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Original Research Paper

Dermatology

ROWELL SYNDROME- A DIAGNOSTIC DILLEMA

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ABSTRACT Rowell syndrome is a rare entity with coexistence of lupus erythematosus and erythema multiforme like lesions with characteristic immunological pattern. The acceptance of Rowell syndrome as a distinct entity is controversial and not widely acknowledged. The syndrome was first described in 1963 by Rowell, Beck and Anderson and till date about 95 cases of erythema multiforme like lesions associated with lupus erythematosus have been described in literature with major being reported in adult females.

KEYWORDS : Rowell syndrome, Lupus erythematosus, Erythema multiforme like lesion.

INTRODUCTION

Rowell syndrome is a constellation of erythema multiforme like lesions in association with lupus erythematosus along with findings of speckled ANA, positive rheumatoid factor (RF), positive anti-La/anti- Ro, and clinical findings of chilblains. The coexistence of erythema multiforme like lesions and lupus erythematosus was first described by Schotz in 1922; however distinct entity of Rowell syndrome was described in 1963.[1,2] The diagnostic criteria of rowell syndrome:

Major Criteria:-
SLE,
EMF-like lesions,
speckled pattern of ANA

Minor Criteria:-
chill bains,
antiRo /antiLa,
positive rheumatoid factor.
All 3 major and at least 1 minor criteria are required for diagnosis[3] Generally, Rowell syndrome affect adult female. Here we present cases of 14 year old girl and 22 year old female.

Case 1

A 14 year old girl presented with red raised lesions on both upper, lower limbs and trunk with fever since 5 days. Fluid filled blisters at the angle of mouth since 4 days. History of photosensitivity present. On cutaneous examination multiple erythematous to hyperpigmented plaques are seen on cheeks, forehead, neck, chest and abdomen with few plaques showing crusting present on upper limb and lower limb. Multiple erythematous patches are present on palms and soles which are targetoid in nature.

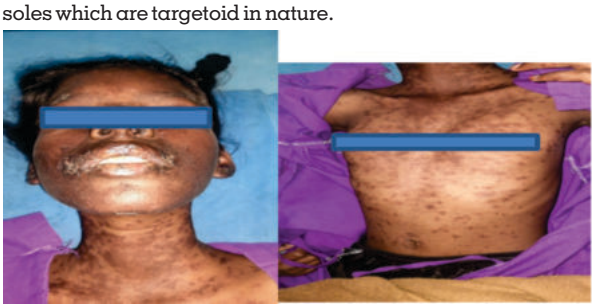

Fig 1 – Multiple erythematous to hyper pigmented plaques present on cheeks, neck&trunk.

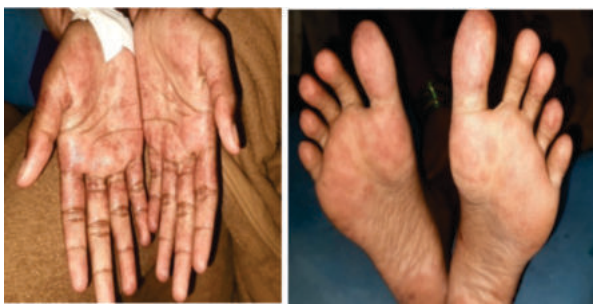

Fig 2- targetoid like lesions seen bilaterally over palms and soles.

Differential diagnosis includes erythema multiforme like lesions, systemic lupus erythematosus, rowell syndrome. On further workup blood investigation showed pancytopenia. Renal parameters, serum electrolytes, coagulation profile are all with in normal limit. ANA were positive with speckled pattern, and ANA profile showed the presence of anti- ds DNA, anti-histone, anti SS-A. Rheumatoid factor was negative. It fulfills the diagnostic criteria of SLE [ACR/EULAR criteria] with 19 points [ANA – 6; fever-2; anaemia-4; leucopenia-4; thrombocytopenia-3]. Therefore given the presence of both LE and EM like lesions with positive ANA and anti-Ro/SSA, a diagnosis of rowels syndrome was determined. The patient was kept on IV methyl prednisolone pulse therapy [750mg in 100 ml NS slow infusion over 1 hour for 3 consecutive days] followed by oral prednisolone 1mg/kg/day and hydroxychloroquine 4mg/kg/day. Significant improvement was noted in the skin, mucosal and palmoplantar lesions after 2 weeks. Azathioprine was added later. Patient is in regular follow up with no recurrence of skin lesions.

Case 2

A 22 year old female presented with red raised lesions on upper limbs, lower limbs, trunk, face since 1 month and painful oral lesions since 15 days. Patient had history of drug intake prior to development of lesions which include antibiotics (ofloxacin, ivermectin, metranidazole) and NSAIDS. History of photosensitivity present. Cutaneous examination revealed multiple annular erythematous scaly plaques with occasional crusting seen on the upper back. Crusted plaques are present on upper and lower lip. Erythematous plaques are seen on forehead, nose, and cheeks. Erythematous plaques are present on both feet and palms which are targetoid in nature. Few targetoid plaques

are seen on extensor aspects of both arms. Multiple erythematous scaly plaques with central crusting seen on extensor aspects of both arms and forearms. Few ulcers of irregular size and shape are present on hard palate with largest being 5*3 cm and smallest being 0.5*1cm.

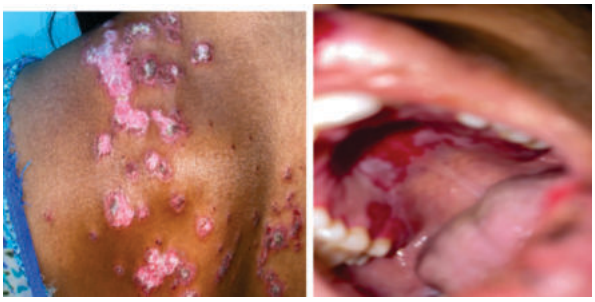

Fig 3 –a) annular to polycyclic erythematous scaly plaques present on upper back. b)ulcers of irregular size and shape present on hard palate.

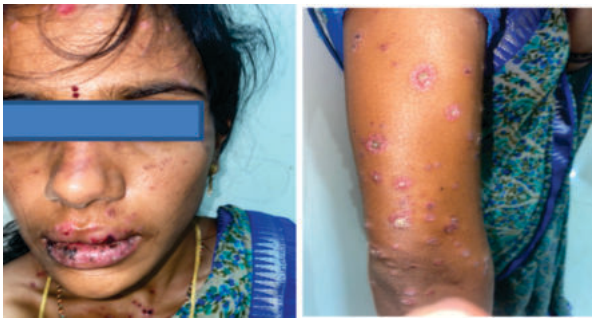

Fig 4– a) crusted plaques present on lips b)erythematous annular scaly plaques with central crusting present on extensor surfaces of B/L upper limbs. Few targetoid lesions are also noted.

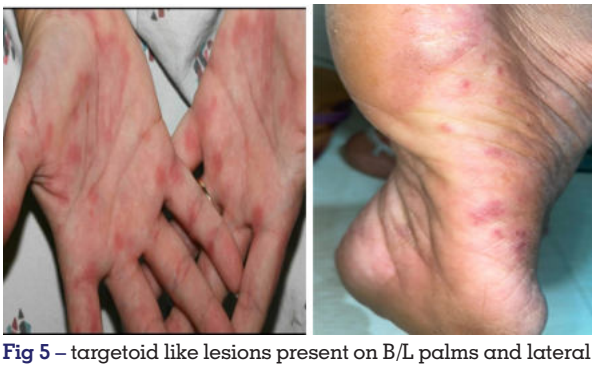

Fig 5 – targetoid like lesions present on B/L palms and lateral surface of B/L feet

Laboratory investigation shown leukocytopenia and raised ESR. Renal parameters, serum electrolytes, coagulation profile are all with in normal limit. On serology - positive ANA with speckled pattern and ANA profile showing positive Anti Sm Ab and Anti U1RNP and rheumatoid factor was positive. It fulfills the diagnostic criteria of SLE [ACR/EULAR criteria] with 15 points [ANA – 6; leucopenia-3; oral ulcer- 2; SCLE- 4]. Given the presence of EM like lesions and SCLE like lesions with speckled pattern of ANA with positive RF, the diagnosis of rowell syndrome is considered. The patient was kept on oral methyl prednisolone 1mg/kg/day and hydroxychloroquine 4mg/kg/day. Potent topical corticosteroids were advised. Significant improvement was noted in the skin, mucosal and palmoplantar lesions after 3 weeks. Azathioprine was added later. Patient is in regular follow up with no recurrence of skin lesions.

DISCUSSION

Rowell syndrome is a disease characterized by both LE and EM-like lesions in subjects with a characteristic immunologic pattern. The median age reported is 32 (range 9–87); however, pediatric or elderly cases have also been described. RS appears to be more prevalent in female patients than male patients (female to male ratio is 8:1) and only eight patients were under 18.[4] The diagnosis of LE usually precedes the appearance of EM-like lesions.[4] In our patients, the appearance of LE and EM lesions was almost simultaneous.

Classic EM is often caused by bacterial and viral infections (e.g., mycoplasma pneumonia, herpes simplex virus), drugs (e.g., antibiotics, anticonvulsants, nonsteroidal anti-inflammatory drugs, tuberculostatic drugs), or malignancies.[5] EM cases associated with LE lesions where an EM trigger factor is missing are considered an RS diagnostic criterion.[2,6] Patients with cutaneous LE may develop coincidental EM. However, if characteristic serological abnormalities are present and there is no obvious precipitating event, the association is known as Rowell's syndrome. In both cases, a possible trigger factor for EM was not identified.

In 1963, Rowell et al[2] first described diagnostic criteria for RS: presence of discoid lupus erythematosus (DLE) and EM-like lesions, positive RF, speckled ANA, and a saline extract of human tissue (anti-SJT), now known as similar to anti-Ro/SSA positivity. In 1995,Lee et al[6] observed the presence of chilblains in cases of RS. In 2000, Zeitouni et al[3] divided the diagnostic criteria into major and minor. The major criteria included LE (acute, subacute, or systemic), EM-like lesions, and antinuclear antibodies positivity. The minor criteria were the presence of chilblains, and presence of anti-Ro/SSA or anti-La/SSB antibodies, and rheumatoid factor. All major criteria and at least one minor criterion are necessary for an RS diagnosis.

Our patients have met the diagnostic criteria of rowell syndrome where case 1 have met the 3 major criteria and positive SSA (minor criteria), case 2 has also met 3 major criteria and positive RF.

The prognosis and treatment are similar to those of systemic lupus erythematosus or DLE that occur alone. In most cases of RS, prednisone combined with azathioprine, antimalarials (e.g., chloroquine, hydroxychloroquine), dapsone, or cyclosporine were frequently reported.

CONCLUSION

Many argue that Rowell syndrome is not a distinct entity and now widely considered to be a variant of SLE[7]. RS is a rare entity which has been regarded as only a coincidence, an overlapping syndrome, a variant of CLE, or a true "ghost syndrome." The therapeutic regimen, response and prognosis in Rowell syndrome are similar to that of SLE. The occurrence of Erythema multiforme with LE do not alter the course, therapy, or prognosis of the disease.

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