The Indian Journal of Dermatology, Venereology and Leprology (IJDVL) is a bimonthly publication of the Indian Association of Dermatologists, Venereologists and Leprologists (IADVL) and is published for IADVL by Medknow Publications.

The Journal is indexed/listed with Science Citation Index Expanded, PUBMED, EMBASE, Bioline International, CAB Abstracts, Global Health, DOAJ, Health and Wellness Research Center, SCOPUS, Health Reference Center Academic, InfoTrac One File, Expanded Academic ASAP, NIWI, INIST, Uncover, JADE (Journal Article Database), IndMed, Indian Science Abstract’s and PubList.

All the rights are reserved. Apart from any fair dealing for the purposes of research or private study, or criticism or review, no part of the publication can be reproduced, stored, or transmitted, in any form or by any means, without the prior permission of the Editor, IJDVL.

The information and opinions presented in the Journal reflect the views of the authors and not of the IADVL or its Editorial Board or the IADVL. Publication does not constitute endorsement by the journal. The IJDVL and/or its publisher cannot be held responsible for errors or for any consequences arising from the use of the information contained in this journal. The appearance of advertising or product information in the various sections in the journal does not constitute an endorsement or approval by the journal and/or its publisher of the quality or value of the said product or of claims made for it by its manufacturer.

The Journal is printed on acid free paper.
A clinicoepidemiological study of polymorphic light eruption
Lata Sharma, A. Basnet

A clinico-epidemiological study of PLE was done for a period of one year to include 220 cases of PLE of skin type between IV and VI. The manifestation of PLE was most common in housewives on sun exposed areas. Most of the patients of PLE presented with mild symptoms and rash around neck, lower forearms and arms which was aggravated on exposure to sunlight. PLE was more prevalent in the months of March and September and the disease was recurrent in 31.36% of cases.

Comparative study of efficacy and safety of hydroxychloroquine and chloroquine in polymorphic light eruption: A randomized, double-blind, multicentric study
Anil Pareek, Uday Khopkar, S. Sacchidanand, Nitin Chandurkar, Geeta S. Naik

In a double-blind randomized, comparative multicentric study evaluating efficacy of antimalarials in polymorphic light eruption, a total of 117 patients of PLE were randomized to receive hydroxychloroquine and chloroquine tablets for a period of 2 months (initial twice daily dose was reduced to once daily after 1 month). A significant reduction in severity scores for burning, itching, and erythema was observed in patients treated with hydroxychloroquine as compared to chloroquine. Hydroxychloroquine was found to be a safe antimalarial in the dosage studied with lesser risk of ocular toxicity.
Many faces of cutaneous leishmaniasis
Arfan Ul Bari, Simeen Ber Rahman

Symptomatic cutaneous leishmaniasis is diverse in its presentation and outcome in a tropical country like Pakistan where the disease is endemic. The study describes the clinical profile and atypical presentations in 41 cases among 718 patients of cutaneous leishmaniasis. Extremity was the most common site of involvement and lupoid cutaneous leishmaniasis was the most common atypical form observed. Authors suggest that clustering of atypical cases in a geographically restricted region could possibly be due to emergence of a new parasite strain.

Forehead plaque: A cutaneous marker of CNS involvement in tuberous sclerosis
G. Raghu Rama Rao, P. V. Krishna Rao, K. V. T. Gopal, Y. Hari Kishan Kumar, B. V. Ramachandra

In a retrospective study of 15 patients of tuberous sclerosis, eight patients had central nervous system involvement. Among these 8 cases, 7 cases had forehead plaque. This small study suggests that presence of forehead plaque is significantly associated with CNS involvement.

Ligand-binding prediction for ErbB2, a key molecule in the pathogenesis of leprosy
Viroj Wiwanitkit

SCORTEN: Does it need modification?
Col. S. S. Vaishampayan, Col. A. L. Das, Col. R. Verma

Universal acquired melanosis (Carbon baby)
P. K. Kaviarasan, P. V. S. Prasad, J. M. Joe, N. Nandana, P. Viswanathan

Adult onset, hypopigmented solitary mastocytoma: Report of two cases
D. Pandhi, A. Singal, S. Aggarwal
Incidental finding of skin deposits of corticosteroids without associated granulomatous inflammation: Report of three cases
Rajiv Joshi ..................................................................................................................................................................... 44

Erythromelanosis follicularis faciei et colli: Relationship with keratosis pilaris
M. Augustine, E. Jayaseelan ........................................................................................................................................ 47

Naxos disease: A rare occurrence of cardiomyopathy with woolly hair and palmoplantar keratoderma
R. Rai, B. Ramachandran, V. S. Sundaram, G. Rajendren, C. R. Srinivas................................................................. 50

Granular parakeratosis presenting with facial keratotic papules
R. Joshi, A. Taneja ......................................................................................................................................................... 53

LETTERS TO THE EDITOR

Extragenital lichen sclerosus of childhood presenting as erythematous patches
N. G. Stavrianeas, A. C. Katoulis, A. I. Kanelleas, E. Bozi, E. Toumbis-Ioannou ..................................................... 59

Leukocytoclastic vasculitis during pegylated interferon and ribavirin treatment of hepatitis C virus infection
Esra Adisen, Murat Dizbay, Kenan Hize, Nilsel Ilter .................................................................................................. 60
Poland’s syndrome
Saurabh Agarwal, Ajay Arya

Hereditary leiomyomatosis with renal cell carcinoma
Sachin S. Soni, Swarnalata Gowrishankar, Gopal Kishan Adikey, Anuradha S. Raman

Infantile onset of Cockayne syndrome in two siblings
Prerna Batra, Abhijeet Saha, Ashok Kumar

Multiple xanthogranulomas in an adult
Surajit Nayak, Basanti Acharjya, Basanti Devi, Manoj Kumar Patra

Bullous pyoderma gangrenosum associated with ulcerative colitis
Naik Chandra Lal, Singh Gurcharan, Kumar Lekshman, Lokanatha K

Sporotrichoid pattern of malignant melanoma
Ranjan C. Rawal, Kanu Mangla

Acitretin for Papillon-Lefèvre syndrome in a five-year-old girl
Didem Didar Balci, Gamze Serarslan, Ozlem Sangun, Seydo Homan

Bilateral Becker’s nevi
Ramesh Bansal, Rajeev Sen

Madarosis: A dermatological marker
Silonie Sachdeva, Pawan Prasher
The copies of the journal to members of the association are sent by ordinary post. The editorial board, association or publisher will not be responsible for non-receipt of copies. If any of the members wish to receive the copies by registered post or courier, kindly contact the journal’s/publisher’s office. If a copy returns due to incomplete, incorrect or changed address of a member on two consecutive occasions, the names of such members will be deleted from the mailing list of the journal. Providing complete, correct and up-to-date address is the responsibility of the members. Copies are sent to subscribers and members directly from the publisher’s address; it is illegal to acquire copies from any other source. If a copy is received for personal use as a member of the association/society, one cannot resale or give-away the copy for commercial or library use.
ABSTRACT

Background: Toxic epidermal necrolysis (TEN) is a drug induced acute life threatening condition with mortality ranging from about 15 to 60%. A ‘severity of illness’ score termed as SCORTEN has been developed to predict mortality in TEN cases at the time of admission. It is calculated by giving one point for each of predetermined seven variables, evaluated during first 24 hours of admission. Total score ranging from 1-7 predicts a probability of mortality from 0.03 to 0.90. Aim: A prospective study was conducted to analyze efficacy of ‘SCORTEN’ in TEN cases to predict mortality during their management. Methods: All cases of TEN reporting for management to the hospital were assessed using ‘SCORTEN’ on day one and day five to predict probable mortality, this data was then compared with ultimate outcome. Results: During the study period, we treated 10 cases of TEN, all induced by drugs, patient’s age ranging from 03 to 70 years and body surface area (BSA) involvement from 10 to 95%. Three cases succumbed to death. These cases were analyzed with SCORTEN to predict probability of mortality at the time of admission and day five. We encountered some variations from the original study. It was observed that if patients are analyzed with SCORTEN on a daily/alternate day basis, it will serve as a better predictor of mortality. Conclusion: Body surface area (BSA) involvement and age probably need more weightage in calculations. Besides malignancy, tuberculosis and pre-existing diabetes also need to be included while predicting mortality.

Key Words: Toxic epidermal necrolysis, SCORTEN, Predictors of mortality

INTRODUCTION

Toxic epidermal necrolysis (TEN) is a drug related, acute life threatening dermatological disease. Apoptosis of cells causes erosion of mucous membranes, extensive detachment of epidermis and severe constitutional symptoms. Stevens-Johnson syndrome (SJS) and TEN are considered variants within a continuous spectrum, SJS or SJS-TEN overlaps being milder forms. This classification is based on percentage of denuded skin.[1]

The incidence of TEN is reported to be 0.4-2 cases per million populations per year.[2] In India exact incidence of this near fatal disease is not known and seems to be increasing due to indiscriminate use of drugs. This severe disease is reported to have a mortality rate ranging from 15 to 40% with frequent disability in survivors. Various prognostic factors already known are advancing age, maximal body surface area (BSA) detachment and increased blood urea levels.[3]

Another major cause of death in TEN has been reported to be bronchial epithelial detachment.

There are several severity-of-illness scores being used in ICUs to estimate the probability of hospital mortality. Specific scores have been developed for burn patients (age plus percentage of BSA burned) since general ICU scores can not be extrapolated on them and so is the case of acute dermatoses like TEN. A score termed as SCORTEN was developed by Basutji-Gatin et al. in year 2000, as a severity-of-illness score for TEN.[4] It is a validated predictor of mortality in TEN patients when seen at the time of admission. The score is calculated by giving 01 point for each of the following 07 clinical variables during the first 24 hours of evaluation;

• Age more than 40 years
• Malignancy
• Heart rate > 120/minute
• Initial epidermal detachment >10% of BSA
• Serum urea level > 28 mgm/dl (40 mgm/dl in Indian settings)
• Serum glucose levels >250 mg/dl and
• Serum bicarbonate levels <20 mEq/dl.

The probability of death predicted by this score is as follows:
0-1 points- 0.03; 2 points- 0.12; 3 points- 0.35; 4 points- 0.58;
5 to 7 points- 0.90. A probability of 0.90 means approximate
90 of 100 patients with TEN are expected to die. SCORTEN has
been used extensively in large number of studies with good
to excellent accuracy in predicting death in TEN cases. Since
this score is relatively new, a study was conducted to find out
applicability of SCORTEN in TEN patients in Indian settings.

METHODS

A prospective study was carried out at Command Hospital, Pune
for 24 months. A total of 10 cases were diagnosed to have TEN/
SJS-TEN out of nearly 23000 cases seen in OPD during the study
period. All the cases diagnosed to have TEN, were subjected
to a standard battery of ICU protocol of investigations on day
one, following which each case was analyzed using SCORTEN.
Same battery of tests and analysis were also carried out on day
five and prediction of mortality on both days as predicted by
SCORTEN was later compared with final outcome.

RESULTS

Since total number of patients was only 10 statistical
analysis could not be done. We treated 10 cases of TEN with
age ranging from 03 to 70 years and BSA involvement being
15 to almost 100% Anticonvulsants were the commonest
(5/10) drugs implicated, anti-tuberculous therapy being the
next (2/10) common causative drug category. Paracetamol
was thought to be the precipitating cause of TEN in one
case. Many patients were on multiple drugs.

Females outnumbered males (6:4) as reported in some text
books. Details of patient data (including SCORTEN and
mortality data) are given below [Tables 1 and 2]. Out of
the ten patients, 5 had stopped probable offending drug
themselves before reporting to us, but 2 of them died. In
the remaining 5, probable offending drugs (mostly multiple)
were stopped by us immediately on reporting.

---

Table 1: Clinical profile of patients

<table>
<thead>
<tr>
<th>No</th>
<th>Age/sex</th>
<th>Time from the onset in days</th>
<th>Offending drug</th>
<th>Medical history</th>
<th>Category of ACDR</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>70/M</td>
<td>12</td>
<td>Antituberculous therapy</td>
<td>TB, DM</td>
<td>TEN</td>
</tr>
<tr>
<td>2</td>
<td>03/F</td>
<td>02</td>
<td>Phenytoin</td>
<td>--</td>
<td>SJS-TEN</td>
</tr>
<tr>
<td>3</td>
<td>12/M</td>
<td>10</td>
<td>Pyrimethamine-sulphamethoxypyridazine</td>
<td>--</td>
<td>TEN</td>
</tr>
<tr>
<td>4</td>
<td>35/F</td>
<td>03</td>
<td>Carbamazepine</td>
<td>--</td>
<td>SJS-TEN</td>
</tr>
<tr>
<td>5</td>
<td>19/F</td>
<td>02</td>
<td>Carbamazepine</td>
<td>--</td>
<td>TEN</td>
</tr>
<tr>
<td>6</td>
<td>45/F</td>
<td>01</td>
<td>Carbamazepine</td>
<td>DM, HTN, Bell's Palsy</td>
<td>TEN</td>
</tr>
<tr>
<td>7</td>
<td>38/M</td>
<td>08</td>
<td>Paracetamol</td>
<td>DM, TB</td>
<td>TEN</td>
</tr>
<tr>
<td>8</td>
<td>70/M</td>
<td>01</td>
<td>Furosemide</td>
<td>DM, HTN, CAD,</td>
<td>SJS-TEN</td>
</tr>
<tr>
<td>9</td>
<td>20/F</td>
<td>02</td>
<td>Rifampicin</td>
<td>TB</td>
<td>TEN</td>
</tr>
<tr>
<td>10</td>
<td>08/F</td>
<td>03</td>
<td>Carbamazepine</td>
<td>--</td>
<td>SJS-TEN</td>
</tr>
</tbody>
</table>

TB - Tuberculosis, DM - Diabetes mellitus, HTN - Hypertension, CAD - Coronary artery disease, ACDR - Adverse cutaneous drug reaction

Table 2: Patient data

<table>
<thead>
<tr>
<th>No</th>
<th>Age (yrs)</th>
<th>BSA Involved</th>
<th>HR</th>
<th>BUN</th>
<th>BSL</th>
<th>HCO3</th>
<th>SCORTEN</th>
<th>Outcome (days)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>D1</td>
<td>D5</td>
<td>D1</td>
<td>D5</td>
<td>D1</td>
<td>D5</td>
<td>D1</td>
</tr>
<tr>
<td>1</td>
<td>70</td>
<td>40</td>
<td>70</td>
<td>↑</td>
<td>N</td>
<td>N</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>2</td>
<td>03</td>
<td>15</td>
<td>&lt;10</td>
<td>↑</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>3</td>
<td>12</td>
<td>80</td>
<td>100</td>
<td>↑</td>
<td>↑</td>
<td>N</td>
<td>↑</td>
<td>↓</td>
</tr>
<tr>
<td>4</td>
<td>35</td>
<td>20</td>
<td>30</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>5</td>
<td>19</td>
<td>30</td>
<td>20</td>
<td>↑</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>6</td>
<td>45</td>
<td>10</td>
<td>80</td>
<td>↑</td>
<td>↑</td>
<td>N</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>7</td>
<td>38</td>
<td>70</td>
<td>100</td>
<td>↑</td>
<td>↑</td>
<td>N</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>8</td>
<td>70</td>
<td>15</td>
<td>&lt;10</td>
<td>↑</td>
<td>N</td>
<td>N</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>9</td>
<td>21</td>
<td>50</td>
<td>30</td>
<td>↑</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>10</td>
<td>08</td>
<td>20</td>
<td>15</td>
<td>↑</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
</tbody>
</table>

BSA - Body surface area, HR - Heart rate, BUN - Blood urea, BSL - Blood sugar level, D1 - Day 1, D5 - Day 5, S - Survived, ↑ - Rise, ↓ - Fall, N - Within normal range
DISCUSSION

After analyzing above data it is clear that none of the patients had malignancy but 4 out of 10 had pre-existing diabetes mellitus and 03 of them died. Tuberculosis was seen in 3 patients and 2 out of 3 died. Thus in Indian sub-continent probably these systemic diseases and other severe systemic illnesses need to be given weightage equal to malignancy. Age had no effect on the ultimate mortality but more male 3/4 died as compared to females 1/6. As is well known, ultimate cause of death was septicemia in all 4 cases who died, despite best possible management in a tertiary care hospital.

In SCORTEN analysis, % of BSA involvement entails 01 point, equal to other six parameters, however it is a well known fact (as observed in this study too) that BSA involvement of >40% had very poor prognosis. Therefore BSA involvement of >40% should be given more weightage as in burn scores.

Analysis of data given in Table numbers 02 and 03 reveals that patient number 1, 3, 5 and 6 who ultimately died had SCORTEN of either 3 or 4 on day 01 (these scores have a predicted mortality of 35 to 50%) and in all of them SCORTEN increased to 5 or 6 on day 05 (at these scores prediction of mortality rate rises to more than 90%). Thus it is apparent that expected mortality based on SCORTEN of day 05 is more accurate than SCORTEN calculated on first day.

Though this study involves only 10 cases, following amendment are recommended keeping in mind the conditions prevalent in the Indian sub-continent. a) Percentage (%) of BSA involvement be given graded points e.g. 10-30% = 01 point, >30% = 02 points. b) Besides malignancy other systemic diseases like pre-existing diabetes mellitus, tuberculosis, cardiac disorders, other severe chronic diseases should also be included while calculating SCORTEN. c) SCORTEN analysis should be done on first as well as fifth day to get more accurate picture and prognosis.

Larger and longer studies are required to further authenticate and confirm these observations so that a modified and better SCORTEN may evolve.

REFERENCES


<table>
<thead>
<tr>
<th>SCORTEN</th>
<th>No. of Pts</th>
<th>Expected Mortality (based on SCORTEN)</th>
<th>Observed Mortality (based on SCORTEN)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>D1</td>
<td>D5</td>
<td>%</td>
</tr>
<tr>
<td>0-1</td>
<td>1</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>2</td>
<td>4</td>
<td>1</td>
<td>12</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>0</td>
<td>35</td>
</tr>
<tr>
<td>4</td>
<td>2</td>
<td>1</td>
<td>58</td>
</tr>
<tr>
<td>&gt;5</td>
<td>-</td>
<td>3</td>
<td>90</td>
</tr>
</tbody>
</table>

Vaishampayan, et al.: SCORTEN