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Systemic Lupus with Erythema Multiforme–so called Rowell's Syndrome

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Abstract

The combination of lupus erythematosus and erythema multiforme has been grouped as Rowell's syndrome. Though there are reports of so called 'Rowell's syndrome' (RS) in the literature, recentlymany authors have questioned the rationale behind regarding Rowell's syndrome as a separate clinical entity. Nevertheless, we describe a 20 year old female patient with features of systemic lupus erythematosus, lupus nephritis, and erythema multiforme. Laboratory investigations showed speckled antinuclear antibody, positive anti-dsDNA and rheumatoid factor positivity. Skin lesions healed with hyperpigmentation within four weeks of therapy.

Keywords: *Rowell's syndrome, Erythema Multiforme, systemic lupus.*

1. Introduction

Rowell Syndrome (RS) is a unique clinical association of lupus erythematosus (LE) with erythema multiforme (EM) like lesions and a characteristic immunologic pattern.[1]We describe a patient whoseclinical picture is consistent with so-called*Rowell*'s syndrome.

Case Report

A 20 year old female presented with history of rash of 15 days duration in both her arms and both post auricular areas. She also gave history of facial puffiness, and pedal edema. There was no history of hematuria, joint pains, facial rash, raynaud's phenomenon, chilblains, oral and genital ulcers. On evaluation she had periorbital edema and pedal edema. There were erythematous targetoid lesions in the lateral aspects of both her arms and both post auricular areas suggestive of erythema multiforme (Figure 1 &2). There were no mucosal lesions. There was no prior history of infection or drug intake.

Laboratory investigations showed hemoglobin 8.2 g/dl, total counts of 3400cells/µl, blood urea 20 mg/dl, serum creatinine 0.7 mg/dl, and serum albumin 3.1g/dl. Urine analysis showed 3+ proteinuria with 6-8 RBcs per hpf. Urine protein creatinine ratio was 4.0 and 24 hour urinary protein excretion was 3.6 grams per day. Peripheral smear showed microcytic hypochromic anemia. ANA was positive with speckled pattern and anti – ds DNAwas positive. Anti – Ro and anti – La antibodies were negative and Rheumatoid factor was positive. Complement levels (C3, C4) were low.

In view of proteinuria, renal biopsy was done. Light microscopy showed diffuse and global endocapillary proliferation along with neutrophilic infiltrates and necrosis in few glomeruli (Figure 3). Active cellular crescents and wire loop lesions were seen in few glomeruli. Immunofluorescence microscopy revealed full house pattern (IgG, IgA, IgM, C3, & C1q) of immune deposits.

Patient was diagnosed to have systemic lupus along with lupus nephritis and erythema multiforme. After discussion with the patient relatives, patient was started on oral mycophenolatemofetil, oral steroids, hydroxychloroquine and ACE inhibitors. One month after treatment, the skin lesions healed with residual hyperpigmentation.

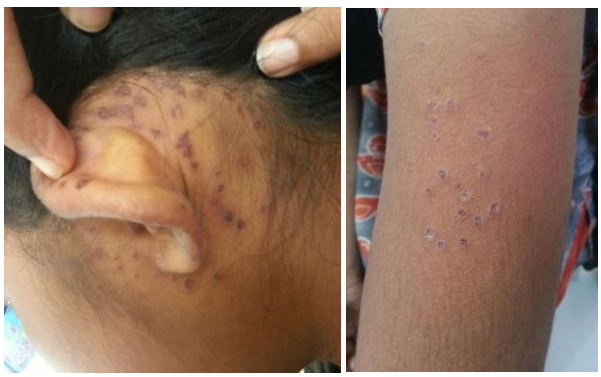

Figure 1 and 2: Erythematous targetoid lesions in the lateral aspects of both her arms and both post auricular areas suggestive of erythema multiforme

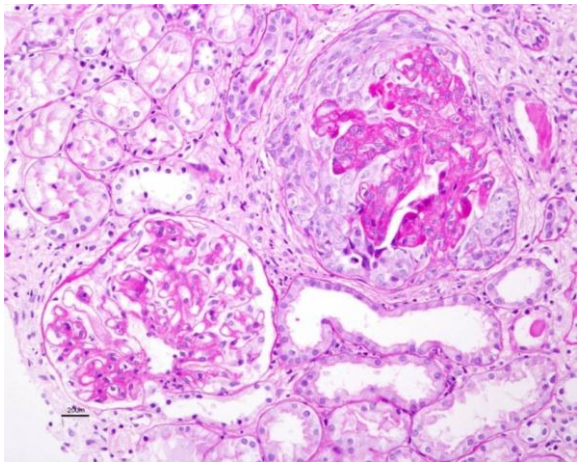

Figure 3: Diffuse and global endocapillary proliferation along with neutrophilic infiltrates and necrosis in few glomeruli

Discussion

In 1963, Rowell et al. described a distinctive subset of patients diagnosed with discoid lupus erythematosus (DLE) associated with Erythema Multiforme (EM) and a characteristic pattern of immunological abnormalities.[4] Zeitouni and coworkers reviewed the diagnostic criteria for RS. The major criteria consist of the presence of lupus erythematosus (systemic, discoid, or subacute lupus), EM-like lesions (with or without involvement of mucous membranes), and speckled pattern of antinuclear antibody. The minor criteria include chilblains, anti-Ro and/or anti-La antibodies, and positive RF. All three major and at least one minor criteria are required to establish the diagnosis of RS.[2]

EM is thought to fall within a spectrum of diseases that affect the skin and mucous membranes, including erythema multiforme, Stevens-Johnson syndrome, and toxic epidermal necrolysis. Erythema multiforme is usually precipitated by infections (e.g., herpes simplex and mycoplasma) and drugs (e.g., penicillin and sulfamide) and is generally not associated with any specific autoimmune abnormality. The skin lesions are often quite targetoid.[3]

Since the original diagnostic criteria were proposed for Rowell syndrome in 1963, over 70 cases have been reported in the literature. However, none of the reported cases entirely conformed to Rowell's original serologic and clinical description.[6]

In majority of the cases, several other types of cutaneous lupus were reported rather than discoid lupus and chilblains as originally described by Rowell et al. Typical raised target lesions of erythema multiforme were reported very rarely, while in several cases the lesions were more consistent with the annular-polycyclic variant of subcutaneous lupus.

Often it is challenging to differentiate erythema multiforme and annular-polycyclic variant of subcutaneous lupus both clinically and histologically. Such misinterpretations have been reported by Mendonca, in which repeat biopsies of lesions previously read as EM showed lupus instead.[7]

With the lack of a consistent and specific serological profile the very existence of Rowell syndrome is questioned. There seems to be enough evidence to classify Rowell's syndrome within the subacute cutaneous lupus erythematosus (SCLE) rather than accepting it as a separate entity.[5]A real association between discoid lupus and erythema multiforme was present only in a minority of cases and could be considered a mere coincidence without any immunological significance.

Our patient was a 20 year old female who presented with features of nephrotic syndrome and targetoid lesions. All three major and one minor criteria were present. Histological diagnosis of erythema multiforme could not be proved as the patient was not willing for skin biopsy.

Conclusion

A more clinically relevant question concern whether RS truly merits distinction as a unique clinical entity. It is quite possible that prior reports of RS may be merely lupus masquerading as EM, and that the true existence of Rowell syndrome remains controversial. However, we describe a patient whose clinical picture was consistent with so-called Rowell's syndrome.

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