Pericardial tamponade presenting as abdominal pain in a patient with systemic lupus erythematosis

Antonio E. Muñiz, MD

ABSTRACT: Abdominal pain is a common complaint in children presenting to the emergency department. Patients with systemic lupus erythematosus (SLE) are at increased risk of infection, bowel necrosis and perforation, especially if they are taking nonsteroidal anti-inflammatory drugs, corticosteroids or immunosuppresive agents, which may mask the signs and symptoms of the underlying condition, hence delay diagnosis and treatment. This article presents the case of an adolescent girl whose abdominal pain was related to cardiac tamponade and provides an overview of the cardiac and gastrointestinal manifestations of SLE.

RÉSUMÉ : La douleur abdominale est un symptôme couramment rencontré chez les enfants reçus à l'urgence. Les patients atteints de lupus érythémateux aigu disséminé (LEAD) courent un risque plus grand d'infection, de nécrose intestinale et de perforation, surtout s'ils reçoivent des anti-inflammatoires non stéroïdiens, des corticostéroïdes ou des immunosuppresseurs qui peuvent masquer les signes et symptômes de la condition sous-jacente, et par le fait même, retarder le diagnostic et le traitement. Le présent article décrit le cas d'une adolescente dont la douleur abdominale était liée à une tamponnade cardiaque et offre un aperçu des manifestations cardiaques et gastro-intestinales du LEAD.

Key words: abdominal pain, systemic lupus erythematosis, pericardial effusion, cardiac tamponade

Introduction

Children frequently present to the emergency department (ED) with abdominal pain.[1] The clinical presentation is often complex and it is not always possible to make a definitive diagnosis during the ED visit, but it is important to recognize emergent conditions that require immediate surgical intervention.

Clinicians should think of common conditions first, but when developing a differential diagnosis they must consider risk factors and comorbid illnesses that predispose patients to uncommon entities. Patients with systemic lupus erythematosus (SLE) are at increased risk of infection, bowel necrosis and perforation, especially if they are taking nonsteroidal anti-inflammatory drugs (NSAIDs), corticosteroids, or immunosuppresive agents, which may mask the clinical findings associated with acute abdominal processes. This paper presents the case of an adolescent woman with systemic lupus who developed pericardial tamponade and presented to the ED with abdominal pain. It then reviews the cardiac and gastrointestinal (GI) manifestations of SLE.

Case report

A 17-year-old adolescent girl presented to the ED with a history of 6 hours of right-sided abdominal pain that had awakened her from sleep. The pain was constant and intermittently sharp. She had vomited 3 times and had 7 epi-sodes of watery, non-bloody diarrhea. Associated symptoms included a bifrontal headache, dysphagia and dry cough. She denied chills, fever, dyspnea, chest pain, vaginal discharge or bleeding, dysuria, frequency or gastrointestinal bleeding.

Department of Emergency Medicine and Pediatrics, Medical College of Virginia, Richmond, Va.
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She related a history of seizures for which she took phenytoin. Five days prior to presentation her phenytoin had been discontinued because of a hypersensitivity reaction, and carbamazepine initiated. Her medical history was also remarkable for systemic lupus, diagnosed 2 months earlier when she developed dyspnea, chest pain and fever. At that time, she was found to have a pleural effusion and pericardial tamponade and underwent a pericardial window procedure. She denied smoking, alcohol consumption or illicit drug use. Her last menstrual period was one week previous and she was sexually active. Her medications at the time of presentation included azathioprine, prednisone and hydroxychloroquine sulfate.

On examination, she had pale conjunctivae and was in mild discomfort. Blood pressure was 128/97 mm Hg, pulse 130 beats/min, respiratory rate 20 breaths/min, temperature 37.2ºC, and oxygen saturation 97% on room air. Breath sounds were diminished at the left lung base, with no rales or wheezes, and heart sounds were distant, without gallops, rubs, clicks or murmurs. Jugular venous pressure was 8 cm. The abdomen was slightly distended and diffusely tender, with right upper quadrant guarding. Bowel sounds were hypoactive. There was mild right costovertebral tenderness and her liver was palpable 5 cm below the right costal margin. Rectal and pelvic examination were normal, and she had no dependent edema. A blanching red maculopapular rash was present over her extremities and the malar area of her face.

Her white blood cell count was 17,000/mm^3, with 62% neutrophils, 2% bands, 17% lymphocytes and 10% monocytes. Hematocrit was 32.6% and platelet count 310,000/mm^3. Serum electrolytes, erythrocyte sedimentation rate, liver function tests and urinalysis were normal. An abdominal film was unremarkable, and the chest x-ray revealed an enlarged cardiac silhouette with prominent vascular markings and a left pleural effusion. The electrocardiogram showed sinus tachycardia at 120 beats/min with low voltage and electrical alternans (Fig.1). Her pregnancy test was negative.

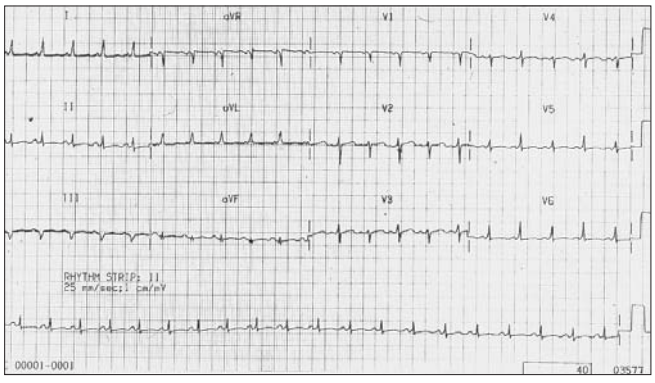

Fig. 1. Electrocardiogram showing sinus tachycardia at 120 beats/min with low voltage and electrical alternans.

A surgeon examined her and felt she did not require surgical intervention. At this point, 2D-echocardiography was performed. This revealed a moderate pericardial effusion with adequate systolic pump function, no right ventricular collapse and no equalization of ventricular pressures. Because the etiology of her symptoms was not apparent, the patient was admitted to the intensive care unit for further monitoring and evaluation.

A few hours later, she developed increasing abdominal pain, dyspnea, confusion and hypotension requiring intravenous fluid boluses. Repeat 2D-echocardiography demonstrated a larger pericardial effusion with right ventricular collapse. The patient was taken to the operating room, where 500 mL of serosanguinous fluid was drained from the pericardial space. Her hemodynamic status normalized and pericardial stripping was performed. Postoperatively, all symptoms, including abdominal pain, resolved. No other cause for the pain was identified, no other treatments were provided to account for her improvement, and the pain was attributed to passive liver congestion due to pericardial tamponade.

Discussion

There are few reported cases of patients with pericardial tamponade who presented with abdominal pain,[2–6] and most of these had concurrent abdominal conditions such as mesenteric ischemia,[3] pancreatitis,[4] or splenic, hepatic and pancreatic contusions[5] to explain their pain. This report chronicles a patient with SLE who presented with abdominal pain due to pericardial tamponade and hepatic congestion; however, patients with systemic lupus are also at risk of various other abdominal pathologies.

Overview

Systemic lupus erythematosus is a chronic multisystem collagen vascular disorder. It is more common in females, African–Americans, Orientals and Hispanics, and tends to be more severe in children than adults.[7] Although 20% of cases begin during childhood, prevalence is only 0.6 per 100,000 children and onset is rarely before age 8.[8] SLE is characterized by abnormal B-cell activation with excess autoantibody production, tissue immune complex deposition and defective T-lymphocyte suppressor function. The cause is unknown, but may be related to genetic, environmental, infectious or hormonal factors affecting immunoregulation. Several drugs have been implicated, including procainamide, isoniazid, hydralazine, phenytoin, anticoagulants, clonidine, penicillin, sulfonamides, tetracycline and methyldopa.[9]

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Clinical presentation

Patients may present with single- or multiple-organ involvement, but children usually present with severe, multisystem disease.[10] Fatigue, fever, malaise, arthritis, arthralgia, anorexia and rash are the most common presenting symptoms,[11] and the disease is characterized by remissions and exacerbations.

Pericarditis is the most common cardiac manifestation of SLE, occurring in 25%–30% of children,[11,12] usually during an acute exacerbation. Pericardial effusions may be asymptomatic and are typically not associated with friction rubs. Diminished heart sounds suggest the presence of pericardial fluid. Low voltage and electrical alternans on the ECG are classic, and chest radiographs may show clear lung fields and an enlarged cardiac silhouette, sometimes with a "water bottle" appearance. Most effusions do not lead to cardiac tamponade, and tamponade as an initial manifestation of SLE is distinctly unusual.[13,14]

The hemodynamic and clinical effects of a pericardial effusion depend on fluid volume, rapidity of accumulation and myocardial competence.[15] When fluid accumulates gradually, compensatory mechanisms come into play and the parietal pericardium stretches to adapt to the increasing volume. In this situation, relatively large pericardial fluid volumes may cause only subtle symptoms. Conversely, rapid accumulation of small volumes may cause dramatic symptoms.

The clinical manifestations of pericardial tamponade are due to reduced cardiac output and systemic venous congestion. Beck's triad (hypotension, faint heart sounds and jugular venous distention) is seen with rapid onset tamponade — for example, traumatic myocardial rupture. When tamponade develops slowly, patients are more likely to exhibit dyspnea, tachypnea, orthopnea, tachycardia, hypotension, jugular venous distention and pulsus paradoxus. These features may initially be absent, but most should be apparent in hemodynamically compromised patients.

Myocarditis occurs in 10% of children with SLE[11] and may cause congestive heart failure, cardiomegaly or electrocardiographic changes (most often flat or inverted T waves). Sterile verrucous (Libman–Sacks) endocarditis has been reported in lupus patients but is generally clinically silent.[16]

Diagnosis

Patients with GI symptoms pose a difficult clinical challenge. The differential diagnosis includes active lupus, acute pathology unrelated to lupus, and adverse effects of lupus therapy. SLE-related enteritis, vasculitis or mesenteric thrombosis may cause bowel ischemia, bloody diarrhea, infarction or perforation requiring surgical intervention, although these are relatively rare.[17] Non-SLE related causes of abdominal pain — including appendicitis, cholelithiasis, pancreatitis, hepatitis, bowel ischemia, obstruction, pneumonia, myocardial ischemia and gynecologic conditions — are at least as frequent as those related to lupus.[8,17] To complicate matters, corticosteroid, antiinflammatory, antimalarial and immunosuppressive drugs used to treat SLE may cause nausea, vomiting, abdominal pain, ulcer formation, GI bleeding and bowel perforation, and may obscure the signs of an evolving surgical abdomen. Therefore, while SLE victims are at increased risk of abdominal catastrophe, the decision to proceed to laparotomy is often difficult and failure to recognize the need for surgical intervention may increase mortality and morbidity.

Management

In the absence of cardiac involvement, treatment for SLE is dictated by disease severity and may include NSAIDs, corticosteroids, antimalarials, and immunosuppressive agents. In cases of suspected pericardial tamponade, urgent echocardiography will confirm the diagnosis, define the size of the effusion and demonstrate right ventricular collapse during diastole. Echocardiography also is useful in assisting with needle guidance during pericardiocentesis.

ED treatment of pericardial tamponade depends on patient stability. In patients who are not in shock, rapid volume infusion may increase ventricular filling pressures and mitigate symptoms until pericardiocentesis can be performed. In critically ill patients, when hemodynamic instability precludes echocardiographic diagnosis, emergent pericardiocentesis is diagnostic and therapeutic.[13] After pericardiocentesis, high-dose corticosteroid therapy is recommended to enhance fluid resolution and clinical remission.[13] Large, resistant or recurrent effusions may require a surgical pericardial window; however, approximately 10% of patients undergoing this procedure will develop recurrent effusions, which are generally treated with immunosuppresive agents.[18,19] Patients who fail a surgical pericardial window and immunosuppressive therapy usually require pericardial stripping.

Conclusion

Comorbid illnesses may predispose patients to rare conditions. Abdominal pain is common in patients with SLE and may be due to active lupus, an unrelated abdominal pathology, or an adverse effect of therapy. A careful history and physical examination are essential in cases where the cause of abdominal pain seems obscure.

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Correspondence to: Dr. Antonio Muñiz, 401 North 12th St., PO Box 980401 Richmond VA 23298-0401, USA; aemuniz@aol.com

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