**Case Report**

**A Case of Deadly “TEN”**

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**Abstract**

A 50 year old male, presented to us with fever and severe skin and mucous membrane lesions, and was diagnosed to be suffering from a deadly medical emergency: Toxic Epidermal Necrolysis (TEN) which has been discussed in detail.

**Introduction**

Toxic Epidermal Necrolysis (TEN; also known as “Lyell’s syndrome”) is a rare, life-threatening dermatological condition that is usually induced by a reaction to medications. It is characterized by the detachment of the epidermis from the dermis all over the body.

There is a broad agreement in medical literature that TEN can be considered as a more severe form of Stevens-Johnson syndrome (SJS). Some authors consider that there is an overlap between the two syndromes. The incidence is between 0.4 and 1.3 cases per million each year.

TEN affects many parts of the body, but it most severely affects the mucous membrane, such as the mouth, eyes and vagina. The severe findings of TEN are often preceded by 1 to 2 weeks of fever. These symptoms may mimic those of a common upper respiratory tract infection. When the rash appears, it may be over large and in varied parts of the body, and it is usually warm and appears red. The skin then begins to sag from the body and can be peeled off in great swaths. The mouth becomes blistered and eroded, making eating difficult and sometimes necessitating nasogastric tube or gastric tube directly into the stomach. The eyes are affected, becoming swollen, crusted and ulcerated and blindness may occur.

**Case Report**

A 50-year-old male, known chronic alcoholic, was admitted in our hospital for fever of 1 week duration. He had no other comorbid illness. On the day of admission, there was diffuse erythematous rash involving the entire body. He also gave history of difficulty in swallowing.

He had a history of severe anemia one month prior to admission and had been treated with five units of fresh blood transfusion. The cause of anaemia was evaluated due to bleeding haemorrhoids and he also underwent haemorrhoidectomy 20 days earlier.

During this admission, his blood count showed low total count (<1100cells/cumm) with neutrophil count less than 400 (neutropaenia). During the next few days, he developed extensive skin blisters over his chest, back and arms (Figures 1–5). He also developed severe oral ulcers with mucosal desquamations (Figure 6), severe crusted conjunctivitis that became toxic.

He was diagnosed to be suffering from TEN with febrile neutropaenia.

The patient underwent extensive investigations including marrow aspiration and HIV status, and we also reviewed the medicines he had received.

Biopsy of the skin showed cell death throughout the epidermis and necrolysis of keratinocytes (Figure 7).

He was treated with:
1. Intravenous steroids
2. Intravenous azithromycin and linezolid
3. Injection Filgrastim Granulocyte Macrophage Colony Stimulating Factor (G-CSF)
4. Intravenous immunoglobulin (IVlg)

On the 14th day, renal shut down occurred, fever persisted, septicemia with septic shock set in and the patient died on the 16th day of admission.

**Discussion**

**Cause**

TEN is a rare and usually severe adverse reaction to certain drugs. History of medication use exists in more than 95% of patients with TEN. The drugs most often implicated in TEN are sulfonamides, non-steroidal anti-inflammatory drugs, allopurinol, antimetabolites (methotrexate), antiretroviral drugs, corticosteroids, chloromezanone (anxiolytic) and anticonvulsants, such as phenobarbital, phenytoin, carbamazepine and valproic acid.

The condition might also result from infection with agents such as *Mycoplasma pneumoniae* or the herpes virus; and transplants of bone marrow or organs. Candidates with SLE and underlying malignancy are more prone to TEN.

**Pathogenesis**

Microscopically, TEN causes cell death throughout the epidermis. Keratinocytes, which are the cells found in the lower epidermis specialized in holding the skin cells together, undergo necrosis (cell death).

**Diagnosis**

Often, the diagnosis can be made clinically. Generally, if the clinical history is consistent with SJS, and the skin lesions cover more than 30% of the body surface area, the diagnosis of TEN is appropriate. Sometimes, however, examination of affected tissue under the microscope may be needed to distinguish it from other entities, such as staphylococcal scalded skin syndrome. Typical histological criteria of TEN include mild infiltrate of lymphocytes which may obscure the dermoepidermal junction and prominent cell death with basal vacuolar change and individual cell necrosis. Nikolsky’s sign is almost always present in TEN (peeling of skin, as rolling over by fingers).

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Prognosis

TEN proper (with less than 10% of body surfaces area involved) has a mortality rate of around 5%. The risk for death can be estimated using the “SCORE of TEN” (SCORTEN) scale, which takes a number of prognostic indicators into account.

The SCORTEN scale is a severity-of-illness scale with which the severity of certain bullous conditions can be systematically determined. It was originally developed for TEN, but can be used for burn victims, sufferers of SJS, cutaneous drug reactions, or exfoliative wounds. These conditions have in common that they compromise the integrity of the skin and/or mucous membranes.

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The term “SCORTEN” stands for SCORE of “Toxic Epidermal Necrolysis”.

Treatment

The first line of treatment is early withdrawal of culprit drugs, early referral and management in burn units or intensive care units, supportive management, and nutritional support.

The second line is IVIg — uncontrolled trials showed promising effect of IVIg on treatment of TEN; a randomized control trial is needed in future to determine the efficacy of IVIg in TEN.

The third line is cyclosporine, cyclophosphamide, plasmapheresis, pentoxifylline, N-acetylcysteine, ulinastatin, infliximab and/or G-CSF (if TEN-associated leukopaenia exists). Systemic steroids are unlikely to offer any benefits.

Conclusion

The mortality due to TEN is 30–40%. Loss of skin makes the patient vulnerable to infections from fungi and bacteria, and can result in sepsis, the leading cause of death in the disease.
Death is caused either by infection or by respiratory distress which is due to either pneumonia or damage to the linings of the airway. Microscopic analysis of tissue (especially the degree of dermal mononuclear inflammation and the degree of inflammation in general) can play a role in determining the prognosis of individual cases. The medico legal implications of TEN/SJS are obvious. The disease can be severe with a high mortality. The initial presenting signs and symptoms may be mild leading to an incorrect diagnosis. We feel an appropriate antibiotic composition with aggressive intensive nursing care with proper and adequate hydration similar to severe burns patient, could have saved life of this patient.

This case report is published to create awareness among the practicing physicians. A high index of suspicion combined with a detailed history and physical examination should lead to early diagnosis2.

<table>
<thead>
<tr>
<th>Risk Factor</th>
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<tbody>
<tr>
<td>Age</td>
<td>&lt;40 years</td>
<td>&gt;40 years</td>
</tr>
<tr>
<td>Associated malignancy</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Heart rate (beats/min)</td>
<td>&lt;120</td>
<td>&gt;120</td>
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<tr>
<td>Serum BUN (mg/dL)</td>
<td>&lt;27</td>
<td>&gt;27</td>
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<tr>
<td>Detached or compromised body surface</td>
<td>&lt;10%</td>
<td>&gt;10%</td>
</tr>
<tr>
<td>Serum bicarbonate (mEq/L)</td>
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<td>&lt;20</td>
</tr>
<tr>
<td>Serum glucose (mg/dL)</td>
<td>&lt;250</td>
<td>&gt;250</td>
</tr>
</tbody>
</table>

Table 1 : In the SCORTEN Scale, 7 independent risk factors for high mortality are systematically scored, so as to determine the mortality rate for that particular patient.

Table 2 : The more risk factors present, the higher the SCORTEN score, and the higher the mortality rate, as shown in the following table.

<table>
<thead>
<tr>
<th>No of risk factors</th>
<th>Mortality rate</th>
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<tbody>
<tr>
<td>0-1</td>
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<tr>
<td>2</td>
<td>12.1%</td>
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<tr>
<td>3</td>
<td>35.3%</td>
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<tr>
<td>4</td>
<td>58.3%</td>
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<tr>
<td>5 or more</td>
<td>&gt;90%</td>
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References