

Case Study

The Great Imitator Strikes Again: A Case of a Lupus Flare-up Presenting Like an Acute Abdomen

Emmanouil Kiriakopoulos, DO,^{1,2} Vanessa Perez, MD,³ Robyn Hoelle, MD^{1,2}

Abstract

Description

Systemic lupus erythematosus (SLE) is an autoimmune disease affecting all age groups and can manifest in various forms, often making the initial or successive presentations difficult to diagnose. Peritonitis secondary to lupus is a rare manifestation of this disease and the prevalence is said to be much lower in children. We present a case report of an adolescent male with a known history of lupus who presented to the emergency department with a clinical picture consistent with an acute surgical abdomen and underwent an appendectomy. Subsequent workup identified the culprit as a lupus-related peritonitis requiring corticosteroids for resolution.

Keywords

SLE; lupus; pediatrics; acute abdomen; peritonitis; lupus peritonitis; appendicitis; systemic lupus erythematosus

Author affiliations are listed at the end of this article.

Correspondence to:

Emmanouil Kiriakopoulos,
DO
Department of Emergency
Medicine
North Florida Regional
Medical Center
6500 Newberry Road
Gainesville, FL 32605
(em.kiriakopoulos@gmail.com)

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease that can involve multiple organ systems leading to significant morbidity and mortality, contributing to over 2,000 deaths annually in the United States.¹ The annual incidence and prevalence have been reported as high as 23.2/100,000 and 241/100,000 respectively depending on age and ethnicity group.² The incidence and prevalence are believed to be much lower in the pediatric population, with previous studies reporting rates of 0.3-0.9/100,000 and 3.3-8.8/100,000 respectively.³ The various manifestations of the disease makes it difficult to diagnose. Even after initial diagnosis, SLE can present in multiple forms, often mimicking other disease processes. While much research has been centered on the adult population, there has been minimal concentration on the pediatric manifestations of SLE. We report a case of an adolescent male presenting to the emergency department with a clinical picture consistent with a common surgical emergency in this age group, but later determined to be a manifestation of SLE.

Case Report

A 16-year-old male with a medical history of poorly-controlled systemic lupus erythematosus (SLE) due to medication noncompliance, previously followed by rheumatology at an outside facility, presented to the emergency department (ED) from a routine appointment at a nephrology clinic. During that visit, his nephrologist found the patient to have abdominal pain, distention, and ascites with concern for an acute abdomen; therefore, he recommended further evaluation in the ED. Upon arrival to the ED, the patient stated that his abdominal pain began several days ago, but worsened yesterday with any movement or when pressure was applied to his abdomen. He and his family also commented that he had associated fevers, abdominal swelling, and nausea at home. The patient stated that he was seen in the ED two days prior for a several week history of general malaise and joint aches. At that time, it was suspected to be a lupus flare-up and he was discharged home with close rheumatology follow-up for re-initiation of his previous immunosuppressive therapy.

On physical exam, the patient appeared uncomfortable, but was afebrile and hemodynamically normal. His abdomen was remarkable for moderate distention, normal bowel sounds, and severe tenderness to the right lower quadrant with guarding and rebound tenderness. Additionally, there was a positive fluid wave consistent with ascites. Labs and blood cultures were drawn, and the patient was empirically started on ceftriaxone and metronidazole for concern of a developing intra-abdominal infectious process. An abdominal ultrasound was ordered and was remarkable for findings suggesting early appendicitis. Surgery was consulted emergently and decided to proceed with an appendectomy. It was noted during the surgery that the appendix was not grossly inflamed, although there was green-colored fluid in the peritoneum and pelvis, consistent with serositis. The fluid was not cultured as it reportedly did not appear purulent. Additionally, pathology evaluation of the appendix did not favor acute appendicitis.

Postoperatively, the patient's distention improved, although his abdominal pain was not well controlled despite intravenous pain medicine. Rheumatology was consulted and determined the patient was having an SLE flare-up. The patient was started on intravenous methylprednisolone which rapidly improved his pain and symptoms. He was transitioned to oral steroids and discharged two days later with a primary diagnosis of lupus peritonitis with instructions to follow up with his rheumatologist to discuss restarting his previous immunosuppressive regimen.

Discussion

Systemic lupus erythematosus is an autoimmune process that can involve any organ system, often making it difficult to initially diagnose due to its ability to mimic other ailments. Common presentations of SLE, both in adult and pediatric populations, include constitutional symptoms such as fatigue, arthralgia, fever and weight loss, and mucocutaneous findings such as the classic butterfly or malar rash of the face. Less commonly, but with a greater risk of morbidity, it can present with nephritis, pericarditis, peritonitis and vasculitis.³ Acute peritonitis secondary to SLE, as in this case study, is rare: some authors have estimated the

prevalence to be less than 10% in the pediatric age group.³⁻⁵ While rare, it should be considered a diagnosis of exclusion and the presentation of ascites with peritoneal signs should warrant a thorough investigation for more common causes such as spontaneous bacterial peritonitis, appendicitis, perforated viscus and malignancy.

Management for lupus peritonitis has been focused around bowel rest, intravenous or oral corticosteroids and possibly the initiation of other immunosuppression medications.^{4,6} Our patient in this case report responded favorably to intravenous glucocorticoids. In cases where ascites does not improve, another case study found success with intraperitoneal steroid injections.⁷

Conclusion

This case report shows that even in a known diagnosis of SLE, uncommon presentations of the disease can suggest a more common etiology of the presenting signs and symptoms. Here, we discussed an adolescent male with known SLE who initially presented with a clinical picture, supported with ultrasonography, concerning for an acute surgical abdomen. The patient underwent appendectomy, however, the appendix was not found to be the culprit. This case should serve as a reminder to maintain a high index of suspicion for the underlying pathophysiology of a disease process such as SLE, to be familiar with all of the manifestations, but to never forget the more common emergencies that may present in a similar fashion.

Conflicts of Interest

The authors declare they have no conflicts of interest.

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Author Affiliations

1. North Florida Regional Medical Center, Department of Graduate Medical Education
2. UCF/HCA Healthcare GME Consortium of North Florida
3. Nemours Children's Hospital, Department of Emergency Medicine

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