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Case Report

ROWELL SYNDROME - A RARE CASE REPORT

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A B S T R A C T

Rowells syndrome is a rare entity consisting of erythema multiforme like lesion with lupus erythematosus with laboratory findings. Here we present a 16 year old girl satisfying the criteria of Rowells with good response to antimalarials and systemic corticosteroids.

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INTRODUCTION

Rowell syndrome is an association of lupus erythematous and erythema multiform (EM) like lesions along with certain laboratory findings. It was first described by Schotlz in 1922. At present time, there were many controversies regarding Rowell as a special entity or should be considered as subacute cutaneous lupus erythematosus subset. . [1],[2]

Case Report

A 16 year old unmarried female attended dermatology department with multiple dark coloured lesions all over the body since 2 months.Initially she developed itchy dark coloured lesions over face and scalp later progressing to involve trunk and upperlimb.Patient had history of fever, oral ulcers, hairloss and periorbital puffiness which started later. There was history of multiple joint pain and photosensitivity. Also she had significant weight loss (12kgs in 2 months) along with loss of appetite and fatiguability. She took sidddha medication (topical) for 15days after which lesions got worsened. No history of drug intake prior to the onset.

On detailed cutaneous examination, bilaterally symmetrical hyperpigmented plaques with few erosions and crusts seen over face involving bilateral ears [Fig. 1, 4]. Few typical target lesions [Fig. 2]were noted over the trunk. There was hemorrhagic crusting and erosions over lips and palate and nasal mucosa [Fig. 3]. Scalp showed patchy alopecia. Genitals, nails, palms and soles were normal. On general examination, she was pale.

With these history and clinical examination ,patient was subjected to baseline investigations. Complete hemogram was normal except anemia (Hb:7.9 g/dl). In urine analysis,urine albumin was 2+ and 24hrs urine protein was 517mg/dl. Other investigations were found to be normal.

Skin biopsy revealed focal sites of degenerative keratinocytes and perivascular lymphocytic infiltrates. Antinuclear antibody blot profile was done which showed positive speckled pattern of Antinuclear antibody, strongly positive Ribosomal P protein, positive Anti-LA(ss-a) and Anti-Ro (ss-b). Direct immunoflourescence was not done because of the unavailability.

Patient was treated with systemic prednisolone of dose 30 mg/day then tapered slowly to 12.5mg/day along with systemic Hydroxychloroquine of dose 200mg/day for 2 months initially and then with the maintenance dose of 100mg /day for 8 months. Significant improvement was noted within 6weeks.

Other supportive measures like photoprotection, systemic antibiotics, oral antihistamines , oral antacids, topical steroids for mucocutaneous lesions and calcium supplements. Ophthalmologic follow-up were done.

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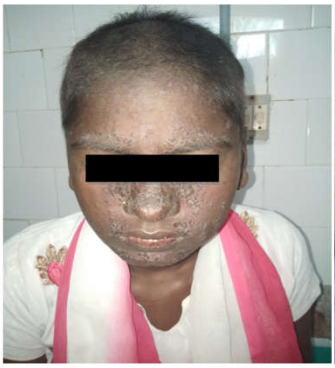

Fig.1

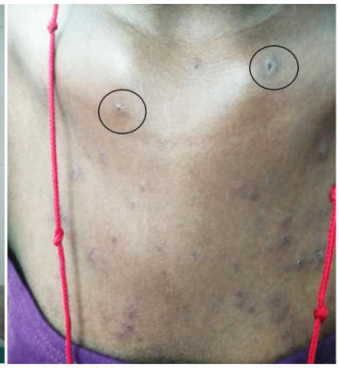

Fig. 2

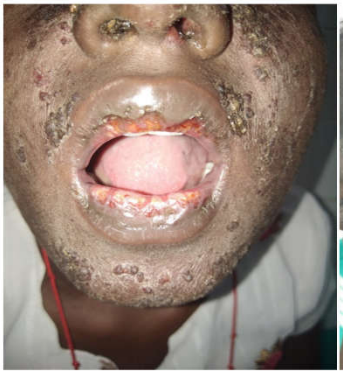

Fig. 3

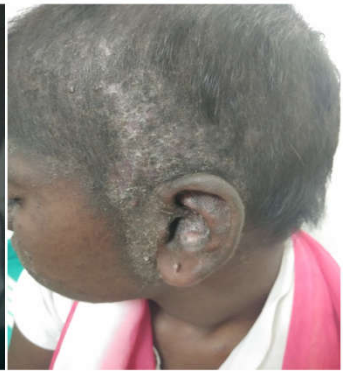

Fig. 4

DISCUSSION

The association of LE with EM was first reported by Shlotz[7] in 1922. In 1963, Rowell et al [6] defined this association as a distinct entity in four patients with discoid lupus erythematosus (DLE).

Then it was redefined by Zeitouni.et al in 2000 as 3 major and 3 minor criteria. The major criteria includes 1) Lupus erythematous 2) Erythema multiforme like lesions 3) Speckled pattern of Antinuclear antibody (ANA). [3] The minor criteria includes 1) Chilblain lupus 2) positive Anti-LA(ss-a) and Anti-Ro (ss-b) 3) Positive Rheumatoild factor. For diagnosis 3major and 1minor criteria is sufficient.

Speckled ANA pattern is the most consistent feature of RS seen in about 88% of the cases, whereas RF is the least preserved feature, found in only 41%. [3],[4],[5] Our patient has fulfilled three major criteria and one minor criteria.

The recent criteria in 2012 was proposed by Torchia et al., based on 95 cases of rowels syndrome with the addition of histopathology of lesions and direct immunofluorescence (DIF)[Table no:1]. [8] Here, our patient fulfilled all the criteria except DIF which was not done.

Table No 1

Major criteria	Minor criteria
Chronic cutaneous lupus erythematosus [DLE , Chilblain] Erthyema multiforme like lesions	No triggering factors [Medications/infections]
Atleast one positive among these [speckled ANA, anti-Ro / SS-A , anti-La /SS-B	Lack of EM lesion on acral or mucosal surfaces
Negative DIF on lesional EM-like lesions	One other diagnostic criteria for SLE excluding discoid or malar rash, ANA, photosensitivity, oral ulcers or chilblains

Erythema multiform is a reaction pattern with multifactorial etiology. It is a hypersensitivity reaction usually triggered by infections, most commonly herpes simplex virus(HSV). It presents with a skin eruption characterised by a typical target lesion. There may be mucous membrane involvement. It is acute and self-limiting, usually resolving without complications.

Erythema multiforme is divided into major and minor forms. Erythema multiforme major is associated with severe oral involvement and prodromal symptoms.

However EMF and RS respond to a similar therapeutic regimen. Azathioprine, antimalarials, prednisone, dapsone, and cyclosporine have been used with good results. [5],[1],[2] Our patient responded well to prednisolone, and hydroxychloroquine.

CONCLUSION

This case illustrates the features of Rowells syndrome satisfying 3 major criteria and 1 minor criteria with good response to antimalarials and systemic corticosteroids.This case is Presented for its rarity.

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