

Toxic Epidermal Necrolysis (TEN) – SCORTEN Scale predicting mortality

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Introduction

Toxic Epidermal Necrolysis (TEN) is a rare potentially life-threatening mucocutaneous blistering reaction. It is nearly always caused by drug reactions, and is characterised by widespread blistering arising from erythematous or purpuric macules with mucosal involvement. Diagnosis is confirmed with a skin biopsy. It affects all age groups, both sexes and all races. TEN lies on a continuum with Stephen Johnson Syndrome (SJS). The total body surface area (TBSA) involvement in TEN is >30%, whereas in SJS it is <10%. Between 10 and 30% is considered a TEN/SJS overlap.

Case

A 34 year-old man developed a temperature, cough and sore throat 3 weeks after starting lamotrigine. He was admitted to a specialist burns unit in London with a maculopapular rash and blistering on his trunk that covered 100% of his TBSA. He had a SCORTEN score of 2 (bicarbonate >20mmol/l). A biopsy confirmed TEN. 5 days later, he developed epidermal sloughing. He deteriorated further, developing respiratory distress, and required a tracheostomy for long term ventilation. He was given fluids, antibiotics, intravenous immunoglobulins (IVIg), ciclosporin and granulocyte-colony stimulation factor (G-CSF). Despite these measures, he died 60 days after admission.

SCORTEN

A predictor of mortality in TEN and SJS. It is calculated within 24 hours of admission. There are seven criteria.

SCORTEN	Predicted mortality
0-1	>3.2%
2	>12.1%
3	>35.3%
4	>58.3%
>5	>90%

Figure 1. Positive Nikolsky's sign



Figure 2. 100% TBSA skin loss



Figure 3. Porcine xenograft



Complications

In the acute setting, patients may develop:

- Dehydration and hypothermia
- Infection
- Shock and multiorgan failure
- Thromboembolism and DIC

** IVIg, ciclosporin, plasmaphoresis, biologics and G-CSF have been associated with enhanced healing, but their effectiveness is uncertain.

Conclusion

This case and case series demonstrates that the SCORTEN score can help to predict mortality in patients with TEN. Antibiotics and anti-epileptic drugs most commonly precipitate this potentially life-threatening reaction. Prompt action is necessary but may not necessitate a good outcome. Current evidence is uncertain of the effectiveness of immunoglobulins, biologics and G-CSF.

References

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Case series

We analysed 11 other cases of TEN during 1999-2010.

	Mean	Range
Time from onset to admission	6 days	1-19 days
Length of admission	33 days	8-84 days
Total body surface area	72%	50-100%
SCORTEN	2	0-2

Precipitating drugs: Lamotrigine, Carbamazepine (x3), Phenytoin, Sulfasalazine (x2), Nevirapine, Tamiflu, Flucloxacillin, Hydroxychloroquine, Lanzoprazole.

3 patients died during admission. Of these, one did not have a SCORTEN recorded and the other two had a SCORTEN of 2. The mortality for this cohort was 25%.

Principles of Management

- Stop the causative drug
- Fluid and nutrition replacement (IV and NG routes) and catheter
- Temperature maintenance (consider skin graft)
- Pain relief
- Sterile handling and isolation
- Skin care, mouth care, eye care, lung care
- Regular assessment for infection