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Original Research Paper

Dermatology

SJS/TEN- LIKE LUPUS ERYTHEMATOSUS PRESENTATION

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ABSTRACT

Stevens–Johnson syndrome (SJS), toxic epidermal necrolysis (TEN) presents as epidermal detachment and mucosal erosions. It is frequently seen secondary to drug ingestion and rarelydue to AIDS, bone marrow transplants, radiotherapy and systemic lupus erythematosus.

Here we describe a case of TEN like lupus presented with photosensitivity, epidermal necrolysis, minimal mucosal involvement, prolonged course and not associated with history of drug ingestion.

Although distinguishing SJS/TEN from TEN like lupus is clinically challenging, systematic approach combining clinical history with laboratory investigations may be helpful in making the diagnosis.

Dermatologist should be aware of this condition and include in the list of differentials while dealing with cases of SJS/TEN as this fatal condition requires aggressive treatment and prevention of potential complications.

KEYWORDS : SJS, TEN, SLE

INTRODUCTION:

Stevens–Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are dermatological emergencies, presents as epidermal detachment and erosions of mucosa.Incidence of TEN is 0.9-1.4 persons per million per year in general population. Most frequent causeis secondary to drug intake[1] and other causes include AIDS, bone marrow transplants, radiotherapy, SLE.

SJS/TEN is a very rare manifestation of LE [2] and can create diagnostic dilemma. Cutaneous lupus erythematosus can be "lupus specific" or "lupus nonspecific" based on histopathological features of interface dermatitis.[3] Lupus specific classified into acute, subacute and chronic cutaneous lupus erythematosus. LE usually presents with cutaneous manifestations of discoid rash, malar rash and alopecia. Most severe form of acute lupus is SJS/TEN. Here we describe a rare case of SLE presenting as SJS/TEN.

Case Report:

- An 18-year-old femalepresented tocasualty with1 week history of fever associated with photosensitivity, rash, alopecia and oral erosions. Multiple erythematous crusted plaques seen all over the body, which are predominantly seen on face. Desquamated areas over trunk, back associated with serous discharge. Similar lesions seen on palms and soles.
- She gave similar complaints 2months back for which she was treated with antibiotics, topical steroids, sunscreen from local hospital and showed mild improvement.
- No history of any drug intake or infections in past one month.
- On examination patient had pallor, hypotension and tachycardia.
- CT chest, ECG, 2D ECHO are normal.
- Routine blood and biochemical investigations showed anemia (6.1g/dl),leucopenia (2600cells/cu mm), thrombocytopenia (90,000/cu mm), raised ESR(60mm/hr). LFT, RBS are normal. Urine Proteins are raised (14mg/24 hours), Protein Creatinine ratio normal. 24-hour urine analysis showed raised creatinine (1750mg/24 hrs) and proteins (275 mg/24 hrs). Direct and Indirect Coombs Test-Negative. Complement levels 3 (13.37 mg/dL) Complement 4 (7.88 mg/dL) are decreased. Urine and blood for Culture and Sensitivity are negative.ANA profile positive with Ribosomal Protein+++, RNP/ Sm- ++,SSA & Ro(52)+.
- Histopathology revealed intraepidermal bulla which consists of eosinophilic necrotic material, lymphocytes, histiocytes and nuclear debri. Epidermis adjacent to bullae showed epidermal atrophy with loss of rete ridges and basal cell vacuolar degeneration.
- DIF- C3 and IgG deposition all dermoepidermal junction.
- EULAR and ACR criteria – score is 28
- Patient kept on Inj. Methyl Prednisolone 1g (in 100ml of NS over 1 hour) for 5 days, oral piperacillin, tazobactam, topical steroids, sunscreen, iv fluids and supplements. Patient showed improvement in 10 days and discharged in hemodynamically stable condition with oral mycophenolate mofetil 500mg tid.

DISCUSSION:

SJS/TEN more commonly seen secondary to drug intake and rarely secondary to lupus erythematosus. Drugs used in management of LE are more likely to cause SJS/TEN.[4,5]

Pathogenesis of TEN-like lupus is still not entirely clear. Clinical features of TEN like lupus include discoid rash, malar rash, photosensitivity, alopecia, bulla formation, shearing of epidermis and constitutional symptoms.

Few common clinical features are seen in TEN and TEN like lupus, TEN patients usually present with history of drug intake, sudden onset of symptoms within 1 to 3 weeks of drug ingestion, shearing of epidermis, severe mucosal involvement associated with constitutional symptoms and positive pseudo nikolsky sign. Histology shows epidermal necrosis due to keratinocyte apoptosis, subepidermal blister& no deposits are seen in DIF. Whereas in TEN like lupus there is no history of drug ingestion, develops gradually over few weeks especially involving photosensitive areas like face and then rash becomes generalized with mucosal involvement. Involvement of different systems and serological diagnostic criteria will guide fordiagnosis of TEN like lupus.[6]Histology shows interface dermatitis, epidermal necrosis, dermal inflammation and DIF may show Ig deposits.[7,8]

Better prognosis is seen for TEN like lupus erythematosus compared to drug induced TEN.[8,9] No treatment guidelines are available due to absence of large randomised and controlled clinical trials for this disease. [9] First line of treatment in TEN like lupus are corticosteroids, some show good response to IV Ig.

Ting et al. in 2004 described concept of 'acute syndrome of apoptotic pan-epidermolysis (ASAP)'. These includes all lethal clinical situations of severeepidermolysisdue to hyperacute apoptotic injury which can be due to either drug ingestion or other causes like Lupus erythematosus, acute graft versus host disease etc.[10] Furthermore, research is needed to substantiate this concept.

CONCLUSION:

Absence of drug intake, characteristic photosensitive rash, chronic course and serology points to the diagnosis of SJS –TEN like lupus erythematosus. Dermatologist should be able to recognise this fatal condition in its early stage and advise appropriate investigations with indispensable treatment to reduce morbidity and mortality of disease.

CASE REPORT IMAGES

Figure1: (A) Clinical image shows hyperpigmented plaques over face, neck, anterior aspect of chest with few of them showing desquamating areas in mid and left lateral aspect of neck (B, C) Desquamating lesions with oozing seen on back, chest and abdomen (D) Few erythematous plaques on forehead, above eyebrows and nonscarring alopecia of scalp are seen.

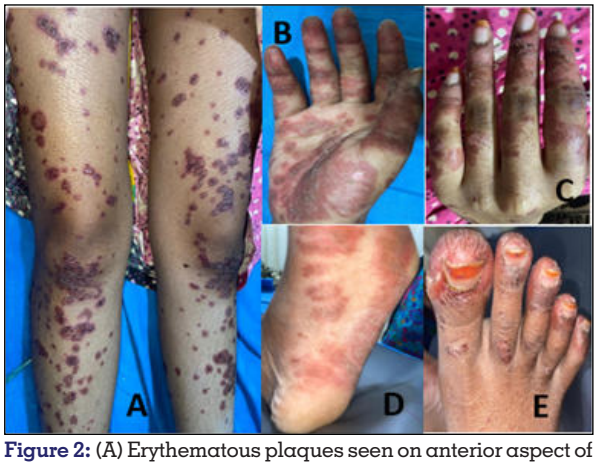

Figure 2: (A) Erythematous plaques seen on anterior aspect of thighs and upper two third of legs. (B) Erythematous plaques with scaling seen on palmar surface of right hand. (C) Erythematous plaques seen on dorsum of left hand. (D, E) Erythematous plaques with scaling seen on plantar, dorsal surface of feet.

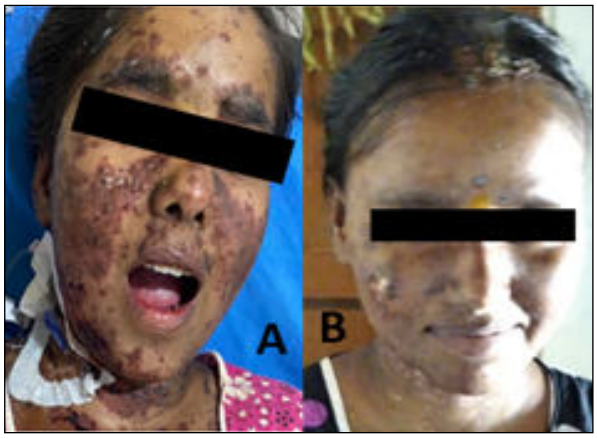

Figure 3: (A) Erythematous plaques seen on malar region with sparing of nasolabial folds. (B) Resolution of malar rash with hyper and hypopigmentation after 2months of treatment

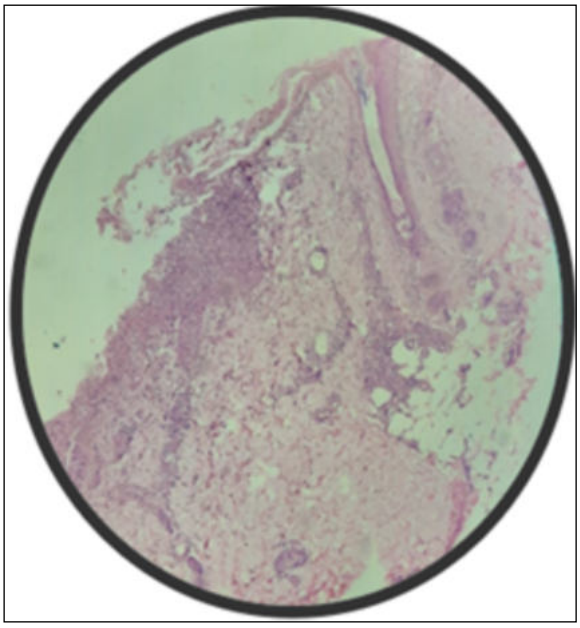

Figure 4: Histopathology on H & E stain shows Intraepidermal bulla which consists of eosinophilic necrotic material, basal cell vacuolar degeneration.

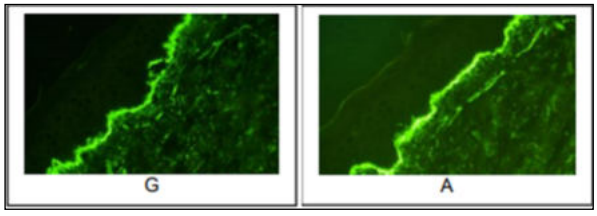

Figure 5: IgG, IgM 3PLUS linear positivity along dermo epidermal junction

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