SELF-ASSESSMENT PROGRAMME

A 10 day old child presented with a three day history of multiple bullous lesions all over the body including the palms and soles. The child was full term, normal delivery, born at home. There was no history of any drugs having been given either to the child or to the mother.

Examination revealed an ill-looking child with multiple bullous and pustular lesions more particularly on the trunk. On the extremities, the skin was coming off in large sheets. The child was toxaemic and died within 12 hours of admission.

1. Which of the following diagnoses is most likely?
   - i. Congenital syphilis
   - ii. Toxic epidermal necrolysis (TEN)
   - iii. Bullous impetigo
   - iv. Epidermolysis bullosa
   - v. Pemphigus neonatorum

2. Which of the following investigations would have been most helpful?
   - i. Skin biopsy
   - ii. Pus for culture and sensitivity
   - iii. Phage typing of the organism isolated
   - iv. Serology of Blood for syphilis of patient and parents

   STS from the patient and the parents were negative. Culture revealed staphylococcus aureus phage type 71. Biopsy revealed an intra epidermal bulla with acantholysis.

3. Which of the following diagnoses is now probable?
   - i. Pemphigus neonatorum
   - ii. Impetigo bullosa
   - iii. Toxic epidermal necrolysis (TEN)

4. What treatment should have been started in this child?
   - i. Systemic corticosteroids
   - ii. Systemic antibiotics
   - iii. A combination of the two
   - iv. No treatment

5. What could have been the prognosis of the child if he had presented earlier and appropriate treatment instituted?
   i. Still fatal
   ii. Serious but not fatal
   iii. Relapses and remissions

ANSWERS

1. Epidermolysis bullosa would seem to be the least likely possibility since the child was delivered normally and the bullous and pustular lesions were present all over. Secondary syphilis could not be entirely excluded though it seemed less likely because of pustules and large sheets of skin peeling off, eight days after birth. Pemphigus neonatorum is synonymous with bullous impetigo but no longer a commonly used term. In view of sheets of skin peeling off, TEN would seem the most probable diagnosis though bullous impetigo could well simulate SSS (Staphylococcal scalded skin) syndrome; perhaps bullous impetigo and SSS syndrome are the same disease.

2. This is an emergency and there is no time to lose in investigations; the treatment must be started straight away. Serology of patient's and parent's blood must be done and phage typing if staphylococci be isolated, would be helpful in diagnosis and management.

3. All the three conditions listed here are more or less the same disease. Isolation of phage type 71 substantiate further the diagnosis of TEN. Acantholysis often occurs in impetigo.

4. The patient should be energetically managed by systemic antibiotics. Unless severe constitutional symptoms and hypotension demand, there is no place for corticosteroid therapy.

5. Prognosis in all patients of TEN is very serious and a number of fatalities occur despite reasonable management. The disease is not necessarily fatal, though and the patient may have been helped if he had presented earlier.

Comment

While this patient presents a typical picture of SSS syndrome, it is not always easy to differentiate between drug induced and staph induced forms of TEN. Histologically SSS syndrome shows a split in upper Malpighian layers; drug induced type involves basal cell destruction. While in adults it is the drug induced that occurs commonly, staph-induced types have also been rarely reported. The history, histology and culture of the specific staph group 2 phage 71 helps in differentiation of the two. While the adult type needs large doses of systemic corticosteroids together with broad spectrum antibiotic cover, the SSS

syndrome mainly needs penicillinase resistant penicillins like Cloxacillin, as the corticosteroids do not at all affect the toxin.

The prognosis in SSS syndrome is reported to be excellent[2], though we see significant number of fatalities, despite energetic and apparently appropriate treatment. Lesions of bullous impetigo are produced by the same toxin as in SSSS and the latter may in fact be regarded as a massive form of bullous impetigo ; one criterion to differentiate between the two being able to culture the toxin producing organism in blood and not necessarily in all the skin lesions in SSSS[1].

References

1. Pearson RW : Advances in the diagnosis and treatment of blistering diseases, a selective review, year book of Dermatology, Edited by Malkinson & Pearson, Year Book Medical Publishers Inc, London, 1977, p 34.

2. Lyell A : A review of toxic epidermal necrolysis in Britain, Br J Dermatol 1967 ; 79 : 662.

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— *Managing Editor*