MIMICS OF TEN
Dr. Rajkumar Kannan DDVL, M.D (DVL)
Associate Professor, Department Of DVL, Chengalpattu Medical College.

ABSTRACT:
Toxic Epidermal necrolysis (TEN) is an Adverse Drug Reaction. Many dermatological conditions may mimic TEN, posing a challenge for the clinician. We would like to highlight the difficulties of establishing differential diagnoses between TEN and "TEN-like" LE and drug reactions.

We report two cases that clinically presented as TEN but turned out to be different entities when we started working out.

KEYWORDS: TEN- like SLE, Adverse drug reaction, Anti-convulsant hypersensitivity syndrome.

INTRODUCTION:
Toxic Epidermal necrolysis is an Adverse Drug Reaction that is clinically characterised by mucocutaneous lesions in the form of mucosal erosions, crustings, raw areas and maculo papular eruptions in the skin. Cutaneous lesions may take a wide array of presentations ranging from mere dusky erythema with skin tenderness, EMF like target and targetoid lesions to sheets of erythema which may coalesce to form large areas of denudation.

CASE 1:
A 30 year old female, home maker presented with complaints of extensive lip erosions and painless oral ulcers. She gave history of over the counter medication for fever three days back. History of fever on and off for the past two months associated with myalgia and arthralgia. History of decreased urine output for the past 3 days. On general examination patient was thin built, febrile, pale with left cervical lymphadenopathy. Dermatological examination revealed extensive lip erosion, vasculitic lesions over the face, ears, palms and soles, erosions over the hard palate (fig.1-fig.4). A provisional diagnosis of SJS/TEN was made. On investigating patient was found to have Hemoglobin - 7.3 g/dl, TC-2400, Platelets – 60000. Peripheral Smear showed mild microcytic hypochromic anaemia. Her blood sugar was 96mg%, serum sodium bicarbonate levels were 23mEq/L, ESR was 118 at one hour and CRP - 12 mg. Her renal and liver function tests were normal. She was found to be ANA positive with low C3 (0.8) and C4(0.08) levels. Renal biopsy showed ISN/ RPS Class II – Mesangial proliferative glomerulonephritis.

Though the clinical lesions were suggestive of SJS/TEN, on investigating she was found to be a case of systemic lupus erythematosus with class II lupus nephritis.

DISCUSSION:
TEN-like rash of lupus, is clinically similar to rash of drug-induced TEN. Less than 50 cases of TEN like SLE has been reported worldwide[1]."Acute Syndrome of Apoptotic Pan-Epidermolysis" (ASAP) is characterized by hyperacute epidermal basal cell apoptotic injury resulting in massive cleavage of the epidermis. It is seen in Toxic Epidermal Necrolysis (TEN), but occasionally occurs in "TEN-like" diseases (SLE, GvHD, pseudoporphyria)[2]. It clinically resembles drug-induced TEN, but differs widely in terms of etiopathogenesis. It has been proposed that exaggerated keratinocyte apoptosis due to increase in Fas expression is the underlying mechanism of many of the cutaneous manifestations of LE.[3,4] TEN occurs with increased frequency in connective tissue disease hence it is difficult to differentiate TEN-like lesions of LE from drug-induced TEN.[5,6] Histopathology of both TEN and TEN like LE shows full-thickness epithelial necrosis and sparse superficial lymphocytic infiltrate. Both conditions respond to steroids. TEN-like rash of LE can be distinguished from drug-induced TEN
DISCUSSION:

The drug hypersensitivity syndrome[7,8] also known as Drug Rash with Eosinophilia and Systemic Symptoms (DRESS) syndrome[9], or as Drug-Induced Delayed Multi Organ Hypersensitivity Syndrome (DIDMOHS) [10] has been reported with the anticonvulsants phenytoin, carbamazepine, Phenobarbital and lamotrigine. This syndrome should be distinguished from drug induced pseudolymphoma syndrome which is also caused by anti convulsants.

Incidence is 2.8% which implies an incidence of 1 in 1000 to 1 in 10000 exposures. The aromatic anti convulsants which are said to be relatively safer than other anti convulsants have also been implicated in the etiopathogenesis of this otherwise fatal syndrome, unless detected and treated early with heroic multi disciplinary interventions.

Clinical features of this syndrome includes fever, mucocutaneous eruptions, lymphadenopathy and hepatitis, one to three months after anti-convulsant therapy. Multi organ involvement can occur with renal and pulmonary lesions[11-15]. Muco-cutaneous eruptions, lymphadenopathy and hepatitis can occur. The lesion may progress into TEN. Generalized pustules may be a manifestation of anti convulsant hypersensitivity[16].

These drugs can cross react with each other in inducing anti convulsant hypersensitivity syndrome. 40-80% cross reactivity has been reported. A severe form of hypersensitivity vasculitis with extensive visceral involvement and poor prognosis is encountered with aromatic anti-convulsants. These anti-convulsants are oxidised by cytochrome P-450 enzyme system into potentially reactive toxic arene oxide intermediates. This syndrome is associated with an inherited deficiency of hepatic microsomal epoxide hydrolase, which converts such reactive intermediates to non toxic dihydrodiols. There may be associated reactivation of herpes viruses[16], and/or ethnic predisposition with certain HLA subtypes[17].

CONCLUSION:

Many dermatological conditions with extensive epidermal necrosis may mimic TEN. A high degree of suspicion, timely diagnosis and dedicated multi-disciplinary intervention could really be life saving to the patients.

REFERENCES:

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