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SLE With Toxic Epidermal Necrolysis Like Presentation Successfully Treated with Plasmapheresis and Methyl Prednisolone Pulse: A Rare Case Report

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ABSTRACT

Toxic epidermal necrolysis (TEN), a life threatening dermatological condition, characterized by mucositis, epidermal detachment and erosions occurs mostly secondary to drugs with rare causes like systemic lupus erythematosus(SLE) have been implicated. TEN like presentation of SLE, a type of ACLE is being rarely reported due to the difficulty in differentiating it from drug induced TEN. Herein, we report a 25 year old male presenting with fever, weight loss, mucosal erosions, generalized fluid filled lesions and detachment of skin diagnosed as TEN like presentation of SLE. The patient showed positive anti Smith antibodies and was successfully treated with methyl prednisolone pulse therapy and plasmapheresis

Keyword: *Anti Smith antibody, male, methylprednisolone pulse, plasmapheresis,SLE,TEN.*

1. INTRODUCTION

Toxic epidermal necrolysis (TEN) is a life threatening dermatological condition characterized by mucositis, epidermal detachment, and erosions.[1] The etiology is secondary to drugs mainly, with other rare causes like SLE, infections, and vaccinations having been implicated. Systemic lupus erythematosus (SLE) is a type of autoimmune disorder with varied

clinical presentations such as acute, subacute and chronic forms. Acute cutaneous lupus erythematosus. (ACLE) is a specific lesion in systemic lupus erythematosus (SLE) patients that is further classified into localized ACLE (malar rash), generalized ACLE, and toxic epidermal necrolysis-like ACLE.[2] TEN like cutaneous LE is a rare occurrence.[3] Differentiation between classic drug-induced TEN and TEN-like cutaneous LE is difficult and challenging. This is a case of newly diagnosed SLE with cutaneous features similar to TEN confirmed by histopathological findings and positive antinuclear antibodies with specific positivity to anti Smith antibody

Case report

A 25 year old male presented to the dermatology OPD with complaints of high-grade fever on and off associated with significant loss of weight for past one month. During the process of evaluation for fever, he developed itchy, red, and dark coloured raised skin lesions all over his body. He also gave a history of painful oral ulcers and burning sensation of skin on exposure to sunlight, which he attributed to fever. A provisional diagnosis of drug reaction was made, and treatment with steroids was initiated. During the course of treatment, he developed sudden onset hair loss involving 50%-60% of his scalp within a span of two days. Multiple flaccid bullae followed by erosions appeared all over the body, including the oral mucosa, with a subsequent episode of seizure. Cutaneous examination revealed sheets of dusky erythema with multiple crusted plaques, erosions, and flaccid bullae present all over the body, predominantly over the sun exposed areas. (Fig 1A,B&C) Pseudo Nikolsky sign was found positive over the dusky necrotic areas. Oral examination showed extensive erosions with diffuse hemorrhagic crusting noted over both the lips. (Fig 1B) Extensive non-scarring patchy alopecia with positive hair pull test was present over the scalp. (Fig 1D) Laboratory evaluation showed pancytopenia (Hemoglobin-9.2g%; totalcount-2,100; platelets-1,41,000), elevated liver enzymes, and positive direct-Coombs test. ANA profile was done, which showed positive anti-nuclear antibodies (3+ speckled pattern), negative anti-dsDNA, and positive anti-Smith antibody. His complement C3 levels (27.10mg/dl) were low. CT brain showed small vessel ischemic changes. Antiphospholipid antibodies and other investigations for systemic involvement were within normal limits.

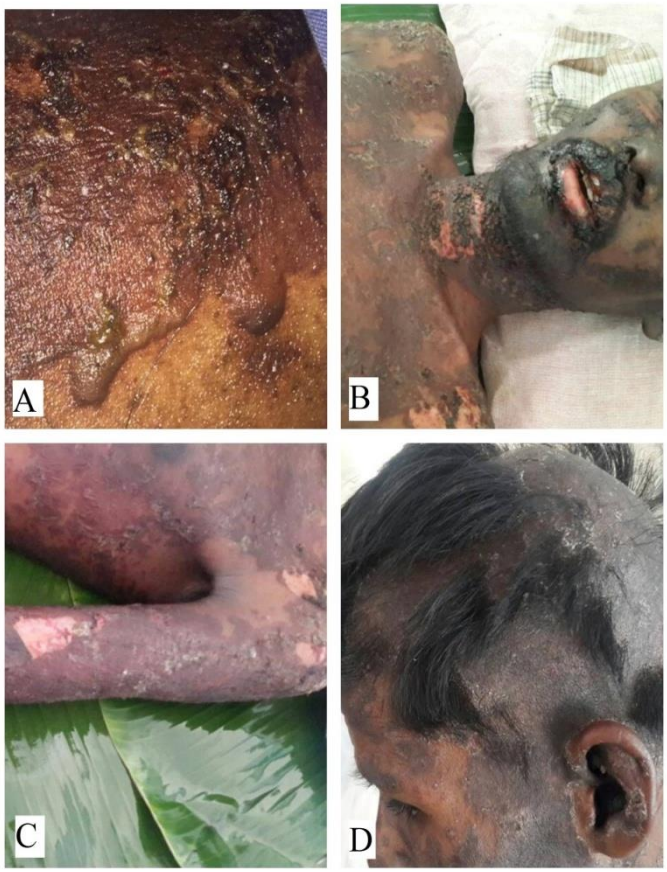

Figure 1: Multiple flaccid bullae,erosions and crusting over trunk (A). Haemorrhagic erosions and crusting over lips with sheets of dusky erythema and erosions involving chest and upper limb (B&C). Extensive non scarring alopecia patches noted over the scalp (D).

Histopathology of the bulla showed epidermal findings of focal disrupted intra-epidermal vesicle filled with RBCs and scanty

inflammatory cells , separation of epidermis at mid granular layer, and necrosis of individual keratinocytes. Basal cell degeneration, scattered chronic inflammatory cells with perivascular lymphocytic infiltration, and melanin incontinence were noted in the upper dermis. (Figure 2)

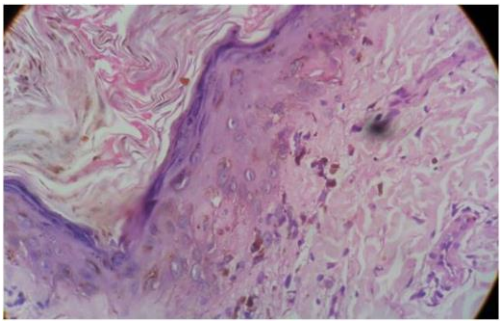

Figure 2: HPE(×40X) showing focal disrupted intra-epidermal vesicle filled with RBCs and scanty inflammatory cells , separation of epidermis at mid granular layer, and necrosis of individual keratinocytes. Basal cell degeneration, scattered chronic inflammatory cells with perivascular lymphocytic infiltration, and melanin incontinence were noted in the upper dermis.

The findings were suggestive of TEN like in the epidermis and SLE like in the dermis. Based on all these features and positive ANA, we came to a final diagnosis of TEN like SLE. The patient was administered IV methylprednisolone 1 gm pulse therapy for 3 days and three sittings of plasmapheresis, following which the lesions started resolving and the general condition of the patient also improved significantly. He was then maintained with injection dexamethasone 8 mg IV once daily, which was subsequently tapered and stopped.

2. DISCUSSION

Similar cases have been reported by a few other persons, among whom most of them were already diagnosed as SLE and were mostly females. The real challenge is when a new undiagnosed SLE case presents with features similar to classic TEN as observed in this report. Even though mucosal involvement is very rare in TEN like presentation of SLE, few cases similar to us with mucosal involvement have been reported previously. [3,4] The classic drug induced TEN and TEN like presentation of SLE can be distinguished from the features as shown in Table 1.

Table 1: Difference between drug induced TEN and TEN like presentation of SLE.

TEN like SLE	TEN
1.Photodistributed areas and history of photosensitivity	Diffuse involvement
2.No H/O any drug intake	Offending drug history+
3.Might have preceding diagnosis of SLE	
4.ANA+ with specific antibodies(Anti-ds DNA + anti-Sm antibody+)	ANA -
5.Involvement of mucosa especially oral mucosa and palms and soles rare and less severe	Involvement of mucosa and palms and soles very common and severe
6.Few necrotic keratinocytes at dermo-epidermal junction, vacuolar degeneration of the basal keratinocytes, moderate to dense perivascular and periadnexal lymphocytic infiltrate in dermis and mucin deposition.	Necrotic keratinocytes throughout the epidermis and sparse inflammatory infiltrate

This patient, in addition to ANA, also had anti-Smith antibody positivity, which is more specific for SLE in the case of a negative anti-ds DNA antibody. Anti-Smith antibody is mostly associated with lupus nephritis, psychosis, vasculitis, and hemolytic anemia.[4] Our patient had an episode of seizure, and CT showed vascular changes indicating CNS involvement.

Most of the TEN like SLE cases have been treated successfully with steroids and IV immunoglobulin, but data regarding plasmapheresis as a treatment option is limited. Simek I et al[5] have reported a case treated successfully with plasmapheresis. Our patient showed remarkable improvement within 3 sittings of plasmapheresis and IV methylprednisolone pulse therapy.

The pathomechanism of TEN is mediated by cytotoxic T cells and by Fas-Fas ligand interaction causing keratinocyte apoptosis.[6] A similar pathomechanism is involved in SLE, where CCL27 mediates the activation of autoimmune T cells and IFN-α-producing dendritic cells, which in turn release effector cytokines, creating a cascade along with upregulated Fas ligand. The removal of FasL by plasmapheresis is the rationale found to help in the treatment of TEN like SLE.[5]

3. CONCLUSION

We report this rare case of SLE presenting as TEN in a male with positive anti-Smith antibody successfully treated with methylprednisolone pulse therapy and plasmapheresis. A high index of suspicion is required for prompt diagnosis and treatment of such cases. This kind of presentation denotes the need for aggressive management of the disease to prevent severe systemic complications

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