CLINICAL PRESENTATION AND MANAGEMENT OF VASCULITIS IN AN ADOLESCENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

PRESENTACIÓN CLÍNICA Y MANEJO DE VASCULITIS EN UN ADOLESCENTE CON LUPUS ERITEMATOSO SISTÉMICO: REPORTE DE UN CASO

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Abstract

Systemic Lupus Erythematosus is a multifaceted autoimmune disease with diverse clinical manifestations, predominantly affecting females. This case report presents the clinical presentation and management of vasculitis in a 16-year-old male with SLE, an atypical demographic for the disease. The study examines the challenges of diagnosing and treating SLE in adolescents. The patient exhibited polyserositis, cutaneous vasculitis, malar rash, and oral ulcers. Diagnostic procedures included clinical evaluation, laboratory tests (ANA, anti-dsDNA, complement levels), and a skin biopsy histopathological examination. Treatment

consisted of hydroxychloroquine, corticosteroids, and immunosuppressive agents. Laboratory findings revealed positive ANA, elevated anti-dsDNA titers, and low complement levels, confirming SLE with associated vasculitis. Histopathology verified skin-limited vasculitis. The patient responded well to the treatment regimen, achieving remission and reducing disease flares. Regular monitoring and a multidisciplinary approach were pivotal in managing the condition. This case underscores the importance of considering SLE in atypical demographics, such as adolescent males. Early diagnosis and comprehensive management are vital for improving outcomes and quality of life. Further research is warranted to address the unique challenges and optimal treatment strategies for pediatric and male SLE patients, emphasizing the need for awareness and tailored therapeutic approaches in managing chronic autoimmune diseases in diverse populations.

Keywords: Systemic Lupus Erythematosus; vasculitis; autoimmune disease; cutaneous manifestations

Resumen

El Lupus Eritematoso Sistémico es una enfermedad autoinmune multifacética con diversas manifestaciones clínicas que afecta predominantemente a mujeres. Este informe de caso presenta la presentación clínica y el tratamiento de vasculitis en un varón de 16 años con LES, un grupo demográfico atípico para la enfermedad. El estudio examina los desafíos de diagnosticar y tratar el LES en adolescentes. El paciente presentó poliserositis, vasculitis cutánea, erupción malar y úlceras orales. Los procedimientos diagnósticos incluyeron evaluación clínica, pruebas de laboratorio (ANA, anti-ADNds, niveles de complemento) y examen histopatológico con biopsia de piel. El tratamiento consistió en hidroxicloroquina, corticosteroides y agentes inmunosupresores. Los hallazgos de laboratorio revelaron ANA positivos, títulos elevados de anti-ADNds y niveles bajos de complemento, lo que confirma LES con vasculitis asociada. La histopatología comprobó vasculitis limitada a la piel. El paciente respondió bien al régimen de tratamiento, logrando la remisión y reduciendo los brotes de la enfermedad. El seguimiento regular y un enfoque multidisciplinario fueron fundamentales para controlar la afección. Este caso subraya la importancia de considerar el LES en grupos demográficos atípicos, como los varones adolescentes. El diagnóstico temprano y el tratamiento integral son vitales para mejorar los resultados y la calidad de vida. Se justifica realizar más investigaciones para abordar los desafíos únicos y las estrategias de tratamiento óptimas para pacientes pediátricos y masculinos con LES, enfatizando la necesidad de concientización y enfoques terapéuticos personalizados en el manejo de enfermedades autoinmunes crónicas en poblaciones diversas.

Palabras clave: *Lupus Eritematoso Sistémico; vasculitis; enfermedad autoinmune; manifestaciones cutáneas*

Fecha de recibido: 16/10/2024
Fecha de aceptado: 02/12/2024
Fecha de publicado: 09/12/2024

Introduction

Systemic Lupus Erythematosus (SLE) is a multifactorial autoimmune disease characterized by a wide array of clinical manifestations that can involve one or more organs (1). The incidence of SLE varies from 0.3 to 31.5 cases per 100,000 persons per year (2), with a global prevalence ranging between 50 and 100 per 100,000 adults. Notably, SLE predominantly affects females, with nearly 10 female patients for every male diagnosed with the disease.

In 2019, the European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) established new classification criteria for SLE that include antinuclear antibody (ANA) positivity. Vasculitis, an inflammation of the blood vessels, occurs in 11% to 20% of SLE patients (3). Despite its significance in the systemic expression of SLE, the clinical characteristics of vasculitis in SLE patients have not been extensively studied (2). Additionally, ANCA-negative AAVs represent a subgroup with clinical and histological features of AAV but without associated antibodies, often showing limited or less severe systemic renal involvement.

The diagnosis and management of SLE in adolescents present unique challenges due to the variability in clinical presentation and the need for tailored therapeutic strategies. Adolescents, especially males, are an atypical demographic for SLE, which complicates the diagnostic process and may delay appropriate treatment. This case report presents the clinical presentation and management of a 16-year-old male with SLE and associated vasculitis, highlighting the importance of considering SLE in atypical demographics and the need for early diagnosis and comprehensive management to improve patient outcomes and quality of life (4). In this report, we present the case of a 16-year-old male who presented with polyserositis and cutaneous vasculitis. Based on current literature, we also discuss SLE's clinical and immunological characteristics in male patients, highlighting the unique challenges in diagnosing and managing SLE in this demographic.

Materials and methods

The study has a qualitative and analytical approach through the presentation of a case study of a 16-year-old adolescent with Systemic Lupus Erythematosus. The study protocol was reviewed and approved by the Institutional Review Board of the Ambato Regional Hospital (Approval Number: HR2023-00124). All procedures performed in this study involving the patient followed the ethical standards of the institutional and national research committee, along with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. Informed consent to participate in this study was obtained from the patient and her legal guardian. The patient and her legal guardian received complete information about the study's objectives, procedures, potential risks, and benefits. They were assured that participation was voluntary and that they could withdraw from the study without consequences for their medical care.

This study was conducted following the guidelines of the Declaration of Helsinki and was approved by the Institutional Review Board of the Hospital Regional Ambato. The patient provided written informed consent to publish this case report and the accompanying images. For patient privacy reasons, the data used in this study are not publicly available. Data sets used during this study are available from the corresponding author upon reasonable request.

Results and discussion

The patient is a 16-year-old male, born and resident of San Miguel de Guaranda, Ecuador. He is a student with no significant personal or family medical history. The patient denies any toxic habits, such as smoking or drug use. The patient presented with symptoms that had evolved over five months, becoming more apparent in the last five days. The primary symptoms included pain and edema in the carpal joints, proximal interphalangeal joints, knees, ankles, and hips. Morning stiffness lasting less than 30 minutes, causing functional limitations. Facial edema, diffuse thinning, hair fragility, non-painful oral lesions, generalized weakness (asthenia).

The initial physical examination recorded the following vital signs and observations: Blood pressure: 117/78 mm. Hg, Heart rate: 100 beats per minute; Respiratory rate: 20 breaths per minute; Oxygen saturation: 89% (FiO2 21%) and Body temperature: 36.5 degrees Celsius (See Table 1)

Table 1. summarizes the physical examination data.

Blood pressure	Heart rate	Respiratory rate	Saturation	Temperature
117/78 mmHg	100 beats per minute	20 breaths per minute	89% FiO2 21%,	36.5 degrees Celsius

Skin: Pale, dry, desquamative skin; diffuse alopecia; bilateral palpebral edema; malar erythema in a butterfly pattern, respecting the nasolabial fold. Figure 1 shows an adolescent with systemic lupus erythematosus (SLE) and vasculitis. The photo shows a person with pale, dry, desquamative skin. Head: diffuse alopecia. Bilateral palpebral edema and malar erythema in butterfly wings that respect the nasolabial fold are characteristics of SLE. The eyes are covered to protect the patient's identity. The clinical context suggests a dermatologic manifestation of SLE accompanied by possible signs of vasculitis.

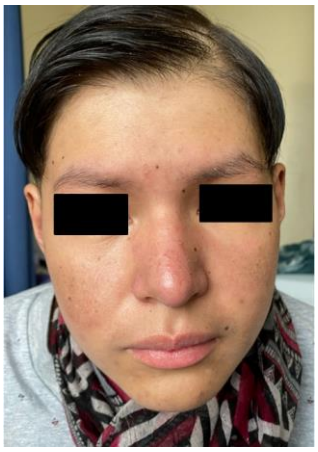

Figure 1. Facial Erythema and Rash in an Adolescent with Systemic Lupus Erythematosus and Vasculitis

Figure 2 shows the open mouth of an adolescent with oral ulcers on the soft palate.

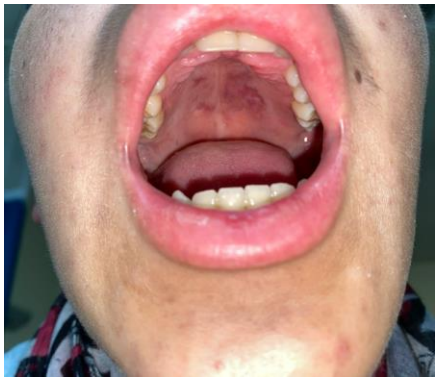

Figure 2. Oral Lesions in an Adolescent with Systemic Lupus Erythematosus and Vasculitis

Figure 3 shows the legs of an adolescent with systemic lupus erythematosus (SLE) and vasculitis. The lower extremities have numerous scattered skin lesions, seen as palpable purpura. These lesions indicate the presence of vasculitis, which is an inflammation of the blood vessels. The lesions are more concentrated in the lower areas of the legs and ankles.

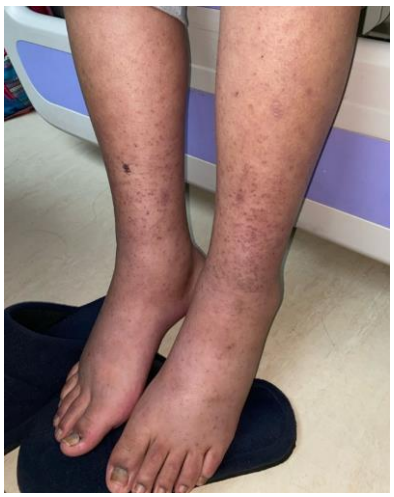

Figure 3. Lower Extremity Purpura in an Adolescent with Systemic Lupus Erythematosus and Vasculitis

Abdomen: no visceromegaly, fluid sounds present, no appendicular or peritoneal signs. Chest: decreased expansibility at bases. Heart: hypophonetic heart sounds. Pulmonary: Vesicular murmur decreased at bases. Table 2 presents the results of complementary tests showing anemia with low hemoglobin (8.8 g/dL) and reduced hematocrit (27.5%). Triglyceride levels are elevated (347.1 mg/dL), and significant liver dysfunction is observed with highly elevated TGO and TGP levels (795.7 U/L and 230.2 U/L, respectively). The immunological analysis reveals the presence of positive ANA (1:1280), elevated anti-double-stranded DNA (>200 IU/mL), and reduced complement C3 (49.13 mg/dL). The 24-hour proteinuria is markedly high (4388.40 mg/24 hours), and the urinalysis shows the presence of granular casts. TSH levels are increased (13 uUI/mL), indicating possible thyroid dysfunction.

Table 2. Results of complementary tests.

Hemogram		
Leukocytes	6.05 10^3/µL	4 - 10
Hematocrit	27.5 %	45 - 55
Hemoglobin	8.8 g/dL	14.5 - 18.5
MCV	93.4 µm³	80 - 100
Platelets	128 10^3/µL	150 - 450
Blood chemistry		
Urea	51 mg/dl	10 - 50
Creatinine	0.84 mg/dl	0.8 - 1.3
Total bilirubin	0.36 mg/dl	0 - 1.2
TGO	795.7 U/L	0 - 38
TGP	230.2 U/L	0 - 42
Triglycerides	347.1 mg/dL	0 - 150
Direct COOMBS	Positive	
Immunological		
ANA	1:1280 Positive	
Anti-DNA C Double	>200 IU/Ml	Positive > 20
Lupus Anticoagulant La	1: 30.50	31-44
Complement C3	49.13mg/dl	90-100
Complement C4	16. 75 mg/dl	9-36
Anti-Cardiolipin Igm	0.5 MPL-UmL	Positive > 25
ANCA C	1.8 U/mL	Positive >5
ANCA P	1.3 U/mL	Positive >5
Urinalysis	granular casts 1-4	
Proteinuria in 24 hours	4388.40 mg/24 hours	28 - 141
TSH	13 uUI/mL	0.35 - 5.1
FT4	0.72 ng/dl	0.5 - 1.4

HBsAg and HCV negatives, Serology for HIV and VDRL: Non-reactive. Abdominal ultrasound: splenomegaly, right pleural effusion. Chest X-ray: scant bilateral pleural effusion. Echocardiogram: preserved left ventricular systolic function LVEF 65%. Histopathology of skin biopsy of the right leg confirmed vasculitis limited to the skin. The diagnosis of systemic lupus erythematosus was made according to the diagnostic criteria used by the American College of Rheumatology. And a SLEDAI lupus activity scale of 40 points, in addition to cutaneous vasculitis and hypothyroidism.

As an induction of treatment, he received pulses of corticosteroids with 1g of methylprednisolone succinate in 2 hours for 3 consecutive days, followed by oral prednisone. As maintenance therapy, he received mycophenolate mofetil 500mg every 8 hours and hydroxychloroquine 200mg per day, prednisone 30mg with weaning. progressive, losartan 50mg as antiproteinuric. After a month he went to follow-up, asymptomatic, with a Hemogram: Leukocytes 10040, Hemoglobin: 12.4 Hematocrit: 39, Platelets: 331000, Glucose: 89 Creatinine: 0.72 mg/dl, TGO: 17 U/L, TGP: 17 U/L, Proteinuria in 24 hours 2084 U/L with SLDAI 8points.

Figure 4 shows an adolescent with systemic lupus erythematosus (SLE) and vasculitis. In the photo, a person with improvement of malar erythema and alopecia.

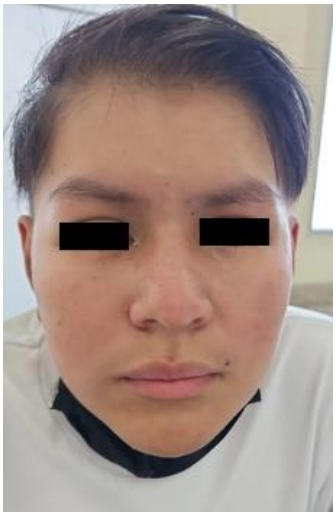

Figure 4. An adolescent with an improvement of malar erythema and alopecia.

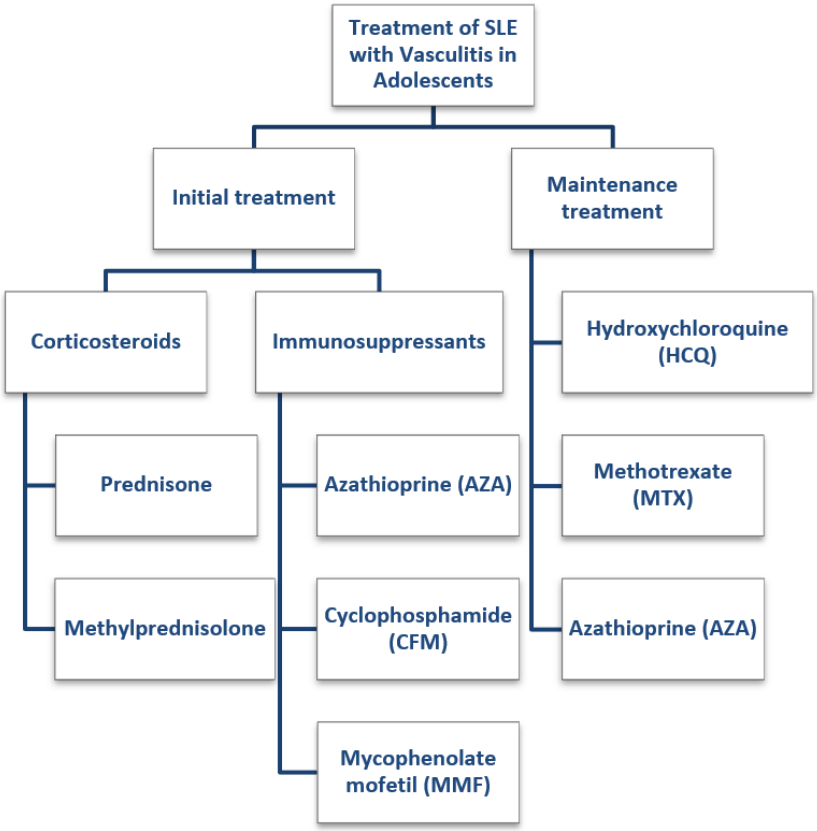

Figure 5. SLE treatment regimen with vasculitis

This case highlights the unusual presentation of SLE in a male adolescent, which is atypical given that the disease predominantly affects females in a 9:1 ratio. This uniqueness underscores the importance of considering SLE in differential diagnoses for young male patients with compatible symptoms (5), (6). The clinical presentation and management of SLE with vasculitis in adolescents pose significant diagnostic and therapeutic challenges. Variability in clinical manifestations can delay proper diagnosis and, thus, initiation of optimal treatment, which is crucial to prevent complications and improve long-term outcomes (2), (7). The reason lupus occurs predominantly in women is difficult to explain. Sex-related genes and hormones may be essential in etiopathogenesis (7). The diagnosis is usually made in young women between 20 and 40 years of age; however, it can begin at any age and will be classified as a juvenile (jSLE) when it begins before the age of 16 years (8), (9).

Vasculitis is an inflammation of the vessel walls. Although it is a characteristic process involved in the systemic expression of SLE, few studies have specifically analyzed the clinical characteristics of vasculitis in patients with SLE. Most studies that analyze the prevalence of vasculitis in large series of patients with SLE show a prevalence that ranges between 11% and 20%, 68 (89%) women and 8 (11%) men (6). Within the broad spectrum of clinical manifestations of SLE, musculoskeletal involvement is reported as the most

frequent (80-95%), followed by skin involvement in 60-85% of cases, the latter being the first sign of the disease in 23-28% of cases (10).

Among the specific symptoms are fever, a wide variety of cardiac and pulmonary pathology; nephropathy, a common complication of SLE that affects 50% of patients; digestive manifestations skin manifestations; and arthritis, 50% of patients with SLE present anemia (11). Newer classification criteria allow earlier classification of SLE, and the combination of the three (ACR-1997, SLICC-2012, and EULAR/ACR-2019) warrants diagnosis of patients. A positive ANA or other immunological (autoantibody or hypocomplementemia) is required to classify SLE according to SLICC-2012 and EULAR/ACR-2019 criteria (12), (13).

ANCA-negative AAVs are a subgroup with clinical and histological characteristics of AAV but with adverse antibody reports. The literature describes these patients' apparent limited or less severe systemic renal involvement (4). The treatment of SLE must be individualized and will depend on the type of manifestation, the organ(s) or systems involved, and the severity of the disease. Severe manifestations include involvement of a major organ and endanger life or function, risk of chronic damage with significant organic sequelae (e.g., lupus glomerulonephritis, severe neurological condition, pulmonary hemorrhage, vasculitis, lupus bullous. These manifestations may be treated with high-dose GC or boluses of cyclophosphamide or, mycophenolic acid or other immunosuppressants (14).

Treatment goals include long-term patient survival, organ damage prevention, and quality of life improvement. Therapy should achieve remission or at least low disease activity and prevention of flares. All patients with lupus should receive hydroxychloroquine at a dose not to exceed 5 mg/kg body weight. During chronic maintenance therapy, glucocorticoids should be minimized to less than 7.5 mg/day (prednisone equivalent) and, when possible, withdrawn. Appropriate initiation of immunomodulatory agents (methotrexate, azathioprine, mycophenolate) may accelerate the gradual reduction or discontinuation of glucocorticoids. In persistently active or exacerbated disease, the addition of belimumab should be considered; Rituximab or cyclophosphamide (CY) may be considered in refractory organ-threatening disease (2).

In the past two years, several new therapies targeting different molecular pathways have shown encouraging results in clinical trials in SLE. After the approval of belimumab, trials with fully humanized B-cell-targeted monoclonals or combinations of biologics were initiated. A second group of drugs targets interferon, either directly, such as nivolumab and sarilumab, or more indirectly, such as BIIB059 or omalizumab (5). Figure 5 shows the treatment options for adolescents with systemic lupus erythematosus (SLE) and vasculitis, dividing them into initial and maintenance treatments and specifying the medications used in each category.

Conclusions

This case report highlights the complexity of diagnosing and managing systemic lupus erythematosus (SLE) with vasculitis in a male adolescent, an unusual demographic for this predominantly female disease. The patient's presentation with polyserositis, cutaneous vasculitis, and typical features of SLE, such as malar rash and oral ulcers, underscores the diverse clinical manifestations of SLE. The diagnostic process was based on clinical evaluation, laboratory findings, and imaging studies. The presence of positive antinuclear antibodies

(ANA), high titers of anti-double-stranded DNA (dsDNA) antibodies, and low complement levels (C3 and C4) were crucial to confirm the diagnosis of SLE. Skin biopsy confirmed the presence of vasculitis, highlighting the importance of histopathologic examination in diagnosing systemic involvement.

Management of SLE in this patient focused on controlling disease activity and preventing organ damage. Hydroxychloroquine, corticosteroids, and immunosuppressive agents were adjusted to achieve remission and reduce the risk of flares. This case also illustrates the importance of regular monitoring and multidisciplinary care in managing chronic autoimmune diseases such as SLE. Finally, this case underscores the need for greater awareness of SLE and its potential complications in adolescents, including males, who may present with atypical manifestations. Early diagnosis and comprehensive management are essential to improve these patients' outcomes and quality of life. More research is needed to explore the unique challenges and optimal treatment strategies for pediatric and male patients with SLE.

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