

# A 28-Year-Old Woman Presenting with a Clinical Flare of Systematic Lupus Erythematosus and Abdominal Pain Due to Rectus Sheath Hematoma

Authors' Contribution:  
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Data Collection B  
Statistical Analysis C  
Data Interpretation D  
Manuscript Preparation E  
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## Conflict of interest:

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None declared

**Patient:** Female, 28-year-old  
**Final Diagnosis:** Class III (A/C) and class V lupus nephritis • neuropsychiatric systemic lupus erythematosus • rectus sheath hematoma • systemic lupus erythematosus  
**Symptoms:** Abdominal pain • edema • abdominal distension • frothy urine • cough • slight dyspnea  
**Medication:** —  
**Clinical Procedure:** —  
**Specialty:** Nephrology • Rheumatology





**Objective:** Rare coexistence of disease or pathology  
**Background:** A flare, or flare-up, of systematic lupus erythematosus (SLE) is diagnosed by an increase in disease activity in one or more organs, new symptoms, or changes in laboratory measurements. A hematoma can occur in the sheath of the rectus abdominis following muscle trauma or rupture of an epigastric vessel, or it can occur spontaneously. This report is of a 28-year-old woman who presented with a clinical flare of SLE and abdominal pain due to rectus sheath hematoma.

**Case Report:** A 28-year-old woman had been suspected of having SLE 9 years ago and had received glucocorticoid therapy combined with hydroxychloroquine. However, lupus flared after she discontinued glucocorticoids, and she was admitted with a 1-month history of marked generalized edema, abdominal distension, frothy urine, and massive ascites. During hospitalization, she abruptly developed a continuous, stabbing abdominal pain and a bulge over the right abdomen as a result of straining during a bowel movement. On examination, a well-demarcated round mass that measured 121×96 mm was detected in the right quadrant. Abdominal emergency computed tomography revealed a right rectus sheath hematoma (21.4×4.7 cm). After her condition improved, the patient underwent an ultrasound-guided renal biopsy and was diagnosed with class III (A/C) and class V lupus nephritis.

**Conclusions:** This case has shown that spontaneous rectus sheath hematoma can occur without a history of trauma in a patient with an exacerbation of SLE. This association appears to be rare, and the cause is unknown.

**Keywords:** Hematoma • Lupus Erythematosus, Systemic • Lupus Nephritis

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 2013  1  5  19



## Background

Systemic lupus erythematosus (SLE) is a multisystemic disease with protean manifestations [1]. SLE flares are common in the disease course and can lead to permanent organ damage [2]. Although this disease is difficult to grasp and define, the new European League Against Rheumatism/American College of Rheumatology classification criteria for SLE added antinuclear antibodies to the entry criterion and provide an improved diagnosis of combined sensitivity and specificity [3]. Treatment of SLE aims at remission or low disease activity and prevention of flares [1]. Lupus nephritis influences the outcomes of SLE and guides the treatment and prognosis by its histologic classification, especially the 2003 revised International Society of Nephrology-Renal Pathology Society (ISN/RPS) classification, which is more advanced in predicting renal outcome [4].

Rectus sheath hematoma (RSH) is another rare clinical entity but has been previously reported in a patient with SLE [5]. RSH is characterized by accumulation of blood within the rectus sheath caused by disruption of the superior epigastric arteries, inferior epigastric arteries, and their branches or by direct damage to the abdominal wall [6]. Although it has been detected in only less than 2% of patients who present with acute abdominal pain, the overall mortality rate has been estimated at about 4% [6]. The mortality rate can be as high as 25% in patients on anticoagulation treatment and is 2 to 3 times higher in pregnant women [7]. Therefore, it is important to promptly identify RSH and make appropriate management.

This report is of a 28-year-old woman who presented with a clinical flare of SLE and abdominal pain due to RSH.

## Case Report

A 28-year-old woman presented with photosensitivity, leukopenia, and positive antinuclear antibodies in 2013 and was suspected of having SLE, according to the 1982 American College of Rheumatology classification criteria for SLE. She received glucocorticoid therapy combined with hydroxychloroquine. Her initial daily dose of glucocorticoid was 25 mg of prednisone, which was tapered to a maintenance dose of 15 mg. The woman had not undergone renal biopsy and did not have a family history of SLE. She did have a history of cesarean delivery and appendectomy conducted in 2008 and 2010, respectively. She also reported noncompliance with the glucocorticoid regimen and had discontinued the use of oral prednisone and hydroxychloroquine since June 2016 because she had experienced acid reflux and indigestion. She refused to undergo regular treatment and was lost to follow-up until she consulted the hospital for severe edema in 2017 and was admitted to the Department of Nephrology in September 2017

with a 1-month history of marked generalized edema, abdominal distension, frothy urine, cough, and slight dyspnea, without oral ulcer or arthritis.

Physical examination showed that her temperature was 36.5°C; respiratory rate, 19 breaths/min; oxygen saturation, 99% in room air; blood pressure, 153/82 mmHg; and weight, 77 kg. She had gained about 13.5 kg body weight within the span of 1 month. She had no rash or lymphadenopathy. Cardiopulmonary examination revealed hydrothorax. Shifting dullness was indicative of ascites, and her abdomen resembled that of a late-term gravid woman. Additionally, severe pitting edema was observed on both legs.

Laboratory tests (**Table 1**) were positive for the autoantibody ANA and related antibodies but negative for anti-dsDNA antibody. Hematuria, cylindruria, pyuria, and massive proteinuria (15.02 g/24 for 24-h urinary protein excretion) were also noted from the urinalysis. Pulmonary computed tomography (CT) examination in the outpatient department indicated pulmonary infection, and ceftriaxone was given.

A renal biopsy was not attempted due to severe edema and orthopnea. Treatment for a presumptive diagnosis of membrane lupus nephritis was commenced and initially comprised intravenous (i.v.) methylprednisolone (48 mg on days 1-15 of hospitalization) and i.v. immunoglobulin (on days 7-9) to block the autoantibody. Hemodialysis with heparin was done on day 6, as acute left heart failure was not relieved with the administration of furosemide and torsemide. On day 10 of admission, she abruptly developed a continuous, stabbing abdominal pain and a bulge over the right abdomen as a result of straining during a bowel movement. On examination, a well-demarcated round mass that measured 121×96 mm was detected in the right quadrant, without any redness, bruising, pulsation, or ecchymoses. An abdominal radiograph showed that the bowel appeared normal, without dilatation or excess air-fluid levels, and her serum lipase and amylase levels were within the reference range. Emergency CT of the abdomen (without administration of i.v. contrast) was performed. The image revealed a right RSH, massive ascites, and edema of the abdominal and intestinal walls (**Figure 1**); type 2 hematoma was classified in line with the bleeding between the fascia transversalis and the muscle from the CT scan [5]. There was no indication for an operation of the hematoma, given that her blood pressure, heart rate, and hemoglobin level were stable.

She experienced acute kidney injury on day 13 as loss of blood caused by RSH. At this point, the SLE disease activity index of the patient was 19 (based on proteinuria, hematuria, cylindruria, pyuria, leukopenia, and pleurisy). A persistent lupus flare was diagnosed, and she was treated with a pulse of 500 mg of i.v. methylprednisolone daily for 3 consecutive days (days

**Table 1.** Laboratory data.

Variable	On admission	Day 42	Day 216	Reference range
WBC ( $\times 10^9/L$ )	3.71	2.35	8.46	3.50-9.50
Hemoglobin (g/L)	118	90	72	115-150
Platelets ( $\times 10^9/L$ )	103	113	112	125-350
ESR (mm/H)	24	–	74	0-15
Creatinine ( $\mu\text{mol/L}$ )	114	179	102	46-92
Albumin (g/L)	12.3	17.7	26.4	40-55
Cholesterol (mmol/L)	12.1	8.1	6.42	0-5.72
Triglyceride (mmol/L)	18.07	5.8	3.36	0-1.70
LDL-C (mmol/L)	5.49	5.36	4.43	0-3.64
24-h urinary protein excretion (g/24 h)	15.02	11.46	1.13	0.05-0.1
Urinalysis				
Protein	4+	4+	–	Negative
WBC (/HP)	20-30	2-5	–	0-5
RBC (/HP)	15-20	4-7	–	0-3
Hyaline cast (/LP)	10-15	1-3	–	0-1
granular cast (/LP)	30-40	0	–	0-1
Rheumatological studies				
ANA	1: 1000	1: 100	<1: 100	<1: 100
dsDNA	Negative	Negative	Negative	Negative
LA	Negative	–	–	Negative
nRNP/Sm	3+	–	3+	Negative
Sm Ab	1+	–	Negative	Negative
Ro-52	3+	–	1+	Negative
SSA	3+	–	2+	Negative
AnuA	1+	–	Negative	Negative
AHA	2+	–	Negative	Negative
aPL	Negative	–	Negative	Negative
ANCA	Negative	–	–	Negative
C3 (g/L)	0.13	0.40	1.27	0.75-1.8
C4(g/L)	0.14	–	0.18	0.09-0.36

WBC – white blood cell; ESR – erythrocyte sedimentation rate; LDL-C – low-density lipoprotein cholesterol; HP – high-power field; RBC – red blood cell; LP – low-power field; ANA – antinuclear antibodies; ds-DNA – double-stranded DNA; LA – lupus anti-coagulant; Ab – antibodies; aPL – antiphospholipid antibodies; ANCA – antineutrophil cytoplasmic antibody.

16-18) followed by 80 mg of i.v. methylprednisolone daily for the next month. Additionally, 600 mg of i.v. cyclophosphamide was administered on day 12, followed by 400 mg administered every 2 weeks. She also experienced relief from neuropsychiatric systemic lupus erythematosus (NPSLE) on day 22, which manifested with visual hallucination and persecutory delusion

and was relieved on day 25 by plasmapheresis and i.v. injection of immunoglobulin.

After 6 weeks of treatment, her inflammatory marker levels had improved, and her abdominal pain was reduced to mild levels. A follow-up CT scan (Figure 2) taken at day 51 indicated



**Figure 1.** Pretreatment computed tomography (CT) imaging in a 28-year-old woman with a flare of systematic lupus erythematosus and abdominal pain. CT scan showing RSH (yellow arrow) that measured 21.4×4.7 cm, massive ascites, and edema of the abdomen wall and intestinal wall (before treatment).

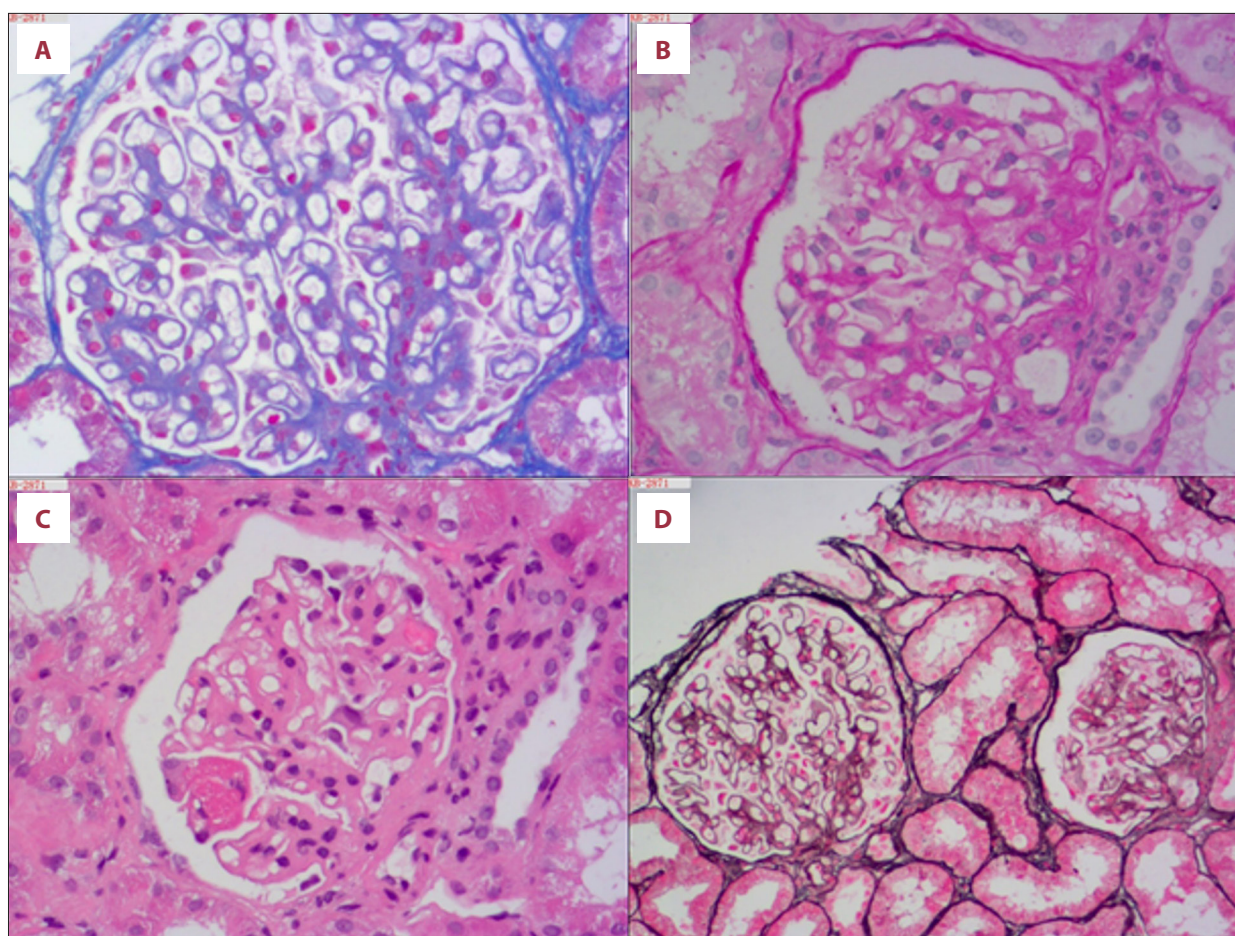


**Figure 2.** Posttreatment computed tomography (CT) imaging in a 28-year-old woman with a flare of systematic lupus erythematosus and abdominal pain. Posttreatment CT scan showed a reduction in the right RSH (yellow arrow) that measured 8.8×1.6 cm.

a reduction that was more than 50% in size of the right RSH, and there was no need for a further procedure. The daily dose of i.v. methylprednisolone was reduced from 80 mg to 24 mg at the rate of 8 mg per week, and oral cyclosporine treatment was commenced (50 mg twice a day) from day 49. The total cyclophosphamide dose reached 2.2 g over the 2-month hospitalization period. Puncture and drainage of ascites were performed on day 75 after her condition improved. The drained fluid was turbid and milky white in appearance and had a volume of 800 mL, and it was positive for the chylous test and Rivalta test, with a protein concentration of 3.6 g/L and a lactate dehydrogenase level of 40 U/L. The nucleated cell count

was  $218 \times 10^6/L$ , with 72% monocytes. The massive chylous ascites gradually disappeared during 1 month of drainage. The patient underwent an ultrasound-guided renal biopsy on day 76. Of the 14 glomerulus samples obtained, 1 was sclerotic and 1 showed segmental sclerosis. The glomeruli displayed mild to moderate segmental mesangial hypercellularity and matrix proliferation, with thickening of the glomerular basement membrane (**Figure 3**). The thickness of the glomerular basement membrane was 1500 nm, as demonstrated by electron microscopy. Immunofluorescence analysis revealed a full-house pattern (**Figure 4**). The patient was diagnosed with class III (A/C) and class V lupus nephritis according to ISN/RPS





**Figure 3.** Photomicrograph of the histopathology analysis of the renal biopsy findings in a 28-year-old woman with a flare of systemic lupus erythematosus. (A) The renal glomeruli show mild to moderate segmental mesangial hypercellularity, increased mesangial cells, and thickening of the glomerular basement membrane. Periodic acid-Schiff. Magnification  $\times 100$ . (B) Thickening of the glomerular basement membrane, segmental and mesangial capillary wall necrosis is present with mild tubular atrophy. Masson's trichrome. Magnification  $\times 60$ . (C) The renal interstitium shows fibrosis and infiltration with mononuclear cells. Hematoxylin and eosin. Magnification  $\times 60$ . (D) Silver staining shows thickening of the tubular and glomerular basement membrane. Methenamine-silver. Magnification  $\times 40$ . These histological features are consistent with SLE membranoproliferative glomerulonephritis, Class III and Class V.

2003 classification [8] and was discharged on day 110 after admission; she was given a prescription of methylprednisolone and cyclosporine (Figure 5). On day 216, her urine protein level had fallen to 1.13 g/day (Table 1).

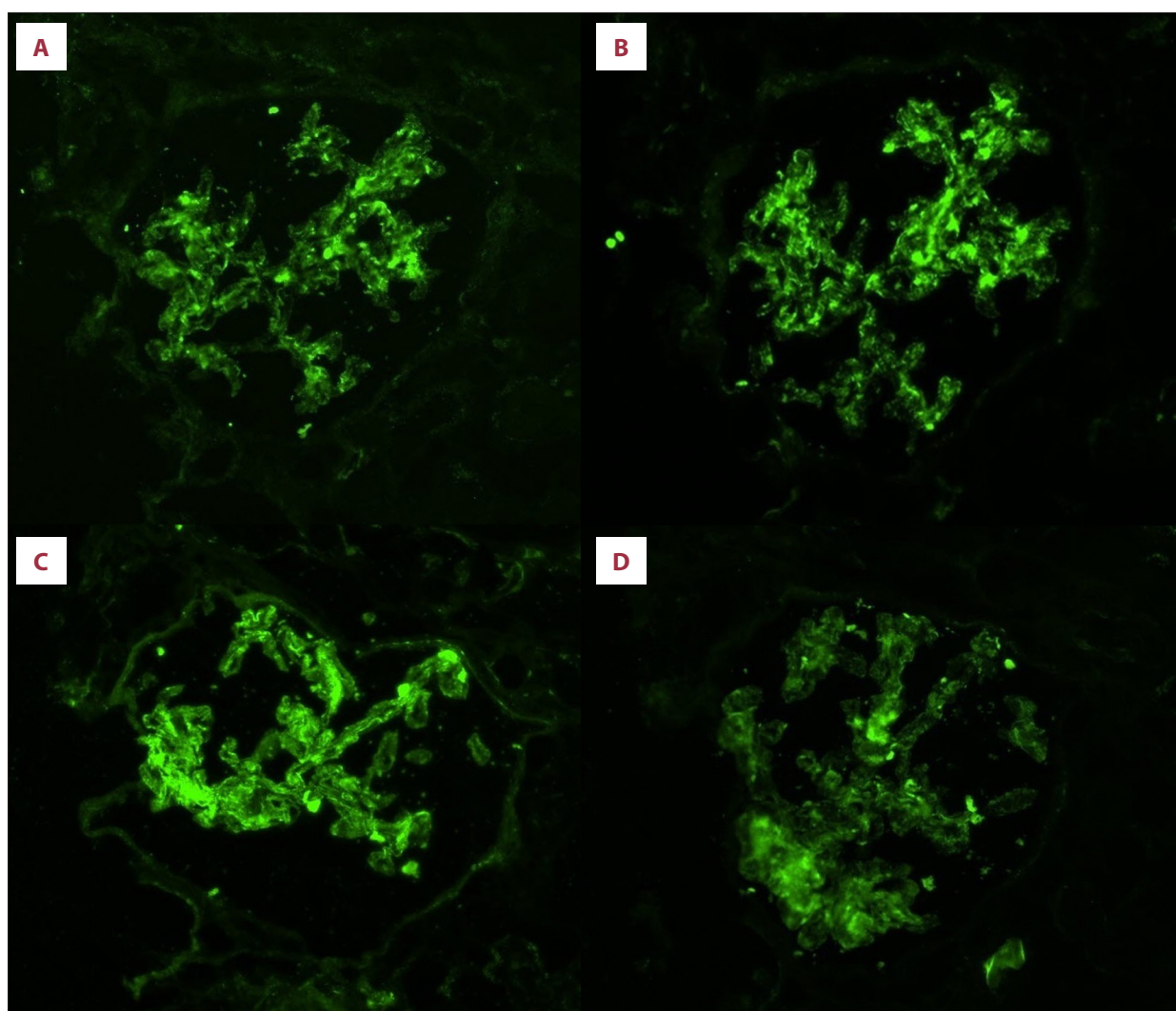
We have received the informed consent of the patient for the publication of this report.

## Discussion

Patients with SLE often experience disease flares of varying severity [9]. Prompt and accurate SLE flare assessment is central in routine clinical practice [2]. The present paper reports a rare onset case of RSH associated with SLE flare, in light of

proteinuria, hematuria, cylindruria, pyuria, leukopenia, pleurisy, and NPSLE in the course of the disease. Although risk factors for RSH such as anticoagulant use and history of prior surgery exist in this case, we propose that the massive ascites contributed to increasing intra-abdominal pressure in the setting of the SLE flare and thus to the development of RSH.

The mechanism of RSH is not completely understood and is believed to be caused by several factors. Anatomy plays a significant role in the formation of RSH. The posterior surface of the rectus abdominis muscle under the arcuate line is covered only by the relatively weak transversalis fascia and the peritoneum [10], and the inferior epigastric arteries are more likely to be injured under the arcuate line without the protection of the rectus sheath. Other risk factors include direct abdominal

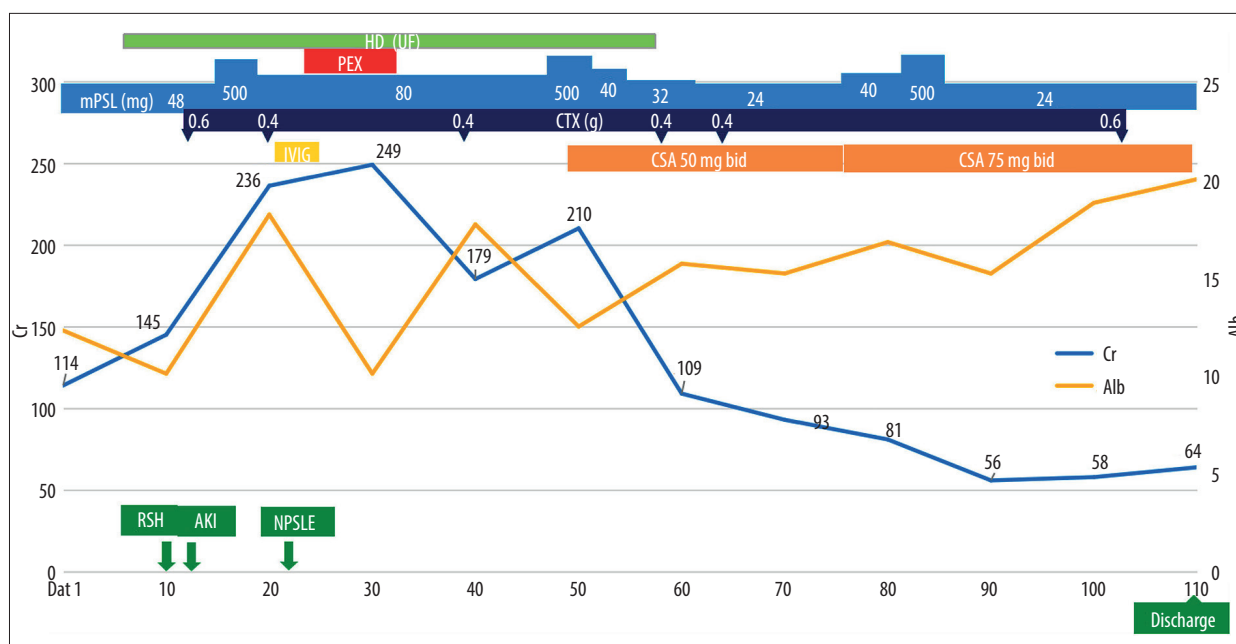


**Figure 4.** Immunofluorescence shows full house positivity for immunoglobulin (IgG, IgA) and complements (C3, C1q). (A) C1q granular deposits in the mesangial region and capillary loops. (B) C3 diffuse and granular deposits in the mesangial area, and segmental positivity along with the capillary loops. (C) IgG granular and segmental deposits in the mesangial region and capillary loops. (D) IgA granular deposits in the mesangial region.

trauma, previous abdominal surgery, the use of anticoagulation therapy, coughing or intense rectus muscle contraction, pregnancy, and infection [5-7,11].

Tutak et al first reported a grade III spontaneous RSH (10×16 cm) concomitant with SLE in a 78-year-old man and used a low dose of anticoagulant for thromboembolism prophylaxis briefly; however, SLE was considered responsible for causing the bleed [5]. Our case was a 28-year old woman with an SLE flare involved in major organs, including cytopenias, neuropsychiatric lupus, and nephritis. The serum C3 was lower than in the previously reported case and anti-double stranded DNA (anti-dsDNA) was negative. The anti-dsDNA negative status seems to identify another subset of patients with peculiar clinical features [12], in particular, serositis. The risks associated

with disease flares were withdrawal of maintenance immunosuppressive treatment and discontinuation of hydroxychloroquine [9]. On the CT scan of our patient, RSH was more giant (21.4×4.7 cm) than the previously reported case and was designated as type II hematoma because it was within the rectus muscle and between the muscle and transversalis fascia [13]. Surgery was not considered in either case, as hemodynamics were not impaired. In the present case, multiple factors may have contributed to RSH. First, the SLE flare may have increased susceptibility, as it is a state associated with complement activation, circulating immune complex formation, in situ immune complex deposition, vascular injury, and, eventually, increased capillary permeability [10]. Second, the patient had been receiving anticoagulation therapy for both venous thromboembolism prophylaxis and intradialytic anticoagulation. Third, as



**Figure 5.** Clinical course in a 28-year-old woman with a flare of systemic lupus erythematosus and abdominal pain. Cr – serum creatine level; Alb – albumin; HD – hemodialysis; PEX – plasma exchange; mPSL – methylprednisolone; CTX – cyclophosphamide; CsA – cyclosporin; RSH – rectus sheath hematoma; AKI – acute kidney injury; NPSLE – neuropsychiatric systemic lupus erythematosus; mg – milligram; g – gram; IVIG – intravenous injection of immunoglobulin; bid – twice a day.

is common during a flare of SLE [14], her platelet count was mildly reduced. Fourth, the patient had severe hypoproteinemia and massive ascites that could lead to increased abdominal pressure and susceptibility to RSH. Ascites is rare in SLE, even rarer in the chylous form. The mechanism of chylous ascites in SLE remains unknown, but one may speculate about lymphatic stenosis or an increase in endoluminal pressure and permeability of vascular walls [15]. Other potential causes of chylous ascites, including tuberculosis, filariasis, liver cirrhosis, neoplastic and traumatic factors, were excluded [16]. Lupus peritonitis was also excluded because of a serum-ascites albumin gradient greater than 1.1 [17]. Finally, coughing and difficulty in defecation were probably the causes in the present RSH [13,18]. This case highlights the importance of simple measures, such as aperients, to reduce the need for abdominal straining in patients with marked ascites, hypoproteinemia, and an inflammatory state.

Management of RSH depends on the patient's clinical status and the underlying cause [6]. Patients who are hemodynamically stable without any changes in their serum hemoglobin are usually closely observed and treated with analgesics if they experience pain [13]. Additionally, anticoagulation therapy is temporarily discontinued, when appropriate [13]. Attention to

ensuring regular and soft consistency of stool is also important to avoid the risk of further bleeding [19]. A supportive blood transfusion may be indicated and, in rare cases, angio-embolization of the bleeding vessel may be required for patients who are in hemorrhagic shock or have a marked decrease in hemoglobin and ongoing bleeding [18]. In our case, the control of the SLE flare with glucocorticoids and immunosuppressants was important, as this alleviated the inflammatory syndrome and associated metabolic derangements that may have contributed to this complication.

## Conclusions

This case has shown that spontaneous RSH can occur without a history of trauma in a patient with an exacerbation of SLE. This association appears to be rare, and the cause is unknown.

## Declaration of Figures' Authenticity

All figures submitted have been created by the authors, who confirm that the images are original with no duplication and have not been previously published in whole or in part.



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