TOXIC EPIDERMAL NECROLYSIS

By

C. BHAKTA VIZIAM * & R. MATHAI ** & A. MAMMEN

Toxic Epidermal Necrolysis is a distinct clinical entity. It is characterised by a short lived febrile illness followed by the sudden appearance of widespread skin lesions, which are the result of necrosis and cleavage at various levels in the epidermis. The Nikolsky sign is markedly positive at this stage. In this paper, we are presenting 3 cases of toxic epidermal necrolysis seen in the department of Dermatology in the past one year.

CASE REPORTS

1. A 34 year old house-wife was perfectly well till 5 days prior to admission when she noticed swelling of her lower lip. This was attributed to an insect bite though there was no objective evidence of it. The same evening, patient developed fever and was treated with anti-histaminic. Three days later, she developed vesicles on the skin which increased rapidly in number and size.

   Past and family histories were non-contributory. On examination, patient appeared toxic There were large areas of skin with blackish discolouration. Many areas were denuded of necrotic epithelium and exposed raw surfaces. Rest of the skin revealed superficial flaccid bullae and vesicles, scattered all over. The largest bullae were on the palms and soles. Some of the bullae showed ecchymosis around them. Nikolsky's sign was strongly positive. Mucous membrane of the mouth and lips showed extensive superficial ulceration. She was running fever of 101°F. Hess test was negative. Other systems did not reveal any abnormality.

   Investigations : Nasal and throat swab cultures showed profuse growth of staph aureus, sensitive to tetracycline. All other investigations were normal. A clinical diagnosis of Toxic Epidermal Necrolysis was made and she was started on 120 mgs. of Prednisolone daily and Tetracycline. Two days after the initiation of therapy patient stopped developing new blisters. Subsequently, she improved remarkably in her general and skin condition. The dose of steroids, was therefore, reduced. Patient developed signs of acute abdomen two weeks after admission and expired within a few hours. Plain x-ray of the abdomen showed evidence of intestinal perforation. A post-mortem could not be obtained.

2. A 40 year old man developed a similar clinical picture while on Tegratol for trigeminal neuralgia. He was started on 150 mgms. of Prednisolone

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** Dept. of Dermatology, Christian Medical College Hospital, Vellore, (South India).

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daily and tetracycline. Patient made a complete recovery in two weeks time. The steroid was discontinued in another 10 days. He has remained well since.

3. A 27 year old female with nephrotic syndrome was on esidrex and some indigenous medicines before the onset of the skin disease. At the time of admission, she was acutely ill and in uremic coma. Her skin showed extensive areas of blackish discolouration on the face and trunk with denudation. In addition, she had several vesicles and bullae, many of them hemorrhagic. Multiple purpuric spots were also seen. Mucous membrane of mouth and eyes showed ulceration (figs. 1 to 3). Prednisone 100 mg. daily was started. But, her uremia bacame worse and expired on the tenth day. The skin lesions did not show any change towards recovery.

HISTOPATHOLOGY

There was moderate hyperkeratosis. The epidermis showed cleavage due to marked cellular necrosis. The necrotic cleavage contained free pigment and erythrocytes (fig. 4). Some areas of epidermis showed intra and inter-cellular oedema. This seemed to be the earliest histologic change in the epidermis leading to cellular necrosis (fig. 5). The basement membrane was intact in the early stage of epidermal necrosis and cleavage (fig. 6) but in the later stages it was destroyed (fig. 4). Changes similar to epidermal necrosis were also seen in the Sebaceous glands and hair follicles. In the upper dermis the blocd vessels showed mild endothelial swelling and perivascular infiltrate (fig. 7). Rest of the dermis looked normal.

PATHOGENESIS

The etiology and pathogenesis of toxic epidermal necrolysis is not clear. Speculations raised so far are, that this may be an expression of an idiosyncrasy, a modified and severe form of Steven-Johnson-Syndrome, a modified Sonarelli-Shwartsman phenon, an acute malignant form of pemphigus or an allergic reaction to food, drug or bacteria. Many drugs have been incriminated in the various cases reported.

In case No. 1 there was no history of drug ingestion. Her nose and throat swab cultures grew staph aureus. It is difficult to say whether this organism was or was not the cause of her illness. The second case was on tegratol and might have been the etiological factor in his disease. The third patient had esidrex and various other drugs for nephrotic syndrome. Drug as an etiological factor in this case can only be a speculation. Taking several findings into consideration toxic epidermal necrolysis appears to be an extreme variety of Steven-Johnson-syndrome due to one or more of the several etiological factors like infection, allergy etc. Individual unu.ual tissue response could be an important additional contributing factor in the pathogenesis.

MANAGEMENT

Case 1 and 2 showed definite clinical response to the steroids. Case 1 wast lost due to intestinal perforation, a rather common complication of steroid therepy.

Case 3 did not show any clinical response to steroids. The unresponsiveness to steroid therapy in this case appears to be due to the pre-existing critical illness of uremia. Although the role of the steroid in the management of this condition is still controversial [2-4], we have reason to believe that steroids are definitely useful, if instituted early in uncomplicated cases [5].

SUMMARY

In summary, Toxic Epidermal Necrolysis is a distinct clinical entity which shows skin with large areas of black discolouration and peeling; and mucosa with severe ulceration. Histologically epidermal necrosis and hemorrhage are its characteristic features. This entity is probably an extreme variety of Steven-Johnson-Syndrome. Steroid therapy is found to be useful in this condition.

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