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Case Report

Systemic lupus erythematosus presenting with bullous lesions, cutaneous vasculitis and laryngeal ulcerations-a rare association

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ABSTRACT

Bullous SLE is an uncommon complication of SLE with autoantibodies to type VII collagen of dermo-epidermal junction. We report a 31-year-old female who presented with vesiculobullous lesions in the photo exposed areas along with vasculitic lesions over palms and soles and ulcerations in the larynx. She also had history of photosensitivity and hair loss. Investigations showed significant anaemia and high titres of antibodies (ANA, anti-ds-DNA, anti Sm) confirming SLE. Histopathology showing sub epidermal blister with neutrophils and immunofluorescence favoured diagnosis of bullous SLE. The patient responded well to high dose parenteral steroids and dapsone. In view of high probability of renal involvement, patient was referred to nephrologist and is under regular follow up.

Keywords: Bullous SLE, Cutaneous vasculitis, Laryngeal ulcerations

INTRODUCTION

Bullous systemic lupus erythematosus (BSLE) is a rare manifestation of SLE. Cutaneous lesions are reported during the course of SLE in 76% patients; however, vesicobullous lesions account for less than 1% of these.[1] Lesions are usually generalized with predilection for sun-exposed areas, neck, upper trunk and proximal extremities.[2] Majority of BSLE patients have autoantibodies against type VII collagen.[3] However, autoantibodies against other target antigens of the DEJ including BP180, BP230 and laminin 332 are also reported in BSLE.[4,5] We present a distinctive case of BSLE associated with cutaneous vasculitis and laryngeal ulcerations.

CASE REPORT

A-31-year-old female, mason by occupation came with chief complaint of painful raw areas in oral cavity since 1 month; multiple fluid filled lesions over neck, lips, face and trunk since 2 weeks and burning sensation over palms and soles since 1 day. The blisters in oral cavity ruptured spontaneously within 2-4 days leaving painful erosions associated with difficulty in swallowing. Few blisters over other areas healed with pigmentation. The patient also developed tender papular lesions over palms and soles associated with burning sensation since 1 day. History of photosensitivity and hair loss were present since 4 months. On general physical examination pallor was present. There was no evidence of any icterus, cyanosis, clubbing, lymphadenopathy or pedal edema. Examination of respiratory, cardiovascular, central nervous system and gastrointestinal system was normal. On dermatological examination, multiple grouped and few discrete vesicles and bullae with erosions and few haemorrhagic crusts present over neck (Figure 1), eyelids, perioral area, right arm, left axilla and umbilical region. Multiple erosions present over lips and buccal mucosa (Figure 2). Tender erythematous papules were present over palms and soles. Few areas of post inflammatory hypopigmentation were noted. Diffuse alopecia was present over the scalp but there was no evidence of any scarring or scaly lesions. Nails and genital mucosa were normal.

Based on history and clinical features, a differential diagnosis of bullous SLE, pemphigus vulgaris, erythema multiformae, pemphigus herpetiformis and IgA pemphigus were considered. Patient was subjected to detailed laboratory investigations and complete blood picture showed presence of anaemia (Hb-8.2 g/dl) and leucocytosis (TLC-16,600cells/cumm) with normal platelet count and ESR. Serum biochemistry profile including RBS, LFT, blood urea and serum creatinine were within normal limits. Collagen profile revealed positive antinuclear antibody, anti-ds-DNA and anti Sm antibody and decreased C3 (0.65g/l), C4 (0.079g/l) complement levels. Radiological imaging including chest X-ray and USG abdomen did not show any abnormality. In view of severe throat pain and dysphagia patient was subjected to videolarygoscopy which revealed multiple ulcerations over lateral tongue, posterior pharyngeal wall, aryepiglottic folds and pyriform fossa along with oedematous arytenoids.

Skin biopsy samples were sent for histopathology and direct immunofluorescence. Histopathology showed thin epidermis with sub epidermal blister containing abundant neutrophils (Figure 3). Superficial dermis showed dilated vessels and mild interstitial infiltration with neutrophils and lymphocytes and deep dermis showed mild peri-eccrine neutrophilic infiltrate. On direct immunofluorescence, a granular pattern of IgG, IgA, IgM and C1q deposits was demonstrated along the dermo-epidermal junction (Figure 4-7).

Based on clinical, serological, histopathological and direct immunofluorescence findings, a diagnosis of bullous SLE was confirmed. The patient was treated with parenteral steroids (Inj. dexamethasone 1 CC IV BD for 1 week with gradual tapering over 1 month), oral dapsone 50 mg OD, oral hydroxychloroquine 200 mg OD along with broad spectrum sunscreen, topical steroids and other supportive treatment which resulted in marked clinical improvement within 10 days (Figure 8). She was later administered inj. methyl prednisolone 500 mg IV pulse therapy for a period of 3 days as suggested by nephrologist.

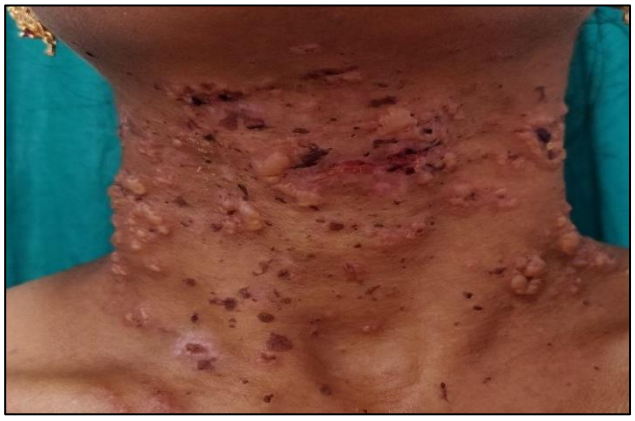

Figure 1: Multiple grouped and few discrete vesicles and bullae with erosions over neck.

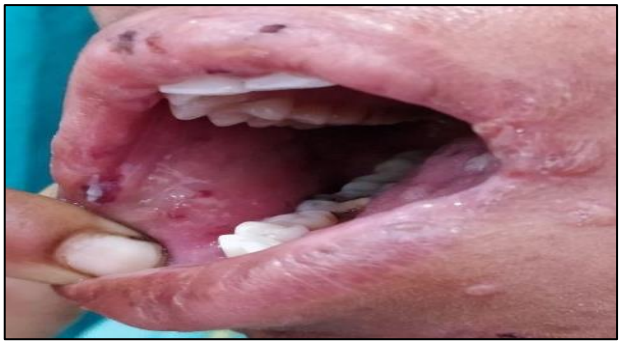

Figure 2: Multiple erosions over lips and buccal mucosa.

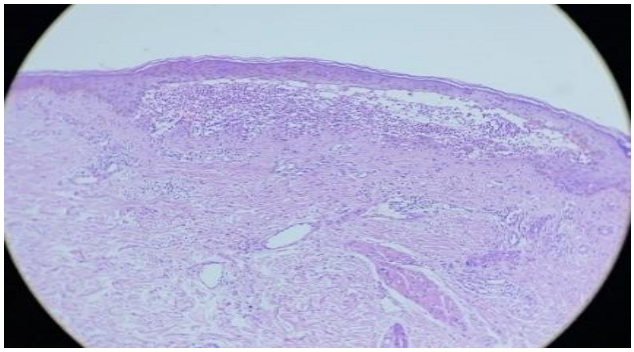

Figure 3: Histopathology showing sub epidermal blister containing abundant neutrophils.

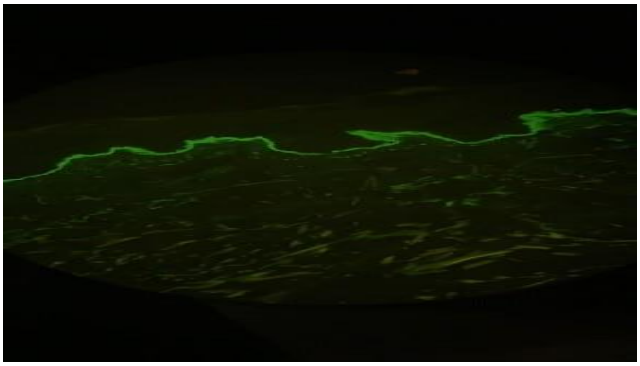

Figure 4: DIF showing granular pattern of IgG deposits along DEJ.

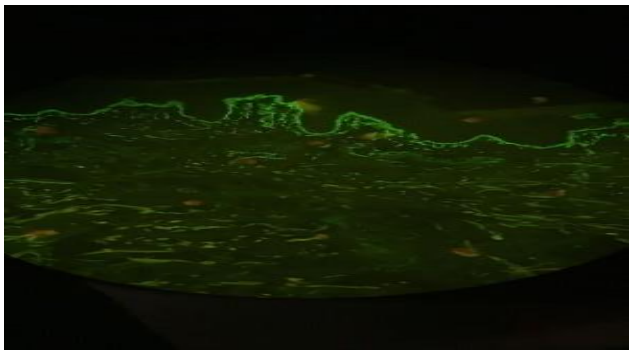

Figure 5: DIF showing granular pattern of IgA deposits along DEJ.

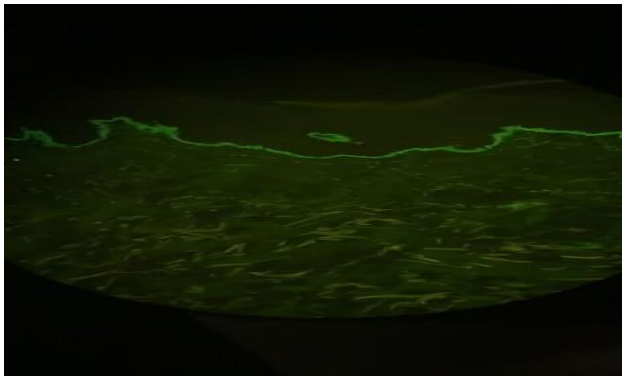

Figure 6: DIF showing granular pattern of IgM deposits along DEJ.

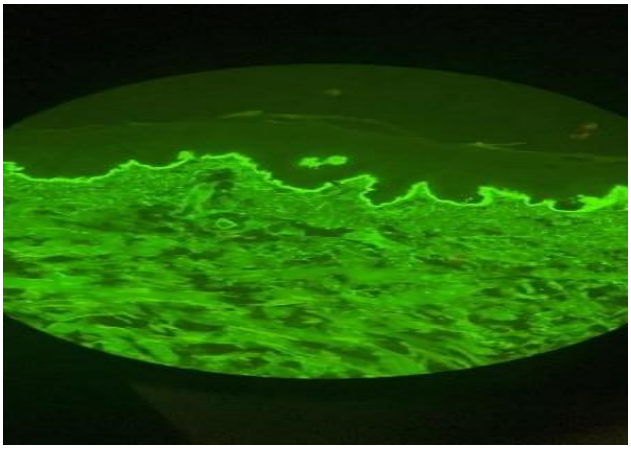

Figure 7: DIF showing granular pattern of C1q deposits along DEJ.

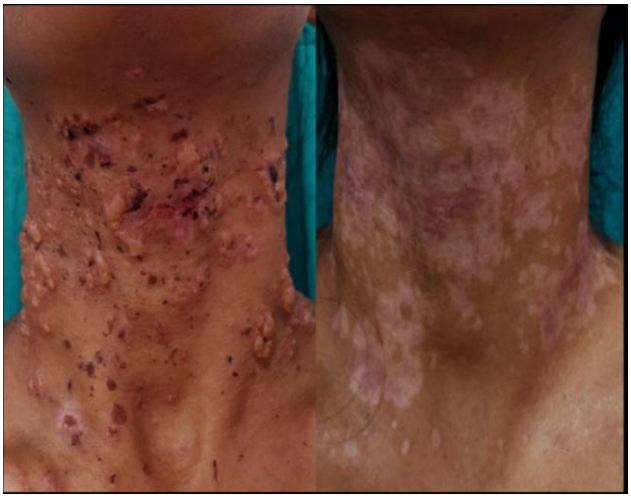

Figure 8: Multiple grouped and few discrete vesicles and bullae with erosions over neck; marked improvement seen 10 days after treatment.

DISCUSSION

The diagnostic criteria of bullous SLE were first described by Camisa and Sharma.[6] They include (1) diagnosis of SLE based on American college of rheumatology (ACR) criteria; (2) vesicles and/or bullae arising upon but not limited to sun exposed areas; (3) histopathologic features similar to dermatitis herpetiformis; (4) DIF with IgG and/or IgM and often IgA at the basement membrane zone; and (5) indirect IF testing that can be negative or positive. In the present case 4 out of 5 criteria were positive and IIF could not be performed.

The modified criteria by Gammon and Briggaman include: (1) fulfilment of American rheumatism association (ARA) criteria for SLE; (2) acquired vesiculobullous eruption on (but not limited to) sun-exposed skin; (3) histologic evidence of sub epidermal blister with neutrophilic infiltrate; (4) presence of IgG, IgM, IgA, and C3 at the BMZ; (5) evidence of antibodies to type VII collagen; and (6) co-distribution of immunoglobulin deposits with anchoring fibrils/type VII collagen by immunoelectron microscopy.[7] On the basis of these, BSLE can be of two distinctive types: type I fulfilling all six criteria, while type II (undetermined antigen or dermal antigen other than type VII collagen) satisfies criteria 1-4 only.[7]

Histopathological findings of BSLE resemble dermatitis herpetiformis with sub epidermal splitting, a dense neutrophil-dominated infiltrate in the upper dermis sometimes accumulating in micro abscesses of the papillary tips, and dermal edema. In addition, mucin depositions are usually seen in the reticular dermis and sometimes signs of leukocytoclastic vasculitis. Basal layer vacuolization characteristic for cutaneous LE is not present. In the present case histopathology favoured as BSLE as most of the above features were present.

In SLE patients with prevalent cutaneous lesions, ANAs have been found positive in 75% of cases. Changes in anti-dsDNA antibody titres along with low complement levels correlate with disease activity and lupus nephritis and can be useful in monitoring disease activity. In the present case ANA, anti-ds DNA, anti-sm antibodies were positive along with low complement levels. Based on above findings patient was referred to nephrologist for evaluation of risk of lupus nephritis.

DIF of BSLE shows linear or granular staining of the DEJ by IgG. Additional linear or granular deposits of IgM, IgA, and/or C3 may also be found.

Previous studies show that prevalence of vasculitis in SLE is 11-36%.[8] In this patient painful and tender erythematous papules suggestive of cutaneous small vessel vasculitis were observed over palms and soles but other skin lesions suggestive of cutaneous small vessel vasculitis were not seen at the time of presentation. Involvement of laryngeal and esophageal mucosa in SLE is extremely rare. Laryngeal involvement in SLE can range from mild ulcerations, vocal cord paralysis, and oedema to necrotizing vasculitis with airway obstruction.[9]

In the present case only superficial mucosal ulcerations were present without any evidence vocal cord paralysis or necrotising vasculitis which resolved completely with the treatment.

Most reported cases of bullous SLE occur concurrently or after an established diagnosis of SLE.[10] Whereas in our case blistering eruption was initial presentation of SLE. Presence of other clinical features such as photosensitivity, hair loss and joint pains which started insidiously 4 months prior to onset of skin lesions also favoured diagnosis of SLE.

CONCLUSION

This case highlights the extremely rare concurrent occurrence of bullous lesions, vasculitic lesions and laryngeal ulcers in SLE. This case emphasizes the importance of prompt recognition and treatment of bullous SLE as the initial manifestation of SLE due to its association with high disease activity and concomitant risk of lupus nephritis and autoimmune hemolytic anemia. Therefore, immediate referral to rheumatologist is also advised.

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