Successful Treatment with Alitretinoin of Dissecting Cellulitis of the Scalp in Keratitis-Ichthyosis-Deafness Syndrome

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Keratitis-ichthyosis-deafness (KID) syndrome is a congenital disorder, which is associated with mutations in the GJB2 gene encoding connexin 26 or in the closely related connexin 30 gene, GJB6. To date, approximately 100 cases have been described, mainly sporadic, but autosomal dominant inheritance has been reported in a small number of families. The syndrome is characterized by profound sensorineural hearing loss accompanied by vascularizing keratitis and erythrokeratoderma-like skin lesions. Palmoplantar hyperkeratosis, alopecia, dystrophic nails, and increased susceptibility to bacterial, viral and fungal infections and squamous cell carcinoma are other common features of this syndrome (1, 2). Treatment of patients with KID syndrome is difficult and often disappointing. Systemic retinoids, acitretin or isotretinoin have been used in patients with KID syndrome with variable success (3–6). We report here a case of KID syndrome where dissecting cellulitis of the scalp was treated successfully with alitretinoin.

CASE REPORT

A 15-year-old boy, born of non-consanguineous healthy parents, presented to our department in 2002 with a history of red and dry skin since birth, sensorineural deafness, red-brown hyperkeratotic plaques on his face and extremities, and sparse hair. Paronychia with nail dystrophy affected most of his fingers and all his toes. In 2003 he was the first Danish patient in whom KID syndrome was verified by mutation analysis, showing a heterozygous missense mutation D50N (148G >A) in GJB2 (6).

The patient had a history of several unsuccessful treatments. Topical treatments with calcipotriol, corticosteroids and tar had been used with no effect. Pulsed dye laser had also been tried with no effect. Etretinate (Tigason, Betapharma, Shanghai) 10 mg per week was introduced when he was only one year of age. Blood tests, including thyroxin (T4) and thyroid-stimulating hormone (TSH), were all within the normal limits. 

He continued this treatment for 4 months with no effect. From the age of 5 to 7 years he was treated with acitretin (Neotigason, Actavis, UK) 10 mg daily, but had to stop the treatment because of erosive skin lesions in his auditory canals. In the following years, his skin lesions and nail surroundings had often been infected with dermatophytes, Candida and pyogenic bacteria, thus he needed frequent treatments with topical and systemic antibiotics and antymycotic drugs. At the age of 17 years, treatment with acitretin was resumed as he showed a follicular occlusion triad, with dissecting cellulitis of the scalp, cystic acne and hidradenitis in his axillary regions and groins. Acitretin 20 mg daily for 2 months did not alter his condition and a higher dose was not possible because of side-effects. As the treatment did not improve his condition, isotretinoin (Roaccutan, Roche, Switzerland) 20–30 mg daily in combination with prednisolone 25 mg daily were initiated and combined with dicloxacillin. In this period he was regularly examined by an ophthalmologist, to ensure that systemic retinoids did not provoke a keratitis or other ophthalmic side-effects. He had continuous painful dissecting cellulitis of his scalp (Fig. 1A) and was regularly receiving wound care in the outpatient clinic. A plastic surgeon was consulted, who considered transplantation of skin on his whole scalp. Photodynamic therapy was also tried with no effect. However, some improvement was seen after systemic treatment with clindamycin (Dalacin) and rifampicin (Rimactan) given for 3 months, followed by the antymycotic drug itraconazole (Sporanox). In 2009 the patient developed an ulcer on his left leg. The sudden onset of the ulcer and its slow healing resulted in several biopsies being taken from the ulcer in order to exclude malignant or premalignant changes. The hyperkeratotic plaques on his leg hampered healing of the ulcer, thus he resumed treatment with acitretin. Within 2 months acitretin was discontinued, because of aggravation of his skin condition. It was clear that the patient’s condition would not improve with acitretin, thus alitretinoin (Toctino, Basilea, Switzerland) 10–20 mg/day for 5 months. Followed by a maintenance dose of 20 mg daily. Blood tests, including thyroxin (T4) and thyroid-stimulating hormone (TSH), were all within the normal limits.

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Fig. 1. Clinical images showing dissecting cellulitis of the scalp (A) before and (B) after treatment with alitretinoin 10–20 mg/day for 5 months.
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In most European countries alitretinoin is hitherto only legislated for chronic hand eczema (8). The effect of alitretinoin in KID syndrome has been published previously in only one case report (5). The patient in this case received alitretinoin 30 mg per day as a maintenance dose for 5 months, and during the course of treatment there were no detectable adverse effects. After 5 months of therapy the infiltrated erythema on the face had decreased, and the hyperkeratotic plaques had almost completely disappeared (5). In our patient the hyperkeratotic skin lesions were treated with topical moisturizers, keratolytic agents, potassium permanganate baths, and alitretinoin. Alitretinoin reduced the severity of the hyperkeratotic plaques during the first months of treatment, and had a positive effect on the dissecting cellulitis, and thus improved our patient’s quality of life. Our patient showed a significant improvement in his scalp and skin symptoms, which indicated that long-term continuous therapy with alitretinoin may be a possibility in patients with KID with severe dissecting cellulitis.

The prognosis of KID syndrome depends on early diagnosis of infections and skin cancers. These patients need lifelong follow-up for early diagnosis of malignant tumours, especially squamous cell carcinomas of the hyperkeratotic skin and mucosa (4, 6). Systemic retinoids reduce the hyperkeratosis and may reduce the incidence of skin cancer. We conclude that treatment with systemic alitretinoin may be an option for KID syndrome in cases complicated with follicular occlusion and dissecting cellulitis. Continuous monitoring for adverse effects remains essential, and more research into alitretinoin therapy is needed in order to determine the most effective dosage and the long-term safety of this medication in disorders of keratinization (9).

REFERENCES


