Failure of Cyclosporine in Netherton's Syndrome

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Key Words

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Cyclosporine

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In 1958, Netherton [1] reported a unique case of trichorrhexis nodosa 'bamboo hairs'.

Netherton's syndrome (NS) consists of the triad of ichthyosiform dermatosis, multiple defects of the hair shaft (trichorrhexis invaginata) and an atopic diathesis [2-4].

By reviewing the literature, we found that the number of articles of NS in adults compared to those in infants and children is very low. Since the first report of NS, various treatments have been proposed. NS is a genodermatosis so that all treatments mentioned should be considered as symptomatic.

Retinoids have been reported to be efficient in the treatment of NS [5-11]. Happel et al. [5], Traupe and Happel [9] and Happel and Kold [10] reported excellent results using long-term etretinate treatment (0.5-0.8 mg/kg/day). Hartschuh et al. [7] reported a case of NS responding to long-term treatment with low doses of acitretin (5 mg/day). Hausser et al. [8] reported a case of NS responding to acitretin treatment (35 mg/day alternating with 10 mg/day). Systemic retinoid treatment is a well-known treatment for ichthyosis. The use of retinoids in NS seems to have more effect on ichthyosis than on the atopic diathesis, which is the limiting factor for this treatment, since it can be worsened by a systemic retinoid treatment [5, 7, 8].

The association of systemic retinoid therapy with PUVA treatment has been recommended by Hintner et al. [12], and Nagata [13] reported improvement of NS only by photochemotherapy. An alternative to systemic therapy are topical treatments. Buxman et al. [14] and Wehretal. [15] reported 2 cases of effective treatment of NS with lac-tate 12% lotion. Skin lesions improved within 2 weeks of topical treatment. The advantage is that this treatment can be considered as a long-term treatment.

We follow a young female patient, expressing the symptom triad of NS [16-18]. Our patient was treated with systemic retinoids: etretinate 25 mg/day during 7 years, isotretinoin 30 mg/day during 3 months, acitretin 20 mg/day during 2 weeks. These treatments had to be interrupted because of the occurrence of a severe facial eruption and for aggravation of ichthyosis. She did not respond to multiple alternative treatments including lactate 12% lotion, PUVA therapy, triamcinolone injections (Kenacort®) or systemic administration of ibuprofen (Brufen®), tetracosactide (Synacthen®), metronidazole (Flagyl®) and ketoconazole (Nizoral®). Since she had a total IgE serum level of 15,526 IU/ml and since Wolach et al. [19] have recently reported the successful treatment of hyperimmunoglobulin E syndrome with cyclosporine, we introduced a treatment of cyclosporine (Neoral®) at 3 mg/kg/day (1 month). We repetitively controlled

cyclosporine plasma levels. They were between 0.1 and 0.3 mg/l. After 2 months we did not see an effect on the skin lesions, so we decided to increase the dose to 4 mg/kg/day. We continued this treatment for 2 months. After a total of 3 months of therapy, we still observed no clinical improvement and decided to interrupt cyclosporine treatment. In our case, cyclosporine therapy had no effect on the skin lesions.

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