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Keratodermia punctata hereditaria Treated with Etretinate (Tigason)

Jørgen V. Christiansen

Department of Dermatology, Marselisborg Hospital, University of Aarhus, Aarhus, Denmark

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Abstract. Nine patients with keratodermia punctata hereditaria were treated with etretinate 0.5 mg/l mg/kg/day for 3 to 13 months. The result was good to moderate in 7 of the 9 patients.

Key words: Keratodermia punctata hereditaria; Etretinate (Tigason)

Bergfeld et al. (1) treated 6 patients with keratosis palmaris et plantaris with isotretinoin, with good results in 5 of 9 patients after 8 to 12 weeks observation period. We have treated 9 patients with keratodermia punctata hereditaria with etretinate (Tigason).

MATERIAL AND METHODS

Nine patients, 4 men and 5 women, were treated for 3 to 39 months with etretinate 0.5 mg/l mg/kg/day. The mean age of the group was 57 years. Measurements of safety and efficacy were made at monthly intervals.

RESULTS

The results were good in 3 patients, moderate in 4, and there was no effect in 2 patients. The treatment was stopped in 6 of 9 patients after 3 to 13 months. One man and 2 women had continued treatment up to 39 months.

Side effects

All the patients had some dryness of the mucous membranes, one man and 2 women had some defluvium and one man and one woman had some pruritus. There was no increase in liver transaminases during the treatment.

DISCUSSION

The treatment of all types of hereditary keratodermia has been unrewarding in the past. We now seem to have some prospect of helping these patients. There seems to be almost the same positive outcome with etretinate as found with isotretinoin. In most cases the treatment had to be continued for a rather long time, but patients seem to tolerate the treatment rather well.

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Ear Ache during Etretinate Treatment

Lennart Juhlin

Department of Dermatology, University Hospital, Uppsala, Sweden

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Abstract. Two patients with ear ache related to etretinate treatment in a dose of 50–75 mg are reported. The ache disappeared within a week after lowering the dose and reappeared when the dose was increased.

Key words: Etretinate; Ear ache; Side effects

Etretinate is known to cause dryness of the lips and mucous membranes. Kramer recently reported excessive cerumen production and otitis externa after etretinate (Tigason) in a patient with Darier's disease (1). This prompted me to report on 2 patients without external otitis but with ear ache related to etretinate therapy.

CASE REPORTS

Patient 1. A 40-year-old woman with pustulosis plantaris and obesitas (81 kg). After treatment with 75 mg Tigason for 2 weeks her lips and mouth were dry and her throat and the right ear were aching. She decreased the dose to 50 mg Tigason and the symptoms disappeared within a week. The patient thought she had merely had a sore throat. She thereafter also received local psoralen-bath plus UV-A treatment. After one month she increased the dose again to 75 mg. Her lips and mouth became dry again after about a week, and her throat and right ear started to ache again. She then reduced the dose to 50 mg. The ache became less severe but did not disappear and her mouth was still dry after a week. She then stopped the treatment and her ear and mouth problems had disappeared completely after 3 weeks.

Patient 2. A 57-year-old man with palmoplantar pustulosis. After 3 weeks' treatment with 75 mg Tigason his lips became dry and tender and he felt pain in his left ear. It was "blocked" and he had difficulty in hearing on the left side. There was no sign of otitis. He reduced the dose to 25 mg. The ear ache disappeared within a few days. When he increased the dose to 50 mg it returned and he therefore reduced the dose to 25 mg which could be kept without problems. He had never before had any trouble with his ears.

COMMENTS

Ear ache has not previously been mentioned during etretinate therapy. The ache in our patients fits best with an obstruction of the auditory (Eustachian) tube. The tube has a pseudostratified epithelium which also could respond to etretinate with an inflammatory reaction. Since the ache occurred on one side only we also have to assume that the tube or its opening into the pharynx is narrower on one side and that by the act of swallowing the lumen is not opened to equalize the pressure in the middle ear. The side effect seems to be rare but it may be overlooked if not asked after, since patients might assume they have a cold.

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