Original Research Paper

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Rheumatology

ROWELL'S SYNDROME-A RARE CLINICAL ENTITY

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ABSTRACT Rowell's syndrome is a rare disease consisting of Erythema multiforme like lesions associated with lupus erythematosus.The present case report presents the case of a nineteen year old female with skin eruptions and investigations suggestive of sub acute cutaneous lupus along with EM-like lesions thereby diagnosed as rowell's syndrome

KEYWORDS : Rowells,erythema Multiforme,lupus Erythematosus,anti-ro/anti-la

INTRODUCTION

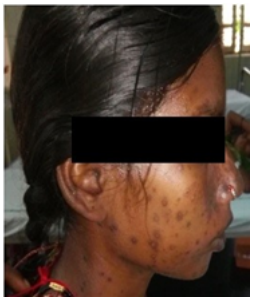
Rowell's syndrome is a rare disease characterized by lupus erythematosus (LE) and erythema multiforme (EM)-like lesions associated with ANA,anti La/ Anti Ro antibodies and rheumatoid factor positivity.Since the original description very few cases have been reported. We describe a patient with lupus erythematosus who developed annular lesions matching those described in original report of Rowells.

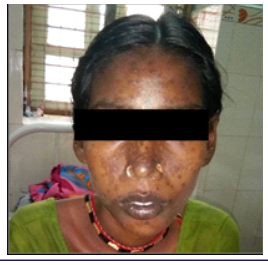
CASE REPORT:

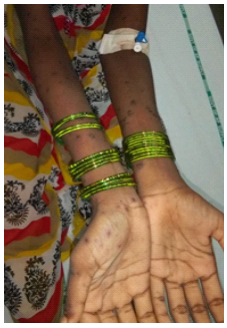
A nineteen year old female presented with complaints of high grade fever,mildly pruritic skin rash, oral ulcers,multiple joint pains,fatigue since one month.There was no history of respiratory tract infection,drug intake or herpetic lesions in the recent past.

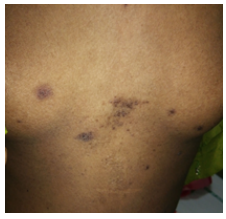
Physical examination revealed non scarring erythematous annular rash over cheeks and nasal bridge, diffuse erythematous papules symmetrically distributed over face predominantly over forehead ,cheeks,chest,lower back,extensor aspects of forearms and palms, along with multiple discrete targetoid lesions predominantly over sun exposed areas ,multiple erosions noted in Buccal mucosa.Vital parameters and systemic examination were normal.

Laboratory investigations showed normocytic normochromic anemia leukopenia,thrombocytopenia.Blood urea and serum creatinine levels were normal.ESR was 50 mm at 1hr.Urine analysis suggestive of mild proteinuria(200mg/d).Immunological profile revealed a positive RF,positive ANA with speckled pattern (titre 1:320), positive anti smith, positive anti Ro antibodies .The patient met the diagnostic criteria for SLE.A diagnosis of Rowell's syndrome was established based on clinical manifestations and positive diagnostic criteria for SLE.









DISCUSSION:

Rowell et al. described a syndrome characterized by lupus erythematosus (LE) and erythema multiforme (EM)-like lesions, positive tests for rheumatoid factor (RF), speckled antinuclear antibody (ANA), and precipitating antibody to saline extract of human tissue (anti-SjT) in 1963.[1].Modifications to original diagnostic criteria have been made due to inconsistent features by Lee et al,Zeitouni et al and Torchia et al.[5]

Lee et al in 1995 suggested the inclusion of chilblains as a diagnostic feature.[3,5]

In 2000,Zeitouni et al proposed the following revised criteria.[2,4]

MAJOR CRITERIA	MINOR CRITERIA
SLE,DLE or SCLE	Chilblains
EM-like lesions	Anti-Ro/anti-La
ANA with speckled pattern	Positive RF
Three major and at least one minor criteria required for diagnosis	

TORCHIA ET AL CRITERIA(2012)

MAJOR CRITERIA	MINOR CRITERIA
Chronic cutaneous LE	No triggering factors
EM-like lesions	Lack of EM lesions on acral or mucosal surfaces
Speckled ANA/anti-Ro/anti-La	Other criteria for SLE excluding discoid or malar rash, ANA ,photosensitivity, oral ulcers or chilblains
Negative DIF on EM like lesions	

All major and one minor criteria required for diagnosis

The non scarring annular and EM- like lesions without follicular plugging distributed in photosensitive areas clinch the diagnosis in this patient as sub acute lupus erythematosus with ROWELLS syndrome. The patient had no precipitating factors for EM like lesions and fit the original and Zeitouni et al crieria.In conclusion, Rowell syndrome, although rare, should be suspected in all patients with LE with EM-like lesions where there is no evidence of a precipitating factor.

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