Antibiotic Therapy in a Boy Affected by Generalized Epidermolytic Hyperkeratosis

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To the Editor

Generalized epidermolytic hyperkeratosis is a rare disorder of keratinization, featuring ichthyosis associated with erythroderma and bullous eruptions. Though most of the cases are sporadic, the inheritance of this condition is assumed to be autosomal dominant [1-3]. The effect of antibiotic therapy on a 9-year-old boy suffering from this disease is reported.

The boy is the third child of non-consanguineous and healthy parents. He was erythrodermic at birth, with extensive bullae and erosions, for which he was treated by a burns care team; he subsequently developed irregular hyperkeratosis. During the first months of life he had pneumonia and a staphylococcal meningitis, which resulted in tetraparesis. A congenital cardiomyopathy (open interven-tricular septum) was also detected. At 1 year of age a skin biopsy showed the characteristic histological and ultrastructural picture of epidermolytic hyperkeratosis. The boy came into observation at the age of 9 years. Physical skin examination revealed erythroderma, desquamation, irregular and yellowish hyperkeratosis with thick verrucous scales and transverse streaks in the area of major folds. All the body was affected except for his face, involved only in the perioral region. No blisters or erosions were detectable. The patient has been taking etretinate (10-15 mg/day) and amoxicillin for more than a year (2x500 mg/day). His family reported that every time the suspension of the antibiotic drug was attempted, soon after new blisters (serous at first and then purulent) occurred. Pyrosis, gastralgia, nausea and regurgitation appeared as side effects of this therapy.

We stopped the treatment with etretinate, because of its ineffectiveness against hyperkeratotic lesions. Amoxicillin was also suspended, but, as expected, bullae and erosions occurred about 36 h after withdrawal and progressively increased. A culture of purulent and serous bullae revealed the presence of *Proteus mirabilis*, *Staphylococcus aureus* and β -haemolytic streptococcus, group A. The boy was then treated with cefotaxime 2×1 g/day for 15 days. This treatment resulted in total regression of blisters and erosions. After 2 months we received a letter from his parents containing same snapshots: the boy was doing well and was still lesion free.

Epidermolytic hyperkeratosis is a tonofilament system disorder [1, 2]. Bullae are one of the characteristic features of this affection [3] and are often associated with fever and malaise. The importance of bacterial infections in the genesis of blisters is yet unclear. Blistering is modernly considered as the consequence of the alteration of the desmosome-keratin unit [2]. Infections should occur only subsequently [3], so that they should not have any pathogenetic role.

We believe that the role of bacterial infections in blister formation should be stressed. Our observation suggests that these patients should always be treated with an adequate antibiotic therapy when bullous lesions are present; patients should also regularly take antiseptic baths for prevention. Moreover, etretinate might negatively act, by augmenting skin fragility, to favour the rising of infections and bullae 121.

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References

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