

STEVEN–JOHNSON SYNDROME (SJS) AND TOXIC EPIDERMAL NECROLYSIS (TEN)

Definitions

Steven Johnson Syndrome (SJS):

- severe erosions of at least two mucosal surfaces with extensive necrosis of lips and mouth, and a purulent conjunctivitis.
- epidermal detachment may occur in SJS, but less than 10% of the body surface area is involved.
- morbidity with this disease is high, and can include photophobia, burning eyes, visual impairment and blindness.

Toxic Epidermal Necrolysis (TEN):

- severe exfoliative disease associated with systemic reaction characterized by rapid onset of widespread erythema and epidermal necrolysis.
- involves more than 30% loss of epidermis.

Aim of treatment:

To remove the cause and prevent complications

Salient features

- acute prodromal flu-like symptoms, fever, conjunctivitis and malaise.
- skin tenderness, morbilliform to diffuse or macular erythema target lesions, vesicles progressing to bullae. Blisters on the face, and upper trunk, then exfoliation with wrinkled skin which peels off by light stroking (Nikolsky' sign).
- buccal mucosa involvement may precede skin lesion by up to 3 days in 30% of cases. Less commonly the genital areas, perianal area, nasal and conjunctival mucosa
- in the gastrointestinal tract, esophageal sloughing is very common, and can cause bleeding and diarrhoea
- in the respiratory tract, tracheobronchial erosions can lead to hyperventilation, interstitial oedema, and acute respiratory disease syndrome
- skin biopsy of TEN - Extensive eosinophilic necrosis of epidermis with subbasal cleavage plane
- renal profile – raised blood urea, hyperkalaemia and creatinine
- glucose - hypoglycaemia

Table 1. Aetiology

Drugs

Antibiotics

sulphonamides, amoxycillin
ampicillin, ethambutol, isoniazid

Anticonvulsants

phenobarbitone, carbamazepine,
phenytoin

Non-Steroidal Anti-Inflammatory Drugs

phenylbutazone, salicylates

Infection

Virus

herpes simplex, enteroviruses,
adenoviruses, measles, mumps

Bacteria

Streptococcus, *Salmonella typhi*
Mycoplasma pneumoniae

Management

Supportive Care

- admit to isolation room where possible
- may need IV fluid resuscitation for shock
- good nursing care (Barrier Nursing and hand washing)
- use of air fluidized bed, avoid bed sores
- adequate nutrition – nasogastric tubes, IV lines, parenteral nutrition if severe mucosal involvement

Specific treatment

- eliminate suspected offending drugs
- IV Immunoglobulins at a dose of 0.4mg/kg/per day for 5 days. IVIG is a safe and effective in treatment for SJS/TEN in children. It arrests the progression of the disease and helps complete re-epithelialization of lesions.

Monitoring

- maintenance of body temperature. Avoid excessive cooling or overheating
- careful monitoring of fluids and electrolytes – BP/PR
- intake / output charts, daily weighing & renal profile

Prevent Complications

Skin care

- cultures of skin, mucocutaneous erosions, tips of Foley's catheter.
- treat infections with appropriate antibiotics.
- topical antiseptic preparations: saline wash followed by topical bacitracin or 10% Chlorhexidine wash
- dressing of denuded areas with paraffin gauze / soffra-tulle
- surgery may be needed to remove necrotic epidermis

Eye care

- frequent eye assessment
- antibiotic or antiseptic eye drops 2 hourly
- synechia should be disrupted

Oral care

- good oral hygiene aimed at early restoration of normal feeds.