Erythema Multiforme (EM)

Background

- Described in 1866 by Ferdinand Von Hebra as an acute self limiting skin disease distributed on the extremities with typical often recurrent concentric target lesions
- Stevens-Johnson syndrome (SJS) was considered an extreme form of EM for many years while toxic epidermal necrolysis (TEN) was considered a different entity
- In 1993, a consensus definition and classification was proposed based on photographic atlas and extent of body surface area involvement
- Essentially SJS and TEN are considered variant of a single disease entity while EM is now considered a different disease entity

EM

Classified into major and minor

- Minor-typical target lesions or raised oedematous papules distributed acrally
- Major-typical target lesions, raised oedematous papules distributed acrally with involvement of one or more mucus membranes or epidermal detachment less than 10% of TBA

SJS/Toxic epidermal necrolysis (TEN)

- Characterised by widespread blisters predominant on the trunk and face presenting with erythematous and oedematous or pruritic macules and one or more mucus membranes erosion.
- There may be epidermal detachment usually less than 1% of total body surface area for SJS and 30% or more for TEN

Pathophysiology

- Appears to involve hypersensitivity reactions triggered by a variety stimuli particularly viral, bacterial or chemical agents
- A major cause is herpes virus especially recent or recurrent infection

Epidemiology

- Global incidence is estimated to be between 0.01-1%
- Male are more affected than females (ratio ranges from 3:2 to 2:1)
- All ages are affected- peak 2nd to 4th decade of life
- 20% in children and adolescents
- Rare under 3 years and over 50 years
- SJS and TEN have equal incidence in males and females

Prognosis

- EM is not usually fatal. Most cases are self- limiting and resolve without any sequelae in 2 to 4 weeks
- SJS has a 5% mortality
- TEN has a 30% mortality

Causative agents

- 50% idiopathic
- EM, SJS and TEN linked to drugs
- Infection-common in children
- Herpes is common in younger patients and is associated with recurrent EM

Infection

Viral

- Herpes simplex 1 and 2
- Adenovirus
- Coxsackie virus
- Echovirus
- Enterovirus
- Epstein-barr
- Hepatitis A and B
- Measles
- Varicella
- Influenza
- Mumps
- polio

bacterial

- Mycoplasma
- Proteus
- Salmonella
- TB
- Vibrio parahaemolyticus
- Psittacosis
- Cat scratch dx
- Brucella
- Tularaemia
- Gonorrhoea
- Typhoid
- Diphtheria
- LGV
- Cholera
- Yersinia enterocoliticus

Causes-continued

Fungal

- Histoplasmosis
- Coccidiodes species

Post vaccination

- BCG
- Oral polio
- Tetanus/diphtheria

Causes

- Drugs
  - Sulphonamide
  - NSAIDS
  - Anticonvulsants
  - Barbiturates
  - Anti TB drugs
  - Antibiotics
  - Pyrazolones
- Malignancy
- Hormonal disorders
- Collagen diseases
- Immune disorders (sarcoidosis, vasculitis, transient selective C4 deficiency
- Physical/Mechanical
  - Tattooing
  - Radiotherapy
  - Cold
  - sunlight

Differential diagnosis

- Behcet's syndrome
- Collagen dx
- Dermatitis herpetiformis
- Drug eruptions
- Figurate erythema
- Granuloma anulare
- Lichen planus
- Meningococcaemia
- Mucocutaneous lymph node syndrome
- Necrotising vasculitis
- Pemphygoid
- Recurrent aphthus ulcer
- Secondary syphilis
- Urticaria

Investigations

These are non-specific

- Raised WCC
- Raised eosinophils (usually >1000/mm3
- Raised ESR –non-specific
- U&E, LFT – may be deranged and indicate organ involvement
- Blood cultures in severe cases
- Herpes simplex virus (HSV) antigen may be detected in keratocytes by immunoflorescence
- HSV DNA may be detected by PCR
- CXR in suspected chest infection
- Skin biopsy in equivocal cases

Treatment in the ED

Usually symptomatic

- Analgesics-NSAIDS
- Cold compress
- Topical steroids
- Oral soothing agents-saline gargles, viscous lidocaine
- Fluid replacement and monitoring
- Empirical antibiotics if infection suspected but prophylactic antibiotic discouraged
- Avoid systemic corticosteroids as prognosis not improved (controversial)
- Treat underlying cause, remove causative agent if identified
- Give acyclovir for herpes-induced EM
- Refer as necessary (dermatology, ICU, paediatrics etc)