ANGAL HOSPITAL

CLINICAL PRESENTATION-MALE WARD

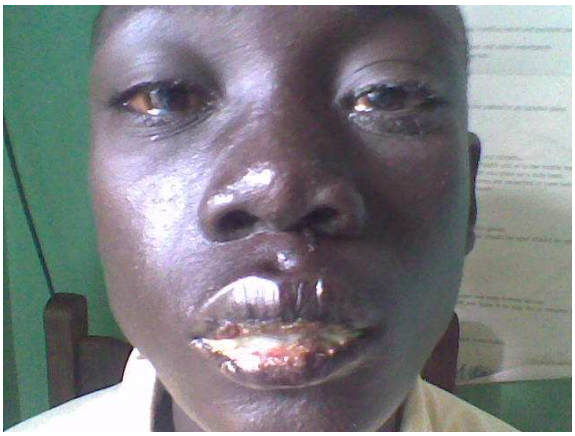
17/04/2010

DR. DAMOI

CASE

15 years old known epileptic presented with h/o generalized body rashes which gradually progressed over 5 days with involvement of the oral mucosae. He developed pus discharge from both eyes and irritation from bright light. No h/o burning minturation.
Past medications: through 2009 he used phenytoin and phenobarbitone (alternated), with no skin rashes reported. He developed frequent fits during the month of january 2010 and recent record shows that he was put on phenytoin 200 mg + carbamazepine 100 mg on the 8/02/2010.
He presented in hospital on the 26/02/2010 with history as above.
O/E: sick looking, T 37.2℃, with purulent eye discharge and reddening of the conjunctiva. There was erosion of the oral mucosa with purulent discharge.
He had erythematous macules most marked on both upper limbs and the face, fewer lesions on the trunk, with crusting and peeling skin on the back (positive Nikolsky's sign), about 28% TBSA involved.
S/E: normal

Stephens-Johnson Syndrome



Is one of the exfoliative/ bullous dermatoses mainly attributted to drugs.
Stevens-Johnson syndrome is also refered to as Erythema Multiforme major(The minor form is also called Hebra's disease)
Some of the sources consider Erythema Multiforme, Stevens-Johnson syndrome and Toxic Epidermal necrolysis(TEN) as one entity, with TEN being the most severe form.

Stevens-Johnson syndrome presents with maculo papular rashes which rapidly progress to form blisters and bullae with subsequent desquamation and large areas of denuded skin.
The areas of desquamation show positive Nikolsky's sign (epidermis is easly removed with slight tangential pressure).
There is involvement of the mucosae, with painful erosions, oral and conjunctival mucosae being the most commonly affected-genital mucosae can also be involved.
There may also be involvement of intestinal and other visceral epithelia.
Associated systemic symptoms include malaise, headache, fever and arthralgia.

Erytema Multiforme is characterized by symmetrically distributed fixed red lesions (maculopapular or bollous) some of which evolve concentric zones of color to form target or iris lesions-which is the hallmark of erytema multiforme. The concentric zones are a central dusky discoloration (bulla), surronded by a pale colored oedematous ring, encircled by erytema.
Oral mucosae lesions occur in only about 25% of cases and is not severe.
Erytema multiforme is a self-limited condition typically lasting only 1 to 4 weeks.

Stevens-Johnson syndrome and Toxic Epidermal Necrolysis are mainly differentiated by the proportion of body surface area affected. SJS-eruptions cover ≤ 30% TBSA, TEN > 50% TBSA, SJS-TEN overlap is when 30-50% TBSA is affected.

ETIOLOGY
SJS and TEN are mainly due to adverse drug reactions, thought to be the result of toxic cutaneous injury.
Most commonly implicated drugs include:

- -sulphonamides
- -anticonvulsants: phenitoine, carbamazepine, phenobarbitone and lamotrigine
- -antibiotics: penicillins, trimethoprim, cefalosporins, ciprofloxacin, doxycycline, erythromycine, vancomycine
- -allopurinol

However SJS may also be precipitated by infectious agents including HSV and Mycoplasma pneumoniae.

For Erytema Multiforme on the other hand the most common precipitating factor is antecedent herpes simplex virus infection, the disease typically develops within 2 days to 3 weeks of an episode of HSV.
EM may be attributed to drugs in a minority of cases.

Skin biopsy and clinical hystory will usually differentiate between SJS/TEN and the other close differentials of Staphylococcal Scalded Skin Syndrome (SSSS), Pemphigus vulgaris, Graft versus Host disease, acute generalized exanthematous pustolosis, thus the importance of sending a skin sample for histology.

In SJS/TEN the histological finding is that of full thickness necrosis of the epidermis.

SSSS-affects mainly neonates and infants under 2 years , however adults can also be affected e.g. those with renal failure.
The histological finding in SSSS is that of necrosis which involves only the superficial corneal layer of the skin.

Acute generalized exantematous pustolosis, commonly associated with hydroxychloroquine-histological finding is of spongiforms superficial intraepidermal pustules (but epidermis is viable), papillary dermal oedema, perivascular dermal infiltrates with eosinophils and neutrophils.

THERAPY FOR SJS/TEN
Elimination and subsequent avoidance of the causative drug + discontinue all unnecessary drugs.
Treatment of the underlying infections.
Management in burns units depending on the extent of cutaneous involvement.
Carefull attention to secondary infections and the potential development of sepsis.
Skin grafting or synthetic skin dressing may provide protection from bacterial infections.
Periodic culture of involved cutaneous sites & blood and initiation of systemic antibiotics as indicated.
Monitoring and replacement of fluid and electrolytes.
High caloric replacement (2200 Kcal/m2/day)
Good pulmonary hygene, suctioning, postural drainage
Pain management
Prompt ophthalmologic consultation
H2 blocking agents and antiacids to avoid gastrointestinal ulceration
Use of steroids controversial

CASE OUTCOME

Patient was admitted on general ward.
Carbamazepine stopped, phenytoin continued at 300mg od.
Analgesia-codeine phosphate 30 mg 6 hrly along with 10 mg nocte of bisacodyl.
TEO, oral cloxacilline 500 mg 6 hrly over 5/7.
Calamine lotion BD applied over affected sites.
Frequent oral gurgles with warm salt water.
IV N/S 2 liters in the first 24 hours.

Gradually improved and was discharged on 15th day on phenytoin, no fits registered during hospitalisation, no new lesions, the crusted areas beginning to heal.