Ind J Dermatol Venereol Leprol 1994; 60 : 294-5

DAPSONE RESPONSIVE JUVENILE PEMPHIGUS FOLIACEUS

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An 8-year-old girl of Pemphigus foliaceus is reported because of the rarity of the condition in childhood. The disease was resistant to steroids and showed a dramatic response to dapsone.

Key Words : Pemphigus foliaceus, Dapsone

Introduction

Pemphigus foliaceus is a bullous disorder that is most commonly seen in the fourth and fifth decades of life. It has been rarely documented in childhood.[1,2] We report Pemphigus foliaceus in a child who failed to respond to steroids but showed a dramatic response to dapsone.

Case Report

An 8-year-old girl presented with numerous, painful erosions and crusted lesions of 2 weeks duration involving face, neck, upper parts of the chest and the adjoining area on both arms. There was no mucosal involvement. There were numerous erosions covered with multilayered yellowish - red crusts. There were no papulovesicular lesions. Nikolsky sign was negative. The investigations including haemogram, urine examination, blood urea, blood sugar and liver functions tests were within normal limits. A pus swab was sent for culture, which revealed Staphylococcus aureus. Clinically, a diagnosis of impetigo contagiosa was made and accordingly patient was put on a combination of ampicillin and cloxacillin. There was partial response to the above treatment. The crusting was less after one week of therapy. However, some vesiculobullous lesions appeared on chest and arms, in the surrounding healthy skin. A skin biopsy taken at this juncture from a represented lesion revealed detachment of the horny and granular layer. The epidermal cells at the base of the clefts showed acantholytic cells (Fig 1). There was a perivascular mononuclear infiltrate in the dermis. Direct immunofluorescence from the perilesional skin revealed pericellular deposition of IgG and C_3 in superficial epidermis. The diagnosis of pemphigus foliaceus was entertained, in view of the progressive lesions and deteriorating condition. Patient was put on steroids (intramuscular dexamethasone 6 mg/day) alongwith an antibiotic. Steroid was increased to 8 mg/day after a week, because of the absence of any response. After being on steroid for another one week, during which new vesicles appeared and the older lesions did not show any repression, oral dapsone was started in a daily dose of 50 mg/day and increased to 100 mg/day after a week. There was a dramatic response to dapsone (100 mg/day) within a week and gradually steroid was decreased and finally stopped over 3 weeks when the patient was controlled completely with dapsone. After 3 weeks of therapy with dapsone, the dose of dapsone was decreased and the patient was discharged on a daily dose of 10 mg/day after 4 weeks of complete cutaneous clearances.

Comments

Pemphigus foliaceus is usually considered

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to be a less severe from of pemphigus, and is invariably treated with prednisolone in low-doses (25 mg/day).[3] Even topical steroids with or without occlusion may abort the disease. On the contrary, there are reports in which patients required high doses of steroids (140 mg/day[4] and 180 mg/day[5]). In one of the study[6] involving 9 adult cases of pemphigus foliaceus, the dramatic efficacy of dapsone was reported in five cases. In our case, dapsone showed an excellent therapeutic response after the initial failure of steroids in combating pemphigus foliaceus. It reiterates the view of Basset et al[6] that dapsone could be employed as first line drug in cases of juvenile pemphigus. In addition, dapsone is far more safer drug than steroids and thus complications of long term steroid therapy can be avoided. The mechanism of action of dapsone in pemphigus foliaceus is still not clear though it has been suggested to act through stabilization of lysosomal membranes or inhibition of polymorphonuclear leukocyte cytotoxicity.

References

1. Perry HO. Pemphigus foliaceus. Arch Dermatol 1961; 83: 52-70.
2. Perry HO, Brunsting LA. Pemphigus foliaceus: Further observation. Arch Dermatol 1965; 91: 10-21.
3. Sotiriou L, Herszenson S, Jordon RE. Childhood Pemphigus foliaceus: Report of a case. Arch Dermatol 1980; 116: 670-80.
4. Schroeter A, Sams WM Jr, Jordon RE. Immunofluorescent studies of Pemphigus foliaceus in a child. Arch Dermatol 1969; 100: 736-40.
5. Levine L, Bornstein JE, Soltani K, Medenica MM, Yung CW. Coexisting childhood pemphigus foliaceus and Grave's disease. Arch Dermatol 1982; 118: 602-4.
6. Basset N, Guillot B, Michel B, Meynadier J, Guilhou JJ. Arch Dermatol 1987; 123: 783-5.