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Case report

# Acute acalculous cholecystitis in systemic lupus erythematosus: a rare initial manifestation☆

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ABSTRACT

Acute acalculous cholecystitis is a very rare gastrointestinal manifestation in systemic lupus erythematosus and becomes rarer as an initial manifestation. There are only two cases reported. The authors report a 20-year-old black woman that presented acute acalculous cholecystitis revealed by abdominal computed tomography. During hospitalization, she was diagnosed systemic lupus erythematosus. Conservative treatment with antibiotics was performed with complete remission of the symptoms. Corticosteroid was started in ambulatory. Cholecystectomy has been the treatment of choice in acute acalculous cholecystitis as a complication of systemic lupus erythematosus. The patient responded well to conservative treatment, and surgery was not required. This case is unique in the way that corticosteroid was started in ambulatory care. We should not forget that the acute acalculous cholecystitis can be the initial presentation of systemic lupus erythematosus although its occurrence is very rare. Conservative treatment should be considered. Abdominal computed tomography was a determinant exam for better assessment of acute acalculous cholecystitis.



## Colecistite aguda acalculosa no lúpus eritematoso sistêmico: uma manifestação inicial rara

*Palavras-chave:*
Lúpus eritematoso sistêmico
Colecistite aguda acalculosa
Tomografia computadorizada do abdome
Manifestação inicial

RESUMO

A colecistite aguda acalculosa é uma manifestação gastrointestinal rara no lúpus eritematoso sistêmico e ainda mais rara como manifestação inicial. Foram descritos apenas dois casos até o momento. Os autores relatam o caso de uma mulher negra de 20 anos, com quadro de colecistite aguda acalculosa revelada pela tomografia computadorizada do abdome. Durante a hospitalização, a paciente foi diagnosticada com lúpus eritematoso sistêmico. Houve remissão completa dos sintomas após tratamento conservador com

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antibióticos. Iniciou-se tratamento com corticosteroides no ambulatório. Embora a colecistectomia seja o tratamento de escolha em casos de colecistite aguda acalculosa como complicação do lúpus eritematoso sistêmico, a paciente respondeu bem ao tratamento conservador; logo, a cirurgia não foi necessária. Este caso é único em razão do modo como o corticosteroide foi iniciado no atendimento ambulatorial. É importante lembrar que a colecistite aguda acalculosa pode ser a manifestação inicial do lúpus eritematoso sistêmico, embora sua ocorrência seja rara. Deve-se considerar a realização de tratamento conservador. A tomografia computadorizada do abdome foi determinante para que fosse feita uma melhor avaliação da colecistite aguda acalculosa.



## Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease found predominantly in female gender[1–3] in which almost all organs can be involved with wide range of clinical manifestations. Gastrointestinal manifestation is usually mild, but gallbladder involvement is an uncommon event. Acute acalculous cholecystitis (AAC) is very rare as a complication of SLE and becomes rarer as an initial presentation.[1–9] Several cases of AAC as a complication of SLE were reported; nevertheless only two cases have been reported as initial manifestation.[5,9]

## Case report

A 20-year-old black woman with history of migratory polyarthralgia was admitted with fever, nausea and vomit, acute abdominal pain in the right upper quadrant and referred loss of appetite. Physical examination evidenced febrile (38.6 °C), tachycardia (102 beats per minute), dehydrated and subicteric mucous membranes. Abdominal pain in the right upper quadrant with hepatomegaly of 8 cm below the costal margin was found. Cardiac auscultation, revealed a grade-II/VI protosystolic murmur in all cardiac focus. Initial laboratory data showed hemoglobin 7.6 g/dL, leucocytes 3300/L, albumin 2.2 g/dL, alanine aminotransferase 86 U/L, aspartate aminotransferase 127 U/L, total bilirubin 1.8 mg/dL; lactate dehydrogenase 1719 U/L and test for *plasmodium* negative. Abdominal ultrasound confirmed hepatomegaly and revealed splenomegaly, gallbladder slightly distended without wall thickening. During hospitalization, the patient evolved with retrosternal pain, orthopnea, surfeit sensation, increased pain in the right upper quadrant, nausea, vomiting, diarrhea and fever. Pulmonary auscultation revealed abolished in both bases. Cardiac auscultation was the same. The abdomen was distended with hepatomegaly of 8 cm below the right costal margin; liquid wave signal and *murphy* were present. Lower limbs presented malleolar edema. The main laboratory findings are shown (Table 1). Teleradiography of chest showed bilateral pleural effusion of small volume. Echocardiography revealed pericardial effusion of 1.5 cm. Abdominal computed tomography revealed increased wall thickness without any evidence of stone and edema around the gallbladder (Fig. 1), and confirmed splenomegaly, hepatomegaly and ascites. Based on the physical examination, laboratory and image findings, a diagnosis of inaugural systemic lupus erythematosus triggered by acute acalculous cholecystitis was established. The patient initiated treatment with ceftriaxone and metronidazole, furosemide and albumin. Within 10 days, there was complete remission of symptoms. The corticosteroid was initiated in ambulatory of rheumatology. Surgical intervention was not performed. According to the rheumatologists' feedback, she looked well and did not have any symptoms until four months after discharge.

**Table 1 – Laboratory findings during admission course.**

| Exam | Results | Reference values |
|---|---|---|
| Test for *Plasmodium* | Negative | Negative |
| Hemoglobin | 6.9 g/dL | 11.5–15.5 |
| Leukocytes | $3.2 \times 10^9$/L | 04–11 |
| Platelets | $342 \times 10^9$/L | 150–400 |
| Total proteins | 5.2 mg/dL | 6.5–8.1 |
| Proteinuria-24 h | 174 mg/dia | <150 |
| Albumin | 2.3 mg/dL | 3.5–5.0 |
| ALT | 80 U/L | <40 |
| AST | 112 U/L | <40 |
| LDH | 1557 U/L | 230–460 |
| Alkaline phosphatase | 155 U/L | 32–92 |
| GGT | 256 U/L | <50 |
| Creatinine | 0.4 mg/dL | 0.6–1.3 |
| Urea | 4 mg/dL | 0.8–26 |
| Amylase | 28 U/L | 36–128 |
| Anti-HBs | 0.44 S/CO | <0.90 |
| Anti-HCV | 0.8 S/CO | <0.9 |
| Toxoplasmosis | 0.2 S/CO | <0.8 |
| ANA[a] | 6 IU/mL | >1.1 positive |
| Anti-dsDNA[a] | 80 IU/mL | >60 positive |
| C3[a] | 45.2 mg/dL | 79–152 |
| C4[a] | 7.1 mg/dL | 16–38 |
| Anticardiolipin IgM[a] | 7.9 U MPL/mL | 0.0–7.0 |
| Anti-β2 glycoprotein 1 IgM[a] | 12.1 U MPL/mL | 0.0–5.0 |
| Anti-β2 glycoprotein 1 IgG[a] | 5.5 U GPL/mL | 0.0–5.0 |
| Smooth muscle antibody[a] | 1:80 | <1:20 |

ALT, alanine aminotransferase; AST, aspartate aminotransferase; LDH, lactate dehydrogenase; GGT, gamma-glutamyl transferase; ANA, antinuclear antibody.

[a] Tested in pathology laboratory support services – AMPATH.

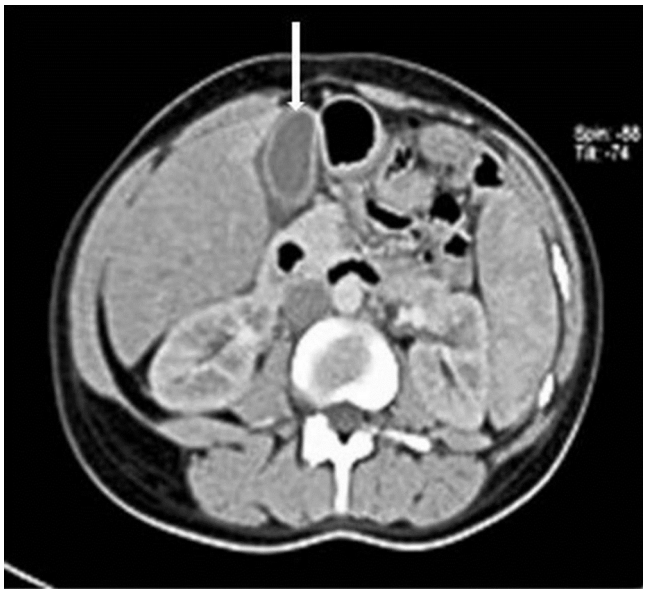

**Fig. 1 – Abdominal CT show gallbladder wall thickness without stone with edema around (arrow).**

## Discussion

Abdominal pain is always a challenge for diagnosis and treatment in patients with SLE; it may be caused by the disease itself, other comorbidities or drug effects.[1–3] The involvement of gastrointestinal and hepatobiliary systems sparing gallbladder have been reported.[1–9] The deposition of immune complexes in blood vessel walls results in acute vasculitis; these events result in ischemia and fibrosis in the target organ.[4–9] Thromboembolic events are most frequently observed in patients with SLE; this risk is increased when anticardiolipin antibody is positive,[10] as in the case of anticardiolipin IgM, anti-β2 glycoprotein 1 positive, although it was a low titer. The patient does not have criteria for diagnosis of antiphospholipid syndrome.[10] Histologically, antiphospholipid syndrome is characterized by multiple thrombi in the vessels of the gallbladder without evidence of vasculite.[5] The gallbladder is rarely involved in patients with SLE.[1–9] Several cases of AAC as a complication of SLE were reported, but only two cases of AAC as an initial manifestation of SLE were described, one in a pediatric patient and other in an adult.[5,9] The most widely used classification criteria for diagnosis of SLE are those proposed by the American College of Rheumatology and Systemic Lupus International Collaborating Clinics that require four or more items to diagnose SLE (cutaneous manifestation, joints, serositis, renal disorder, hematologic disorder, immunologic abnormality).[11,12] In this case, the patient presented with pain in the peripheral joints, pleural and pericardial effusion, proteinuria, hemolytic anemia, leukopenia, positive ANA, positive anti-dsDNA, positive anti-β2 glycoprotein 1, positive Sm and low complement. When there is a suspicion of involvement of the gallbladder as the cause of abdominal pain in these patients, abdominal ultrasound and computed tomography (CT) are indicated for better assessment of AAC.[13] In this case, the abdominal ultrasound performed at admission was inconclusive. The abdominal CT performed was essential for revealing the gallbladder wall thickness with edema around the gallbladder without the presence of stones, important findings for the diagnosis of AAC cases that have not been disclosed by the ultrasound made in the admission. No other risk factors for cholecystitis were found. In this case, AAC may be due to multiple thrombi in the gallbladder vessels.[8] The treatment of AAC has been controversial. There are four cases reported of patients with AAC in SLE, which did not have cholecystectomy, and responded well to medical treatment with high doses of corticosteroid.[4,5,7,9] In this case, due to major improvement of the symptoms with antibiotics, corticosteroid was started in outpatient follow-up. Surgery was not performed which justify no histological description. High doses of corticosteroids are usually suggested as first-line treatment if patients have a good general condition without other cholecystitis risk factors, no serious health complications and no infections.[7]

## Conclusions

Despite the rarity, we should not forget that acute acalculous cholecystitis, besides being a complication, can be the initial manifestation of SLE. Conservative treatment should be considered. In this case, corticosteroid was started after patient discharge; it made this case unique. Computed tomography is a good exam for better assessment of AAC.

## Conflicts of interest

The authors declare no conflicts of interest.

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