CASE REPORT

Rowell Syndrome in Pregnancy: A Unique Case Report

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Abstract

Rowell syndrome is one of the peculiar diseases of the skin. It is characterized by its resemblance with systemic lupus erythematosus (SLE) and erythema-multiforme (EM)-like lesions. Criteria of its diagnosis as given by Rowell reflected similitude with that of SLE. Recently, two more diagnostic criteria were reanalyzed by Zeitouni and Torchia.

Complications during pregnancy cause poor maternal and fetal outcomes. Mother suffers from exacerbation of the lupus, renal impairment, development of hypertension, or venous thromboembolism. Antepartum complications include prematurity, intrauterine growth restriction, fetal miscarriage, neonatal lupus syndrome. Neonatal lupus may also present in its severe form as congenital heart block in up to 2% of newborns in women who are Anti-RO positive.

Keywords: Case report, Erythema multiforme, Pregnancy, Rowell syndrome.

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Case Presentation

A 20-year-old female patient with the pregnancy of 20 weeks presented with complaints of skin lesions worsening over time for 20 days. Patient already had the skin condition since her teenage. She had taken treatment for infertility as she was a known case of polycystic ovarian disease. The lesions were hemorrhagic crusted target-like lesions all over the body involving the face and extremities (Fig. 1). There was only redness on the palm and soles. The patient was referred to dermatology for its management. There was no history of any co-morbidity or any chronic illness. Her family history was also not significant. The patients biochemistry profile of the patient is elaborated in Table 1. Her skin biopsy showed orthokeratosis, mild spongiosis, and vacuolization of the basal cell layer and findings were suggestive of erythema multiforme (EM). Her immunofluorescence was negative for IgG, IgM, IgA, and C3. The patient was started on steroids and hydroxychloroquine. As Torchia criteria was fulfilled, the patient was diagnosed with Rowell's syndrome. As Anti-RO antibodies were present, fetal ECHO was done and strict antenatal monitoring was done. All the skin lesions recovered with residual hyperpigmentation (Fig. 2). At 37 weeks of gestation on routine antenatal check-up per abdomen examination pointed towards fetal growth restriction and less amount of liquor clinically. USG was done and amniotic fluid index (AFI) was 1.7 cm and fetal weight was under 1.8 kg so cesarean section was performed in the presence of pediatrician. The neonate is being followed for the risk of neonatal lupus and congenital heart block.

Discussion

Rowell's syndrome is portrayed by the presence of both lupus erythematosus and EM-like lesions. The disease mainly occurs in young adults with female preponderance (female to male ratio 8:1).[1] Classic EM is caused by bacterial and viral infections (such as herpes simplex, mycoplasma, etc.), drugs (anticonvulsants, non steroidal anti-inflammatory drugs (NSAIDs), anti-tubercular drugs), malignancies, etc.[2]

Our patient met the criteria given by Torchia et al.[3] Moreover, no identifiable precipitating factor other than the patient being pregnant has been identified. Due to the change of the immunity

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Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details and related images.

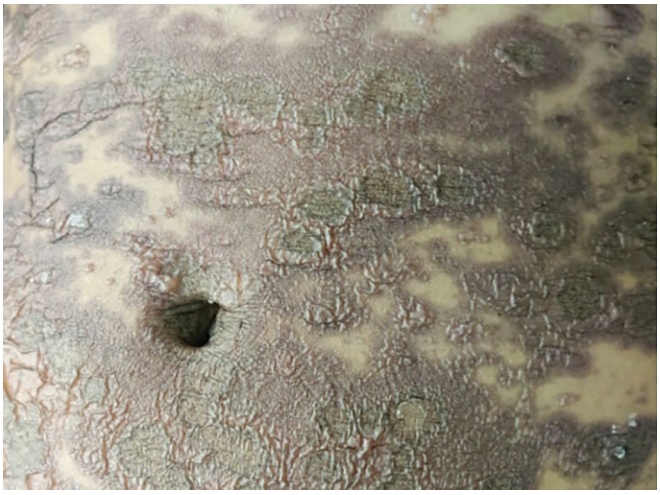

Fig. 1: Erythema-multiforme (target-like) lesion on the ventral surface of the body

pattern in pregnancy from TH1 to TH2 cell-directed, autoimmune disease tend to flare-up. The incidence of flare-up ranged from 13.5 to 65%.[4] Such patients are required to be treated with immunosuppressive drugs like prednisolone, hydroxychloroquine, etc. Our patient also recovered with residual hyperpigmentation. Prolonged usage of steroids for the remission of the disease, lead to intrauterine growth retardation and oligohydramnios necessitating surgical delivery (Table 2).

Table 1: Lab parameters and the result

Lab parameter	*Result*	*Normal value*
Hemoglobin	8.9	12–16 gm/dL
Platelet count	303×10^3	150–350 (10^3/μL)
Leukocyte	5.14×10^3	4–10 (10^3/μL)
C Reactive protein	5.11	<6 mg/L
Antinuclear antibody	Positive +++ (11.20)	<1.5
Anti-SSA/Ro	Positive +++ (dilution 1.101)	<1/10 titer
Anti-SSB/La	Negative	<1/10 titer
Anti-DsDNA	Negative	<1/10 titer
Anti-nucleosome	Negative	<1/10 titer
Anti-histone	Negative	<1/10 titer

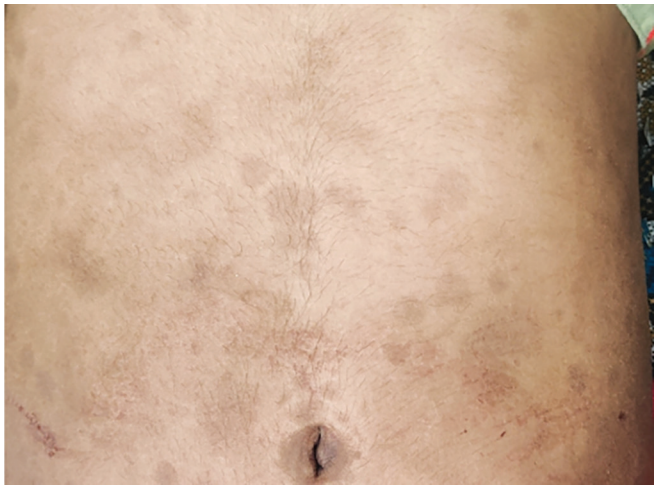

Fig. 2: Healed lesions with residual hyperpigmentation

Diagnostic Criteria

A—Torchia et al.[3]

Major Criteria

- The presence of chronic cutaneous lupus erythematosus (Discoid Lupus Erythematosus (DLE) and/or chilblain).
- Erythema–multiforme-like lesions (typical or atypical targets).
- At least one positivity among speckled ANA, Anti-RO/SSA, and anti-La/SSB antibodies.
- Negative direct immunofluorescence assay on lesional EM-like lesions.

Minor Criteria

- Absence of infectious or pharmacologic triggers.
- Absence of typical EM locations (acral and mucosal).
- Presence of at least one additional American rheumatism association (ARA) criterion for diagnosis of SLE besides discoid rash and ANA and excluding photosensitivity, malar rash, and oral ulcers.

Diagnosis

All four major criteria and at least one minor criteria.

B—Zeitouni et al.[5]

Major Criteria

- Presence of SLE, DLE, or subacute cutaneous LE.
- Erythema–multiforme-like lesions with or without mucous involvement.
- Speckled pattern of ANA.

Minor Criteria

- Chilblains.
- Anti-RO/SSA or Anti-La/SSB.
- Positive Rheumatoid factor.

Diagnosis

All three major criteria and one minor criteria.

Table 2: Shows the difference between Rowell syndrome, systemic lupus erythematosus, and erythema multiforme

Parameter	*Rowell syndrome*	*Systemic lupus erythematosus*	*Erythema multiforme*
1. Presentation	Combination of erythema multiforme-like lesions and characteristic immunologic pattern	Fever, joint pain, rash (discoid rash, photosensitivity)	Symmetrical, discrete, round erythematous macule/papules may even evolve into target lesions showing different stages of resolving disease
2. Diagnosis	Torchia et al. criteria[3] Zeitouni et al. criteria[5]	European league against rheumatism/ American college of rheumatology (EULAR)/ACR 2019 criteria	Clinical presentation + history of recent infection (HSV, *M. pneumoniae*) or drug intake. Mucosal involvement
3. Histopathology	Epidermal necrosis (picture like EM) + Periadnexal lymphocytic infiltrate + direct immunofluorescence positive	Apoptotic keratinocytes with marked basement membrane thickening. Vacuolar interface change. Follicular keratotic plugging	Apoptotic individual keratinocytes. Hydropic degeneration of basal keratinocytes. Spongiosis
4. Treatment	Topical and oral steroids, Immunosuppressive drugs, Dapsone	Antimalarials like hydroxychloroquine Topical steroids	Stop medicine if trigger present. Treat underlying infection. High-potency topical steroids, iv fluids, electrolyte repletion, prolonged prednisolone (in severe cases)

According to Zeitouni et al.[5] all three major and one minor criteria are fulfilled in our case. But the diagnosis of this condition is so confusing other authors have also worked on whose criteria need to be evaluated as well. Since erythema multiforme, systemic lupus erythematosus and Rowell syndrome have overlapping characteristics so (Table 2) elaborates their distinct differences.

Pandhi and workers suggested that even though Rowell syndrome is mostly seen in middle age pregnancy being in a vulnerable state also manifests this condition in the form of flare-up.[6]

Mandelcorn and workers have found that the presentation of this spectrum of disease is variable. It may range from simple EM-like lesions with positive serology to dangerous toxic epidermal necrolysis.[7]

Aydogan et al. suggested that this condition having an immunogenic association may show recurrence without any triggering factor and its control might seem impossible. However, our patient recovered well with steroids and hydroxychloroquine and was disease-free for almost the last 2 years.[8]

The diagnosis of Rowell syndrome is a tricky one as its clinical and histopathological patterns are overlapping with SLE and EM. Histopathology along with serology should always be considered and aid of the formed criteria of diagnosis will lead to prompt diagnosis and treatment.

Conclusion

Rowell syndrome is a rare entity. It should be suspected in females with a history of LUPUS or the presence of EM-like lesions, with no evidence of a precipitating factor. Moreover, strict neonatal monitoring is to be done in lieu of the possibility of neonatal lupus or congenital heart block. As a very limited number of cases have been reported, more studies are required to be undertaken. Proper careful examination and correct investigation panel need to be decided to make exact diagnosis and management.

The presentation of Rowell syndrome coincides with SLE. Both can flare-up during states like pregnancy. Thus, timely diagnosis and starting the patient on immunosuppressive-like hydroxychloroquine can prevent adverse maternal and fetal outcomes.[9]

Delivery of the fetus should take place in a multidisciplinary unit as sometimes there can be multiorgan involvement of the mother and moreover good NICU facility can prevent and manage congenital heart block effectively.[10]

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