Dermoabrasion - method of choice in treatment of morbus Darier

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SUMMARY

Presented are patients with typical changes in morbus Darier, successfully treated with the method of deep abrasion. After experimental abrasions the regenerated epidermis revealed all the characteristics of morbus Darier, while the regenerated epidermis after deep abrasion, when the regeneration resulted from remnants of epidermal adnexes, became completely normal both clinically and pathohistologically. Abrasion of large surfaces is performed under complete anesthesia. Morbus Darier is the only hereditary keratoderma in which after deep dermoabrasion the clinically and pathohistologically normal epidermis is regenerated and remains permanently unchanged. A highly probable explanation is that during the embriogenesis the epithelial adnexes had been formed before the pathological process characteristic for morbus Darier started.

KEY WORDS

Darier's disease, therapy, dermabrasion, deep

Introduction

Morbus Darier or dyskeratosis follicularis is a special form of the disturbance of the follicular keratinization, which appears clinically on the skin, mucous membranes and nails. Approximately, it affects one to two persons in 100.000, and is predominantly inherited by anauto somal dominant gene (1). Its genetic expressivity is variable, and often spontaneous mutations occur. The deficiency of the tonofilament-desmosome complex is responsible for this anomaly. The credit for the description of the disease belongs to Darier and White who had provided it in 1889 (2,3). But since Darier had been the first to describe the pathohistological changes, his name is used as eponym for the disease. The beginning of this disease is gradual, and it occurs mostly between the ages of 10 and 15, appearing as keratotic follicular papules of dirty brown color up to the length of 5 mm. With the removal of the pink plug or small warts, which also appear at times, there emerge warty vegetations emitting an unpleasant smell. Patients also complain of itching. The rudimentary forms are also described. The buccal mucous membrane, tongue, pharynx, larynx and rectum may also be involved. An abnormal function of T-cells as well as mental retarda-

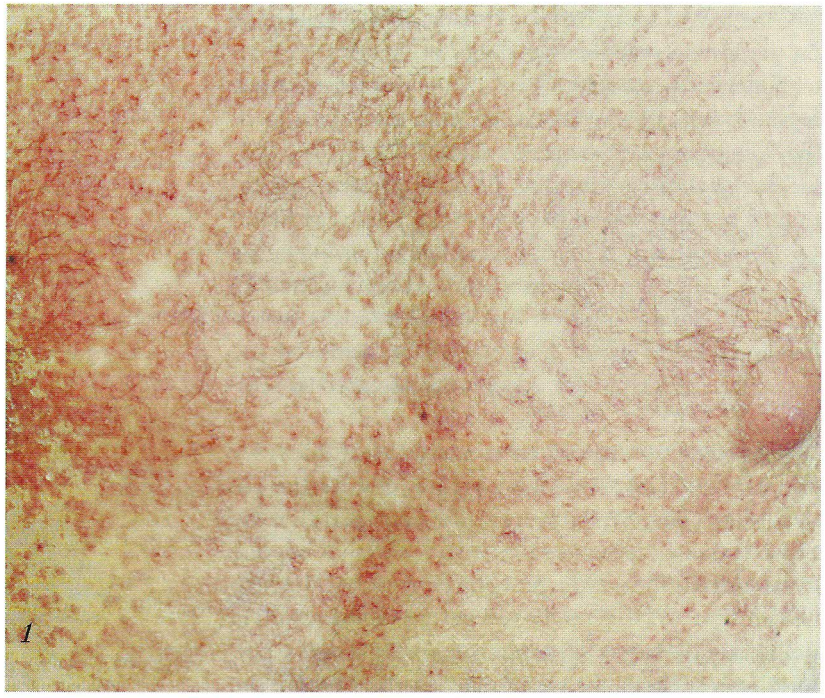

Figure 1. Male, 25 years, with characteristic manifestations of morbus Darier on the chest.

Figure 2. The same patient on whom a test abrasion had been performed six months ago with completely regenerated normal skin (the smaller clear part) and with the second recent test abrasion (bloody area)

tion and cystic changes in the bones are also mentioned. The nails display white or reddish streaks, thickenings or subungual keratosis. With time the symptoms may become more prominent. External influences, warmth and UV exposure may have a negative effect on the disease. Diagnosis is made clinically and pathohistologically. In the epidermis, round the follicle, dyskeratotic cells are seen, usually in the stratum spinosum (*corps ronds*) or in the stratum granulosum (*grains*). With the electron microscopy the damaged tonofilament-desmosome complex is visible. By differential diagnosis should be excluded the seborrheic dermatitis, acanthosis nigricans, pemphigus chronicus familiaris benignus (Hailey-Hailey), hyperkeratosis follicularis et perifollicularis in cutem penetrans (morbus Kyrle). Verrucous lesions on the dorsa of hands and legs are described by some writers as acrokeratosis verruciformis Hopf. Prognosis of the disease is not favorable. Keratolytics, the retinoic acid (tretinoin) or corticosteroid preparations are used locally. Oral retinoids have a favorable effect, unfortunately lasting only during the treatment (4).

Patients and treatment

Male, 25 years, with negative family anamnesis. In his case the changes began to appear at the age of 15 when he noticed small brownish papules, first on the chest, that in the following years spread all over the trunk, they were fewer on the back than on the chest. He was unsuccessfully treated with various ointments until he received Neotigason, which was effective, but only while he was taking it. Upon his admission to the clinic the trunk was covered with thickly disseminated brownish papules. They were ranged in a follicular pattern, the size of millet grains (Figure 1). Individual papules could also be observed on the proximal parts of upper arms. The lesions caused intense itching. His psychical state was normal. Two test dermoabrasions were preformed: one small six months ago revealing normally regenerated healthy skin (the clear, round part), and a freshly abraded larger surface. (Figure 2).

Female, 34 years, married. Family anamnesis negative. The disease started at the age of 12 on the chest and the lateral parts of neck where minute papules appeared and were slowly spreading. The changes worsen in summer, especially during sun bathing. She was treated with various ointments and retinoids that were helping her, but only while she was taking them. Upon her admittance erythematous skin on her breasts, below and between them was visible, with numerous small papules of a red-

dish color located follicularly. (Figure 3). Individual papules have also been observed on the lateral parts of her neck and below her navel. The psychic condition was regular. In Figure 3 we can also observe the result of a test dermoabrasion performed 6 months earlier.

Male, 34 years. His history contained the information that one of his uncles suffered from this disease. In our patient the disease appeared in puberty on the chest, and at the age of 20 it spread over the entire trunk reaching the inguinal region. (Figure 4). Moreover, smaller individual lesions developed on the dorsal parts of the lower extremities. He was treated with various ointments and Tigason orally that was of help until he was taking it. The changes caused itching. The trunk was diffusely covered with brownish minute papules, more to the fore and placed follicularly. Two smaller surfaces covered with similar changes are displayed on the extensory parts on both shins. The nails on both hands were thickened with longitudinal streaks. A few papules could also be observed on the oral mucosa. According to the anamnesis the patient's condition worsened considerably after a sunning spell. His psychic condition was regular.

All the patients were completely examined to exclude possible contraindications concerning dermoabrasion. After biopsy and pathohistological examination, a test abrasion was first performed under local anesthesia on the site of the most pronounced skin changes (Figure 1). The patients, seeing the results of abrasions after 5 to 6 months, agreed to the procedure. Naturally, large surfaces could not be abraded under local anesthesia. The operation was performed under general anesthesia at the surgical clinic. When the bleeding stopped, the abraded part was strewn with antibiotic powder and left to dry in the air, regardless of the size of the planed surface. From under the crust, which developed already the next day, epithelization developed quickly. The crusts began to fall off after ten days. After 6 months, in Figure 5, a border can be seen between the treated area and the limiting part that have not been abraded. It must be stressed that the patients with a large abraded surface suffered great pains during the postoperative period. The crust was cracking at the slightest stir, sneezing, coughing and strain causing much pain to them. During the first week they were continually receiving analgesics. After five to six months, when all the patients were checked, the abraded parts were clinically and pathohistologically completely normal.

Discussion

Curtin's technique of dermoabrasion, which in the world at large has attracted numerous followers, has also been used for decades in Croatia. (5-11). It is based on the lasting regenerative ability of the skin. Full attention must be paid to prevent too deep abrasions since regeneration is possible starting from the remnants of the skin adnexes. The surface planing above the papillae does not cause bleeding. Bleeding starts when corium is reached where the superficial plexus of blood vessels is placed. If during abrasion fat droplets are noticed, it means that the subcutis has been reached. If, however, during abrasion epidermal adnexes (sebaceous and sweat glands, hair follicles) are removed, the defect does not regenerate, but is just repaired by scarring.

Treatment of keratoderma is a great problem for any dermatologist, but even a greater one for the patient, depending on the localization and spread of the disease. Great hopes were pinned on retinoids, but due to side effects the treatment will sooner or later have to be cut short. (4). Over the years we applied abrasion in numerous disorders of keratinization by always performing test abrasions on a limited surface where the pathological changes were most pronounced (ichtyosis, mal de Meleda, karatodermia figurata variabilis, psoriasis vulgaris, etc.). Unfortunately, the results were disappointing, on the abraded surface the original disease with its typical changes kept reappearing. Satisfactory results cannot be achieved in any other keratoderma.

That is why we were pleasantly surprised when by deep dermoabrasion a clinically and pathohistologically normal skin was obtained in Darier's disease. A highly probable explanation is that during the embriogenesis the epithelial adnexes had been formed before the pathological process characteristic for morbus Darier started. Those were patients in completely normal psychic conditions who were firmly decided to recover and regain their normal appearances. After the treatment their life according to their own words acquired a higher quality. Now, even after 12 years, on the abraded parts no pathological changes are visible. Therefore we suggest that dermabrasion should be more widely used for treatment of morbus Darier.

Conclusion

Thme patient must be an adult, in a period after twenty years of age that tends to regain his or her normal appearance and has no psychic problems. The team performing dermabrasion must be protected (protective devices, masks, surgical gowns), since with the spouting of blood droplets and particles of abraded tissue, an infection with AIDS or hepatitis may take place (12).

Figure 3. Female, 34 years, with a performed test abrasion and normal regenerated ski after six months.

Figure 4. Male, 34 years, with characteristic changes of morbus Darier from the neck to inguinal regions

Figure 5. The same patient after six months. The limit of the abraded frontal part with completely regenerated normal skin, and the posterior still non-abraded part on which the changes are less expressed.

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