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Case Report

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Unmasking Vasculitis: An Atypical Presentation of Systemic Lupus Erythematosus with Fascial Rash

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Abstract

Background

Systemic lupus erythematosus (SLE) is a complex autoimmune disease with diverse clinical presentations. Rowell syndrome, a rare overlap of SLE and erythema multiforme (EM), presents diagnostic challenges due to its atypical features. It is characterized by EM-like lesions, positive antinuclear antibodies (ANA), and anti-Ro/SSA antibodies.

Case Presentation

A 29-year-old Filipino female developed a four-week history of EM-like rashes, followed by fever, lymphadenopathy, vomiting, and epigastric pain. After ruling out infectious and drug-induced causes, immunological testing revealed ANA >1280 (speckled pattern), elevated anti-Smith and anti-SSA antibodies, and low C3. A skin biopsy confirmed leukocytoclastic vasculitis, consistent with cutaneous lupus. The patient met both the EULAR/ACR 2019 criteria for SLE and Zeitouni's criteria for Rowell syndrome.

Discussion

This case highlights the diagnostic challenges of Rowell syndrome, particularly when EM-like lesions precede systemic symptoms. A thorough workup led to early diagnosis and treatment with hydroxychloroquine and corticosteroids, resulting in clinical improvement.

Conclusion

Rowell syndrome, though rare, should be considered in patients with persistent EM-like lesions and autoimmune markers. Greater awareness among clinicians can facilitate early recognition, timely management, and improved outcomes for patients with this rare subset of SLE.

Keywords: Physical Vapor Deposition (PVD), Coating Equipment, Vacuum Technology, Heating Source Technology, Coating Material

1. Introduction

Systemic lupus erythematosus (SLE) is a complex autoimmune disorder with diverse clinical and immunological manifestations, affecting 5.14 (1.4 to 15.13) per 100,000 person-years and 0.40 million people annually [1]. While it primarily involves the skin, joints, kidneys, and hematological systems, atypical presentations such as Rhupus and other overlap syndromes can complicate diagnosis [2].

Rowell syndrome, first described in 1963, is a rare overlap syndrome combining features of erythema multiforme (EM) and SLE [3]. It is characterized by EM-like rashes, speckled ANA, anti-Ro/SSA and anti-La/SSB antibodies, and rheumatoid factor [4]. Its rarity and variable presentation often delay diagnosis. We report an atypical case of Rowell syndrome, presenting a diagnostic challenge due to its unusual sequence of onset. The patient experienced a 4-week history of an isolated EM-like skin rash, preceding any overt lupus manifestations. This presentation required a thorough

evaluation to exclude viral, infectious, and drug-related causes of EM. Immunological profiling and histopathological analysis via skin biopsy were crucial in confirming the diagnosis.

This case is notable not only for the rarity of Rowell syndrome but also for its atypical disease progression, with EM as the initial and predominant manifestation—contrasting the typical pattern where SLE precedes EM [5]. We aim to raise awareness among clinicians, highlight the importance of systematic diagnostics, and contribute to the evolving understanding of atypical Rowell syndrome presentations. Persistent EM should prompt consideration of autoimmune etiologies beyond isolated dermatological conditions.

2. Case Presentation

We reported a case of 29-year-old Filipino female presented with a four-week history of a progressive maculopapular rash involving her scalp, face Figure (1-2), trunk and back Figure (3), and upper limbs, preceded by a complicated molar extraction. Two weeks later, she developed a high-grade, continuous fever (peaking at 39.5°C), bilateral facial swelling, and shoulder pain. Despite a course of azithromycin and amoxicillin-clavulanate (Augmentin), her fever persisted, accompanied by vomiting, epigastric pain, dry cough, and headache. The patient denied any history of fatigue, fever, photosensitivity, oral or nasal ulcers, malar rash, and alopecia. History should include joint pain or swelling, Raynaud's phenomenon, serositis (pleuritic chest pain or pericarditis), and renal symptoms like hematuria or edema. Neurological symptoms, including headaches, seizures, or cognitive changes, should be noted. Family history of autoimmune diseases and potential triggers like infections, and sun exposure are also relevant. She denied smoking, alcohol use, recent travel, or sick contacts.

On physical examination, the patient was alert, oriented, and appeared well. She was febrile (39.5°C) with a heart rate of 112 bpm, blood pressure of 118/87 mmHg, respiratory rate of 18 bpm, and oxygen saturation of 97% on room air.

Skin examination revealed a widespread maculopapular rash on the scalp, face, trunk, upper limbs, and palms (figure1). Tender submandibular, supraclavicular, and left axillary lymph nodes (<1 cm) were noted. Joint examination showed no deformities, swelling, tenderness, or restricted range of motion. All joints were pain-free with normal movement, and no synovitis, effusion, or crepitus was detected. Head and neck examination showed no pallor, icterus, dry eyes, or throat ulcers. Cardiovascular, respiratory, and neurological examinations were normal. Abdominal exam revealed epigastric and right lower quadrant tenderness without organmegaly or peritoneal signs. Extremities showed no cyanosis, edema, or deep vein thrombosis

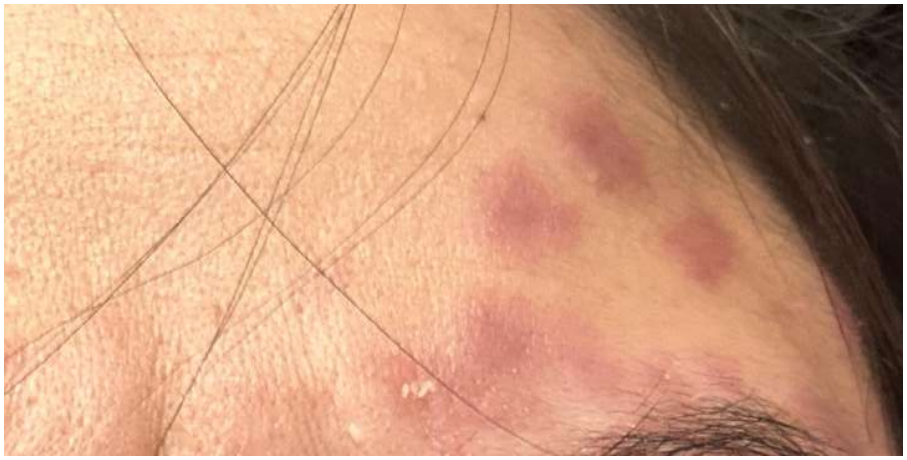

Figure 1: Annular Erythematous Involving the Head

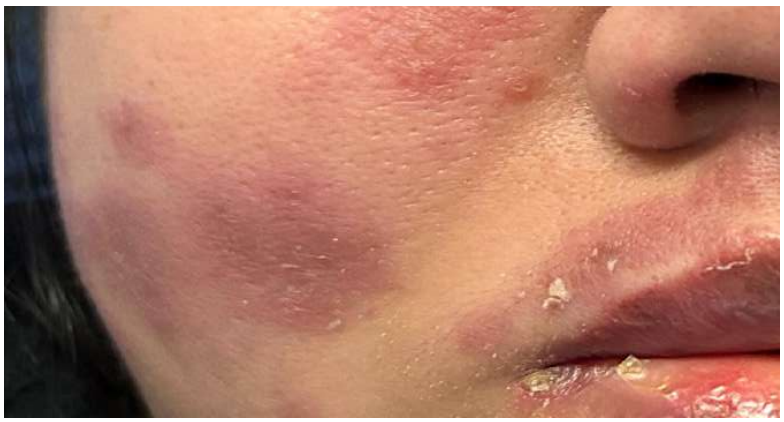

Figure 2: Annular Erythematous Scaly Plaques Involving the Face

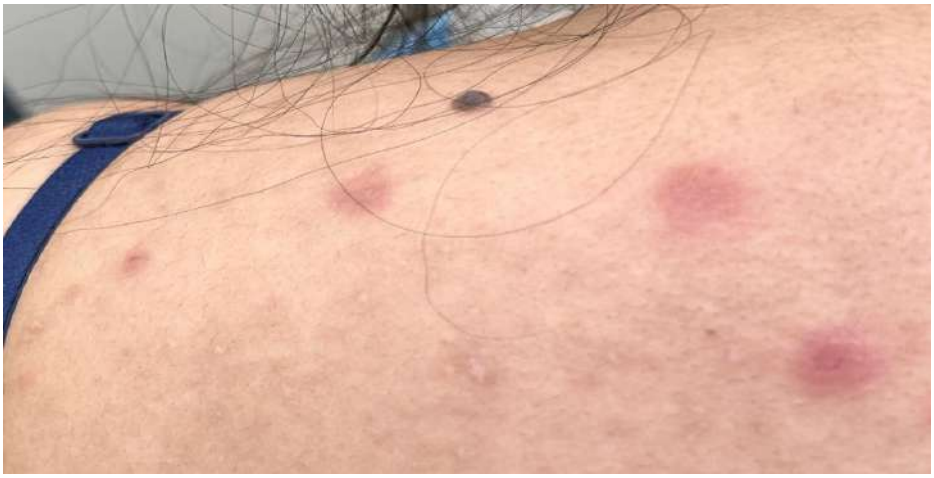

Figure 3: Annular Erythematous Scaly Plaques Involving the Back

All pictures are for educational purposes only and patient were consented and gave a written consent for using her pictures.

3. Workup

The patient underwent an extensive diagnostic workup to determine the cause of her fever and rash, focusing on infectious, autoimmune, and systemic etiologies. Initial laboratory results revealed leukocytosis with neutrophilia and mild normocytic anemia, suggesting inflammation or autoimmune disease. ESR and CRP were significantly elevated, indicating active inflammation, while platelet counts were normal. Liver and renal function tests were unremarkable, ruling out organ involvement.

Microbiological testing, including blood and urine cultures, and virology tests for HIV, Hepatitis A/B/C, EBV, CMV, parvovirus B19, rubella, measles, syphilis, malaria, and brucellosis, all returned negative, excluding common infectious causes.

An autoimmune workup revealed strongly positive ANA with a speckled pattern (titer >1280), elevated anti- Smith antibodies, and positive anti-SSA (Ro) antibodies. Anti-dsDNA antibodies were negative. Complement levels showed mildly decreased C3, suggesting mild complement activation, while C4 was normal. Rheumatoid factor was negative, but anti-RNP antibodies were elevated. The antiphospholipid antibody panel was negative, excluding antiphospholipid syndrome. These findings strongly supported a diagnosis of SLE.

Imaging studies included a chest X-ray, which showed no abnormalities, and a CT scan of the chest, abdomen, and pelvis, revealing left axillary lymphadenopathy. A non-contrast CT scan of the head, following complaints of epistaxis and headache, showed no intracranial abnormalities. Lumbar puncture revealed normal cerebrospinal fluid, excluding CNS infection or inflammation.

To further investigate the patient's rash, a skin biopsy was performed, revealing leukocytoclastic vasculitis (Figure 4), consistent with cutaneous lupus erythematosus and supporting the diagnosis of systemic lupus erythematosus (SLE). The presence of erythema multiforme (EM)-like lesions prompted consideration of Rowell syndrome, a rare overlap condition associated with lupus erythematosus.

Diagnosis was based on the Zeitouni criteria, which require three major criteria and at least one minor criterion. The major criteria include the presence of lupus erythematosus (confirmed through serological and histopathological evidence), EM-like lesions (widespread rash involving the scalp, face, trunk, and upper limbs), and a speckled ANA pattern (titer >1280). The patient met all three major criteria. For minor
criteria, chilblains were absent, but the patient tested positive for anti-Ro (SSA) antibodies, fulfilling one minor criterion. Rheumatoid factor was negative, but this does not exclude the diagnosis, as all major criteria and one minor criterion were met.

This comprehensive workup, combining clinical, serological, and histopathological findings, confirmed SLE with Rowell syndrome, highlighting the utility of the Zeitouni criteria in diagnosing this rare condition. The diagnosis was further corroborated by exclusion of infectious, neoplastic, and other autoimmune causes.

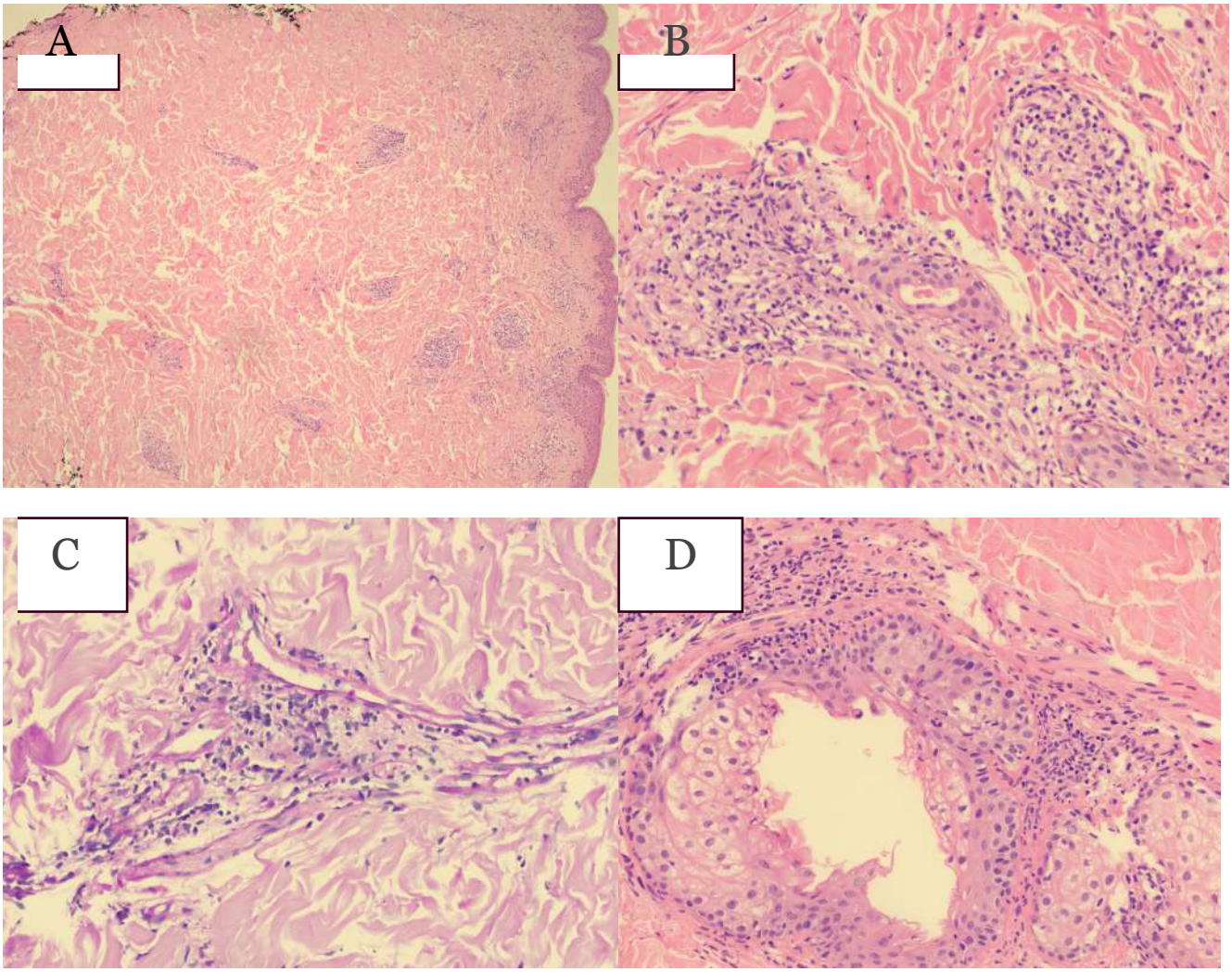

Figure 4: Legend: Skin Biopsy Showing

- Superficial and deep perivascular dermal inflammatory cell infiltration (H&E, 4x).
- The infiltration is composed mainly of neutrophils with nuclear dust ((H&E, 20x).
- Transmural neutrophilic infiltration with fibrin deposition in the vessel wall (PAS, 20x).
- Periadnexal infiltrates show karyorrhexis and fragmentation of neutrophils expressed as nuclear dust (H&E, 20x).

The minor criteria for Rowell syndrome include chilblains, positive anti-Ro (SSA) or anti-La (SSB) antibodies, and rheumatoid factor (RF) positivity. Although the patient did not have chilblains, she tested positive for anti-Ro (SSA) antibodies, fulfilling one minor criterion. Rheumatoid factor was negative, but its absence does not exclude the diagnosis, as all major criteria and one minor criterion were met.

This comprehensive workup, incorporating clinical, serological, and histopathological findings, confirmed the diagnosis of systemic lupus erythematosus with Rowell syndrome. The application of the Zeitouni criteria was instrumental in identifying this rare entity. The diagnosis was further solidified by systematically excluding infectious, neoplastic, and other autoimmune conditions through targeted investigations.

4. Discussion

Systemic lupus erythematosus (SLE) is a complex autoimmune disease, often presenting with diverse clinical features. The patient in this case met the criteria for SLE, with a cumulative score of 18, exceeding the 10-point threshold for diagnosis, driven by constitutional symptoms, hematologic abnormalities, mucocutaneous manifestations, and SLE-specific antibodies. The 2019 EULAR/ACR classification criteria were used to confirm the diagnosis, with ANA positivity being a key criterion.

Rowell syndrome (RS) is a rare overlap of SLE and erythema multiforme (EM), characterized by EM-like lesions, lupus erythematosus, and specific immunological findings (positive ANA, anti-Ro/La antibodies, and rheumatoid factor). Using Zeitouni's criteria, the patient fulfilled all major criteria: confirmed SLE, EM-like rash, and ANA titer >1280. Anti-Ro (SSA) positivity fulfilled one minor criterion, supporting the diagnosis despite a negative rheumatoid factor.

The concept of Rowell syndrome remains debated, with some suggesting it may be a distinct form of subacute cutaneous lupus. Clinical differentiation between EM and subacute cutaneous lupus can be challenging.

5. Conclusion

SLE is often difficult to diagnose, especially in the presence of rare overlaps like Rowell syndrome. This case highlights the importance of considering EM-like lesions in diagnosing SLE and the value of a thorough diagnostic approach. Timely treatment with hydroxychloroquine and corticosteroids resulted in significant improvement, emphasizing the need for awareness and management of atypical SLE presentations [6].

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