similarly noted in the published cases (60%, 15/25). Besides typical autoantibodies associated with cSLE including anti-dsDNA and anti-Sm antibodies, our institutional cohort of patients exhibited various other autoantibodies including anti-SSA Ro52/60 (67%), anti-SSB (33%), anti-RNP (67%), anti-Scl70 (33%), and anti-MPO (33%) antibodies. Many of these patients required significant immunosuppression as induction therapy for cSLE including agents such as cyclophosphamide and rituximab in addition to corticosteroids. All patients in our series received supportive therapies including prolonged bowel rest and parenteral nutrition to prevent bowel ischemia and perforation. One patient with chronic pancreatitis complicated by pseudocyst required transluminal stent placement which

highlights the importance of multi-specialty care to limit morbidity and preserve a high quality of life for these children.

Lastly, all patients at our institution, besides one (Patient 3) who has shortest length of follow-up, achieved complete remission of cSLE (SLEDAI-2K of 0) and complete resolution of GI manifestations between one to thirteen months after initial diagnosis. Patient 1 had a flare of SLE while he was off all medications requiring re-initiation of immunomodulatory treatment. Long-term outcomes in the published cases were often not well defined or discussed in case reports, however available data shows 15 out of 25 patients had remission without flares (60%), 6 had at least one documented flare or readmission to the hospital for GI symptoms (24%), and 4 passed away (16%).

Conclusions

Childhood-onset SLE is a complex autoimmune condition characterized by immune dysregulation with a wide array of clinical symptoms. Diagnosis of cSLE can be challenging and even more so when patients present atypically with predominant GI symptoms leading to delays in diagnosis and treatment. This delay results in possibly preventable progression of disease burden and severe clinical complications. For this reason, it is important that providers maintain a broad differential that considers cSLE when facing GI symptoms in pediatric patients and thoroughly assess for other signs or laboratory abnormalities which may suggest a systemic process including renal or hematologic manifestations. Patients with cSLE and severe GI manifestations also benefit from multi-specialty care with close collaboration between rheumatologists and gastroenterologists.

Author contributions

Drs. James Orozco, Jessica Nguyen, Sara M. M. Yasrebi, Marnie Blalock, and Sara L. Grisales contributed to the conception and design of the report, reviewed the course data, performed literature review and appraisal, drafted the initial manuscript, and critically reviewed and revised the manuscript. Drs. Craig L. Jensen and Marietta M. DeGuzman contributed to the conception and design of the report and critically reviewed and revised the manuscript. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

Declaration of conflicting interests

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Ethical statement

Ethical approval

This study received ethical approval from the Baylor College of Medicine IRB (H-37047). This is an IRB-approved retrospective study, all patient information was de-identified and patient consent was not required. Patient data will not be shared with third parties.

Informed consent

Requirement for informed consent to participate has been waived by the Baylor College of Medicine IRB. The Baylor College of Medicine IRB waived the need for ethics approval and patient consent for the collection, analysis and publication of the retrospectively obtained and anonymised data for this non-interventional study.

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Data Availability Statement

All authors confirm that the data supporting the findings of this study are available within the article.

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