

ROWELL SYNDROME IN MALE: A RARE CASE REPORT

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ABSTRACT

Rowell's Syndrome [RS] was first described by Rowell and coworkers in 1963. Since then there are very few case reports and some authors have even questioned the existence of RS as distinct entity. This is a rare syndrome in which patients with Systemic lupus Erythematosus [SLE] present with Erythema Multiforme [EM]. SLE presenting only with target lesions of EM is uncommon especially in males as more than 90% of SLE patients are females of child bearing age group. Here we are reporting a rare case of RS in a 30 year old male which will add to the fact that RS is not a mere overlap syndrome but a distinct entity.

KEYWORDS: Rowell Syndrome, Erythema Multiforme, Systemic Lupus Erythematosus.

INTRODUCTION

Rowell syndrome [RS] is a rare entity in which patients with Systemic Lupus Erythematosus [SLE] develop Erythema Multiforme [EM] – like lesions. It was first described in 1963 by Rowell and co-workers as a syndrome characterized by EM like lesions, discoid lupus, positive test for rheumatoid factor [RF], speckled Anti-Nuclear Antibody [ANA] and positive precipitating antibody to saline extract of human tissue [anti-SjT].^[1] Later, Zeitouni and co-workers redefined this syndrome with major [SLE, EM like lesions, speckled pattern of ANA] and minor [Chilblains, positive anti La/ anti Ro antibody, positive RF factor] diagnostic criteria.^[2] To diagnose Rowell Syndrome all three major and at least one minor criteria should be present. Very few cases of RS have been reported till now in literature and most of them have been described in females as SLE is more common in females.³ Here, we are reporting a rare case of 30 year old male diagnosed with RS.

CASE HISTORY

A 30 year old male presented to medicine OPD with chief complaints of intermittent fever recorded upto 100 degree Fahrenheit for two months, pain in both ankle joints for two months, multiple target like erythematous lesions over both upper limbs [below elbow on flexor surface] and anterior abdominal wall for one month. He had history of [h/o] recurrent oral ulcerations and chilblains. There was no h/o chronic cough, genital ulcerations, eye lesions, hair fall, recurrent jaundice,

generalized body swelling, decreased urine output or any alleged drug intake like sulfa drugs, steroids, anticonvulsants, antitubercular drugs or any other antibiotic. There was no h/o any other autoimmune disease or chronic illness in past. On general examination, typical target lesions [Fig.1] were found on flexor surface of both upper limbs (below elbow) and anterior abdominal wall suggestive of EM along with few oral ulcers and mild pallor. His ankle joints were tender but not swollen. His vitals and systemic examination was within normal limits. On investigation following was recorded – Haemoglobin level of 10.6g/dl, total leucocyte count of 9100/cu mm, platelet count of 1.36 lac/cu mm, serum C-reactive protein was moderately raised, serum ANA- 139 U with speckled pattern, serum Anti double stranded DNA – 136 IU/ml, rheumatoid factor was negative, serum creatinine- 1.2mg/dl, serum total protein/albumin- 6.7/3.4 g/dl, serum Na/K- 137/3.5mmol/l, urine microscopy had red cell casts with 4+ proteinuria, 24 hr urinary protein was 3 gm. Urine culture, coagulation profile and Ultrasound abdomen reports were normal. Further, renal biopsy was done which showed features suggestive of lupus nephritis type IV, segmental in nature, predominantly active lesion. On the basis of history, physical examination and biochemical parameters, patient satisfied SLICC⁴ criteria described for SLE. Since our patient had SLE and EM with speckled pattern of ANA and history of chilblains, we diagnosed him as RS based on Zeitouni and coworker's diagnostic criteria and managed him conservatively. Initially oral prednisolone

40 mg was started following which skin lesions gradually improved (Fig 2) within two to three weeks. Renal biopsy report was obtained three weeks following clinical diagnosis. Accordingly IV cyclophosphamide (500 mg/m² once month) was added to the regimen for 6 months after which his biochemical parameters improved and proteinuria reduced. Currently the patient is on maintenance therapy with oral prednisolone (5mg/day) plus azathioprine (100 mg/day) with regular monitoring for side effects.

DISCUSSION

90% of patients diagnosed with SLE are females of child bearing age group.^[4] Most common SLE specific skin lesions are malar rash, photosensitive dermatitis, generalized macula-papular rash, subacute cutaneous lupus erythematous, discoid lupus erythematous.^[5] EM has been documented among only 6% of patients with SLE.⁵ Musculoskeletal manifestations of SLE are mainly intermittent polyarthritis commonly involving hand, feet and knee joint. Our patient was male presenting with persistent pain in ankle joint and target skin lesions [EM like] further leading to diagnostic dilemma. EM is hypersensitivity skin reaction which is usually, either drug induced (barbiturates, phenytoin, penicillin, sulfonamides) or due to infections (herpes simplex, mycoplasma).^[6] SLE presenting with EM is uncommon. Except for target lesions, this male patient had very non specific symptoms. Most important point was that he had presented with EM but without any history of precipitating event. Finally both clinically and biochemically our patient met Zeitouni's criteria for RS. Although treatment for SLE and RS is same but this kind of presentation especially in a male patients very rare leading to diagnostic confusion. Our patient responded well to prednisolone and cyclophosphamide. Now whether RS is either an overlap syndrome or a real association or just a co-incidence is still a debatable issue.^[7] Since very few case reports of RS are available in literature, its still a matter of controversy. A number of case series are needed to end up in a firm conclusion. This case satisfying all diagnostic criteria of RS strongly suggests that it is not a mere coincidence. Rather it is a distinct entity which should be always kept in mind in all patients including males presenting with EM-like lesions where there is no evidence of any precipitating factor. Thus, high index of suspicion is needed to diagnose RS.



Figure-1.



Figure-2.

REFERENCES

1. Rowell NR, Beck JS, Anderson JR. Lupus erythematosus and erythema multiforme-like lesions. A syndrome with characteristic immunological abnormalities. *Arch Dermatol*, 1963; 88: 176-80.
2. Zeitouni NC, Funaro D, Cloutier RA, Gagné E, Claveau J. Redefining Rowell' syndrome. *Br J Dermatol*, 2000; 142: 343-6.
3. Aydogan K, Karadogan S, BalabanAdim S, Tunali S. Lupus erythematosus associated with erythema multiforme: Report of two cases and review of the literature. *J Eur Acad Dermatol Venereol*, 2005; 19: 621.
4. Gender Differences in Systemic Lupus Erythematosus. *Gender Medicine* Aug 1 2004.
5. Alakes Kumar Kole, Alakendu Ghosh. Cutaneous manifestations of SLE in a tertiary referral center. *Indian J Dermatol*, 2009 Apr-Jun; 54(2): 132-136.
6. Medline plus. Reviewed on 11/12/2014.
7. Shteyngarts AR, Warner MR, Camisa C. Lupus erythematosus associated with erythema multiforme: Does Rowell syndrome exist? *J Am Acad Dermatol*, 1999; 40: 773.