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Management of Acute Gastrointestinal Complications in a Paediatric Patient with Overlapping Systemic Lupus Erythematosus, Scleroderma, and Polymyositis Overlap

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Abstract: ***Gastrointestinal complications in Systemic Lupus Erythematosus (SLE) are relatively common and can range from mild symptoms like nausea and abdominal pain to severe conditions such as bowel infarction, driven by the disease's systemic inflammatory effects and associated treatment regimens. This case study outlines the management of acute gastrointestinal complications in a 15-year-old female with overlapping SLE, scleroderma, and polymyositis. She presented with severe abdominal symptoms, treated initially with fluids, antibiotics, and stool disimpaction. Persistent issues led to a diagnosis of colonic pseudo-obstruction, managed with prucalopride, laxatives, and supportive care. Immunosuppressive therapy and nutritional support via TPN were crucial. This case emphasizes the complexities of managing gastrointestinal issues in pediatric patients with multiple autoimmune disorders, highlighting the need for a multidisciplinary and tailored approach.***

Keywords: SLE, Scleroderma, Polymyositis, chronic pseudo intestinal obstruction, gastrointestinal complications

1. Background

Chronic intestinal pseudo-obstruction (CIPO) is a severe gastrointestinal motility disorder characterized by significant impairment of gastrointestinal propulsion, leading to symptoms that mimic mechanical bowel obstruction despite the absence of physical blockage[(1,2)]. In pediatric patients with systemic lupus erythematosus (SLE) and scleroderma, CIPO presents with recurrent and severe abdominal pain, distension, nausea, vomiting, constipation, and occasionally diarrhea. The chronic nature of CIPO can result in substantial weight loss, malnutrition, and growth retardation, thereby profoundly affecting the patient's health and quality of life.[(3)]Diagnosing chronic intestinal pseudo-obstruction (CIPO) in patients with systemic lupus erythematosus (SLE) and scleroderma involves a comprehensive clinical evaluation. Imaging studies, including abdominal X-rays, typically reveal dilated bowel loops and air-fluid levels indicative of obstruction. Advanced diagnostic methods, such as gastrointestinal manometry, assess intestinal motility, while endoscopy or exploratory surgery are utilized to exclude mechanical obstruction. Blood tests and biopsies further help evaluate inflammation and fibrosis in the intestinal walls[(4)].

The treatment of chronic intestinal pseudo-obstruction (CIPO) in pediatric patients with systemic lupus erythematosus (SLE) and scleroderma is complex, requiring a multidisciplinary approach. Prokinetic agents like prucalopride are used to enhance bowel motility, while nutritional support, including total parenteral nutrition (TPN), is essential during severe episodes. Immunosuppressive therapies, such as corticosteroids and monoclonal antibodies, play a crucial role in controlling the underlying autoimmune activity. Effective management also involves pain control, maintaining electrolyte balance, and, in some cases, surgical interventions.[(5,6)]

Chronic intestinal pseudo-obstruction (CIPO) is a rare condition, but its incidence is elevated among pediatric patients with systemic lupus erythematosus (SLE) and scleroderma, reflecting the gastrointestinal involvement of these autoimmune diseases. The pathophysiology of CIPO in this context is primarily due to autoimmune-mediated damage

to the gastrointestinal tract's smooth muscle and nerves, leading to atrophy, fibrosis, and autonomic dysregulation. These pathological changes impair peristalsis, causing chronic pseudo-obstruction[7,2]. The incidence of chronic intestinal pseudo-obstruction (CIPO) among children under 15 years is reported to be 0.5-0.6 cases per 100,000 persons. Prevalence rates vary widely, from 4 to 250 cases per 100,000 persons, with higher prevalence observed in Native American, Asian American, Latin American, and African American populations..[8,3]This condition poses significant challenges in management due to its complexity and the need for a personalized treatment plan that addresses both the gastrointestinal symptoms and the underlying autoimmune disorders. Early diagnosis and intervention are crucial in preventing severe complications, improving the patient's quality of life, and ensuring better long-term outcomes.

2. Case Report

A 15-year-old female with a history of Systemic Lupus Erythematosus (SLE) overlapping with scleroderma and polymyositis, diagnosed in 2019, was admitted with a three-day history of sudden-onset, colicky abdominal pain, constipation, and oliguria. She also reported multiple episodes of non-bilious, non-projectile vomiting over the past day, with reduced oral intake. She had a similar episode one month prior, requiring hospitalization. Her current treatment regimen includes antirheumatics, immunosuppressive agents, Phosphodiesterase 5 (PDE5) inhibitors, corticosteroids, and monoclonal antibodies administered biannually.On examination, she was afebrile with a temperature of 97°F, tachycardic with a heart rate of 132 beats per minute, hypotensive with a blood pressure of 96/60 mmHg, and tachypneic with a respiratory rate of 24 breaths per minute. Oxygen saturation was 99% on room air. She appeared dull with tachycardia but had normal peripheral pulses. Systemic examination revealed normal heart sounds, clear lung fields, a distended abdomen with palpable fecoliths, diffuse tenderness, and sluggish bowel sounds. Neurologically, she was alert and fully oriented, with a Glasgow Coma Scale (GCS) score of 15/15. Her weight was recorded at 33 kilograms.

Course in the hospital

The patient was admitted with above mentioned complaints and her Initial treatment included a normal saline (NS) bolus (20 ml/kg), followed by maintenance intravenous fluids and intravenousAntibiotics.Immunosuppressive medications were withheld due to the patient's NPO status. An abdominal X-ray revealed fecal loading in the colon, and a Looz enema was administered. Laboratory investigations indicated hypokalemia (serum potassium 2.8 meq/L), leading to the addition of potassium chloride (KCl) to her maintenance fluids. Low ionized calcium on blood gas analysis prompted the administration of 10% calcium gluconate under cardiac monitoring.Following a paediatric surgical consultation, stool disimpaction and supportive care were advised. Post-enema, the patient passed stools, leading to reduced distension. Stool analysis detected *Entamoeba histolytica*, but in the absence of invasive infection, the infectious disease specialist recommended continuing antibiotics without antiparasitic treatment. The patient was kept NPO, and TPN was initiated due to difficulties with peripheral cannulation. Potassium levels were closely monitored, and KCl was titrated as needed. As NG aspirates decreased and repeate abdominal Xrays showed reduced fecal impaction, oral laxatives and enemas were maintained. ,Due to persistent vomiting and constipation,medical gastroenterologist consultation was taken and colonic pseudo-obstruction was suspected and Osmotically acting laxatives was advised to enhance motility. The NG tube was eventually removed as the patient tolerated clear liquids.Despite initial treatment, the patient continued to experience recurrent constipation and abdominal distension, necessitating proctoclysis enemas. Prucalopride was introduced to enhance gut motility, resulting in improved bowel movements. Imaging, including an MR enterography and abdominal ultrasound, revealed fecal impaction and dilated bowel loops without obstruction. The gastroenterology team recommended continuing prucalopride and laxatives. Attempts to discontinue prucalopride resulted in the return of constipation, leading to the resumption of the medication once daily at bedtime.During her hospital stay, the patient received two doses of intravenous Monoclonal antibodies with premedications, including anti histamines and corticosteroids, administered over 8–9 hours under close monitoring. Mild hypersensitivity reactions, such as fever and rash, were managed symptomatically. She also received intravenous immunoglobulin (IVIg) at a dose of 0.5 g/kg over three days without complications. Follow-up blood tests indicated corrected electrolyte imbalances, with normalized potassium levels and appropriate management of calcium and magnesium. At discharge, the patient was hemodynamically stable, on a fiber-rich diet, and ambulating regularly. She was passing stools once or twice daily with the aid of prucalopride and laxatives.

Table 1: CBP (Complete Blood Picture)

Parameters	Day 1	Day 4	Day 11	Reference range
Haemoglobin	10.5 ↓	8.9 ↓	10.5 ↓	12.0-15.0gm%
PCV (Haematocrits)	34.6 ↓	26.3 ↓	32.5 ↓	36.0-46.0 vol%
RBC (Red blood cells)	5.63 ↑	4.63 ↑	5.36 ↑	3.8-4.8 mill/cumm

Table 2: Serum Electrolytes

Parameters	Day 3	Day 4	Day 11	Day 15	Day 17	Reference value
Sodium	131.0 ↓	131.0 ↓	132.0 ↓	132.0 ↓	134.0 ↓	135-146 mmol/L
Potassium	3.3 ↓	3.4 ↓	5.3 ↑	4.1	3.3 ↓	3.5-5.1 mmol/L

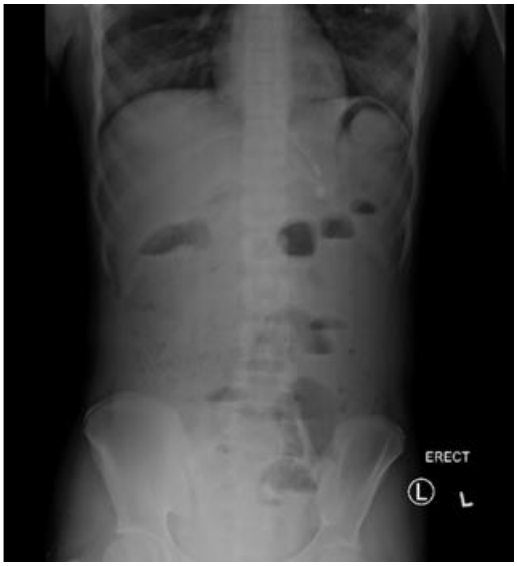

Figure 1

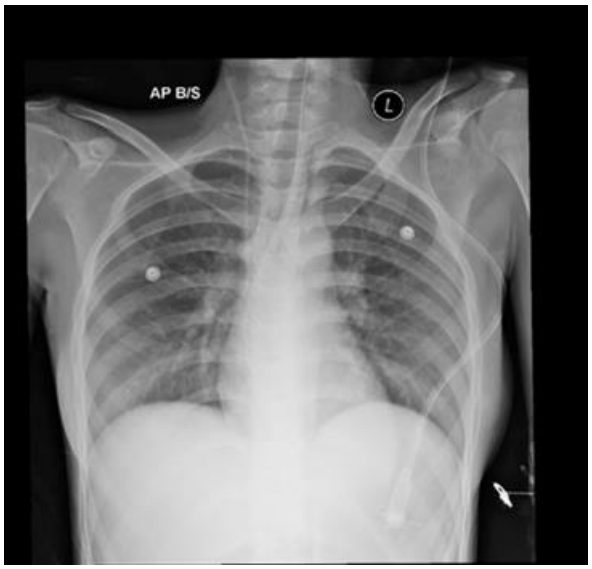

Figure 2

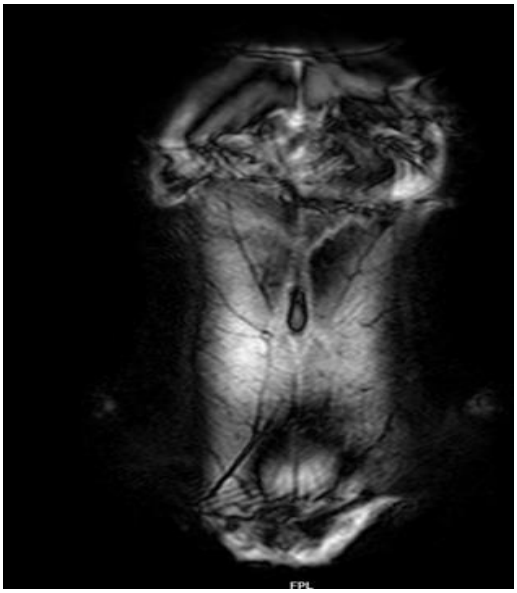

Figure 3

Figure 1: CT whole abdomen was performed and visualized nasogastric tube seen in in situ with distal end coiled in stomach. Visualized distal thoracic oesophagus appears distended with air fluid levels. No abnormal wall thickening. IC junction appears patulous. Distal ileum and caecum are well distended with fluid and show minimal wall thickening with mucosal enhancement. A solitary enlarged right ileac node noted which is reactive/inflammatory. Large bowel loops and rectum appears prominent and distended with faecal matter. Rest of the visualized bowel loops show normal wall thickness, attenuation and enhancement pattern. No imaging features of intestinal obstruction**.**

Figure 2: X ray chest AP view rotation present, mid expiratory film. Central venous line nasogastric tube insitu. Soft tissues and thoracic cage are normal

Figure 3: MRI whole abdomen shows jejunal and ileal loops are normal in caliber wall thickness signal intensity and enhancement pattern. The terminal ileaum is mildly dilated maximal diameter measuring 3.2cms., large bowel loops are loaded with fecal matter. Few small volume ileocolic lymphnodes are seen. Fluid is seen in the visualized lower esophagus.

3. Discussion

This case underscores the complexity of managing gastrointestinal complications in a pediatric patient with a history of Systemic Lupus Erythematosus (SLE) overlapping with scleroderma and polymyositis. Gastrointestinal involvement in SLE is well-documented, ranging from mild symptoms to severe complications like bowel infarction. A study conducted in France by Halabi et al. (2019) highlighted that up to 50% of SLE patients experience some form of gastrointestinal involvement, often exacerbated by overlapping conditions like scleroderma, which contributes to motility issues due to smooth muscle atrophy and fibrosis[(9)]. In this case, the presence of polymyositis likely worsened the gastrointestinal symptoms by impairing muscle function in the GI tract, leading to colonic pseudo-obstruction. This condition, also known as Ogilvie syndrome, is characterized by severe constipation, abdominal distension, and vomiting, and was managed in this patient with supportive care, prucalopride, and osmotic laxatives. The effectiveness of prucalopride in enhancing bowel motility is supported by a study by Camilleri et al. (2008), which demonstrated its efficacy in treating chronic constipation in patients with gastrointestinal dysmotility.[(10)]The patient's immunosuppressive therapy, including corticosteroids and monoclonal antibodies, was critical in controlling the underlying autoimmune activity, though it required careful administration to avoid exacerbating gastrointestinal issues. A systematic review conducted found that Rituximab, a monoclonal antibody, is effective in managing severe SLE cases, though it carries a risk of hypersensitivity reactions, which were observed and managed in this patient[(11)]. Nutritional support, provided through total parenteral nutrition (TPN) and later a fiber-rich diet, was essential for maintaining the patient's health during acute episodes of gastrointestinal distress. In another study conducted emphasized the importance of nutritional management in patients with systemic autoimmune diseases, particularly when gastrointestinal complications are present.[(12)] This case highlights the importance of a multidisciplinary approach and

individualized treatment plans in managing complex gastrointestinal symptoms in patients with SLE and overlapping autoimmune conditions. The successful resolution of symptoms through a combination of supportive care, prokinetic agents, and careful nutritional management reflects the need for tailored therapeutic strategies, as supported by current literature and studies in this field.

4. Conclusion

This case report illustrates the challenges in managing gastrointestinal complications in a pediatric patient with Systemic Lupus Erythematosus (SLE) overlapping with scleroderma and polymyositis. The patient presented with severe gastrointestinal symptoms, including colonic pseudo-obstruction, which were successfully managed through a multidisciplinary approach involving supportive care, prokinetic agents like prucalopride, and osmotic laxatives. The complexities of balancing immunosuppressive therapy, managing hypersensitivity reactions, and ensuring adequate nutritional support were critical to the patient's recovery. The tailored dietary approach underscores the critical role of nutrition in the comprehensive management of complex autoimmune conditions, emphasizing its importance in both symptom management and overall patient recovery. The outcome underscores the necessity of individualized treatment strategies in managing such complex autoimmune conditions, aligning with current research that emphasizes the importance of tailored therapeutic interventions in similar cases.

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