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Case Note

Toxic epidermal necrolysis masquerading as scald burn

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Toxic epidermal necrolysis (TEN) is a severe and potentially fatal drug reaction characterized by an extensive skin rash with blisters and exfoliation resembling scalding. Mucosal involvement is present in up to 90% of cases. Epidermal detachment is generally greater than 30% of BSA. We present a case of TEN which presented in EM Dept. as painful superficial wound over trunk, limbs and face and was misinterpreted as scald burn.

Case report

A 64-year-old woman presented with painful wound on trunk, upper/lower limb, periorbital area for 4 days. She had h/o increased alcohol intake and over-the-counter medication use (nimusulide, clonazepam, amox-clox, azithromycin, alprazolam).

On examination she was ill-looking with irrelevant talk, dehydrated, tachycardiac. She had superficial partial thickness *like* (second-degree) burn and desquamation of skin over trunk, upper and lower limb, face associated with bullae involving 35% of BSA. The bullae were flaccid with clear fluids. It also involved the conjunctiva, genital area. Chest and abdominal examination were normal. In investigation initially counts, Hb%, RFT, LFT, urine routine, CXR were within normal limit.

The clinical impression was scald burn with alcohol withdrawal syndrome (mild). After admission on 2nd day patient had *new bullous eruption* on hands and lower limbs, which made of suspicious of adverse drug reaction and all the IV antibiotics (cephalosporin) were stopped and patient managed conservatively with IV fluids, O2, steroid, NG feeding, strict output monitoring, daily dressing of wound with normal saline in ICU. 3rd day onwards patient had no new eruptions.

On 8th day of admission, patient suddenly deteriorated with persistent tachycardia, spiky fever, tachypnoea. Investigation showed WBC- 56,700 cells/mm3, polymorphs-90%, repeat blood c/s were negative. Broad spectrum antibiotic was started (meropenem) for underlying occult sepsis and subsequently the patient response to treatment was excellent with epithelization of wound in 3 weeks time. Patient was discharged on 26th day of admission without any medication.

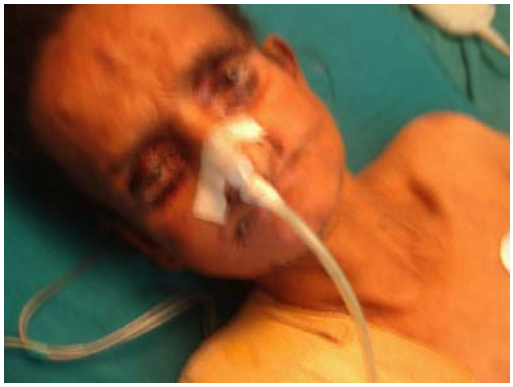

Fig 1: At time of admission

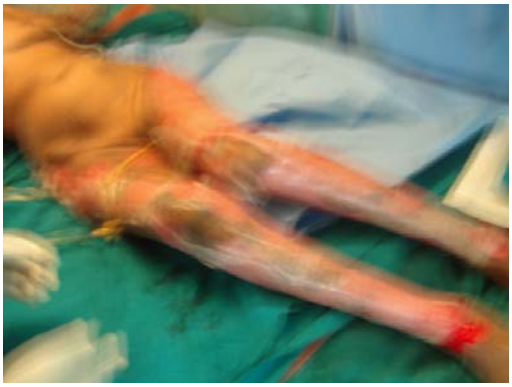

Fig 2: lesion in the legs

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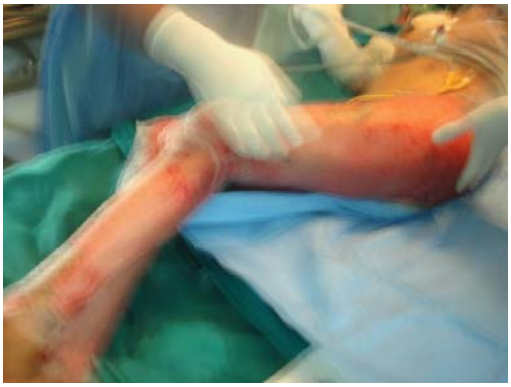

Fig 3: Dressing of the wounds

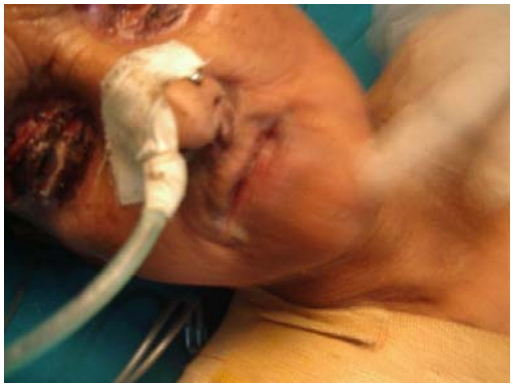

Fig 4: Conjunctivitis and mucositis

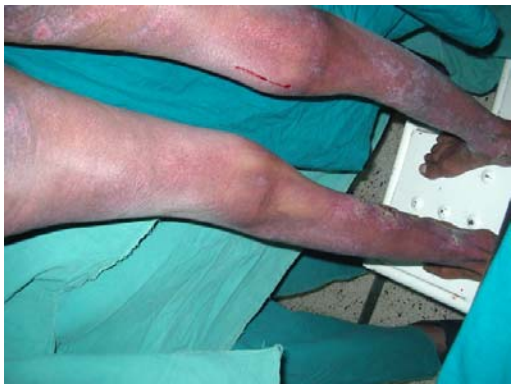

Fig 5: Healing wounds

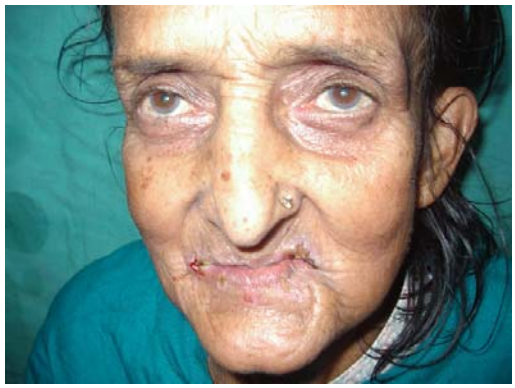

Fig 6: Folllow up after cure

Discussion

TEN is rapidly evolving mucocutaneous reaction characterized by widespread erythema, necrosis, and bullous detachment of epidermis resembling scalding. In majority of cases there is a h/o recent drug ingestion. There is often severe involvement of mucosal surfaces. In some cases there may be extensive involvement of GIT and respiratory system (30%). Reported mortality varies from 30-50%.

Epithelial loss results in vulnerability to bacterial and fungal infections and predisposes to septicaemia, the leading cause of morbidity and mortality. Significant fluid loss from extensive skin lesions as well as inability to tolerate oral intake can lead to hypovolaemia, acute tubular necrosis and shock.

The pathogenesis of TEN is not clear but the basic pathology of large-scale epidermal death results from *apoptosis of keratinocytes* which in turn is due to abundance of TNF in epidermis resulting from reactive drug metabolites. Medications are the major cause of TEN. A causal link is suggested when TEN occurs during the first 4 weeks of medication use. Common medications are sulphonamides, penicillin, NSAIDs, anticonvulsant, allopurinol, antiretroviral.

A prodromal phase of fever, cough and malaise is followed by an acute macular exanthema which rapidly becomes NIkolsky positive (epidermal separation induced by gentle lateral pressure). Increasing age, significant comorbidity, and greater extent of skin involvement correlate with a worse prognosis. *Bastuji-Garin et* al have devised a validated measure of disease severity the SCORTEN. This includes:

Age > 40yrs
Heart rate>120/min
Comorbidity (malignancy)
Involved BSA>10%
BUN>10mmol/L
Serum bicarbonate < 20mmol/L
Blood glucose 14mmol/L

Mortality rate based upon the number of positive criteria are as follows:

0 to 1= 3%
2 factor = 12%
3 factor = 35%
4 factor = 58%
5 or more factor = 90%

The most important aspect of managing TEN are *prompt diagnosis, early withdrawal of all suspect drug(s), good supportive care, specialized nursing care, management in ICU/Burn unit.* Supportive care includes protection of exposed dermis and eroded mucosal surfaces, prevention, early detection and treatment of infection, careful monitoring of fluid and electrolyte status and nutritional support. There is no evidence that prophylactic antibiotics provide benefit and most authors reserve antibiotic for treatment of proven infection. Care must be taken in screening for sepsis and surveillance of lines/catheter to allow prompt intervention.

TEN is highly catabolic state requiring appropriate nutritional support. Energy ad protein requirement appears to be related to amount of BSA affected. Eating may be painful esp. with oral/mucosal involvement and NG tube feeding may be required (enteral feeding is always preferable to parenteral nutrition). Hyperglycemia in critically ill patient has been shown to be associated with increased morbidity and mortality. So strict blood glucose control with insulin sliding scale infusion should be applied. Respiratory support and care is critical; as pulmonary involvement is common. Treatment includes nebulised saline, bronchodilators and physiotherapy. Dressing with normal saline should be done daily. Wound healed completely within 14-21 days with no significant scarring or need for skin grafting.

Immunosupression:

As the initial process in TEN is immunological, there is a good reason to consider the use of immunosuppressant in treatment. These include:

1. Corticosteroids: It has anti-inflammatory, immunosuppressant and antiapoptotic effect. In a retrospective study of 15 patient with TEN treated with parenteral dexamethasone (8mg daily in two divided doses) in Kerela, India, all made an uneventful recovery. But there are also retrospective studies reporting adverse outcomes and enhanced morbidity (infection, GIT bleeding, prolonged hospital stay). Other drugs are ciclosporin, cyclophosphamide but their use is still controversial.
2. Intravenous Immunoglobulin: It has been shown to stop the apoptosis process by addition of human pooled IVIg. The study are conflicting ad any mortality benefit is likely to be small.
3. Others: Zinc, it functions as antioxidant and protects against apoptosis induced by diverse physical, chemical and immunological stimuli including drugs.

Conclusion

In conclusion, TEN is a rare but serious dermatological disorder with significant mortality which needs early recognition, immediate withdrawal of any potential causative agent(s) and management in ICU/Burn unit when TEN is suspected.

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