EPIDERMODYSPLASIA VERRUCIFORMIS
(A case report)

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Summary

A case of epidermodysplasia verruciformis is presented. Empirical therapeutic trials with Sulphaphenazole, Dapsons and Thiosemicarbazone failed to improve the lesions; however, temporary response was obtained with 5% Salicylic acid ointment. Literature on its etiology has been briefly reviewed.

Epidermodysplasia verruciformis is a rare entity and was first described by Lewandowsky and Lutz in 1922. The nomenclature literally means faulty (dys) development (plasia) of epidermis in the warty (verruci) form (formis). The lesions in this dermatosis vary from papules resembling flat warts to more markedly verruciform papular elements depending largely on the area of localisation. Usually the skin lesions are limited to backs of the hands, face, neck, extremities and feet, but may be generalised. Mucous membranes are usually spared. The condition usually begins in early childhood, often at birth, but in some it may appear later (Allen, 1967).

A typical case of this condition came under observation and we tried some drugs to assess their therapeutic value in this disorder. We find there is paucity of reports of this disorder in the literature and, therefore, consider this case to be of sufficient interest.

Case Report

A thirteen year old boy presented with asymptomatic multiple discrete papular lesions on the face, trunk and extremities for the last eight years (Fig. 1).- Lesions started on the extremities and gradually appeared on other parts of the body. There was no history of such disease in any of the family members and history of consanguinity in parents was also absent.

Fig. 1

Back of the patient showing wide spread papular-warty lesions

On examination the lesions were skin coloured, discrete, round, flat topped and papulo verrucous varying in size

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from 2 to 5 mm, distributed over forehead, cheeks, trunk (both anterior and posterior aspects), arms, dorsum of the hands, thighs and dorsum of the feet. There was no mucous membrane involvement. Systemic examination did not reveal any abnormality. Haemogram, urine, stool and radiological examination of chest were found to be normal. Skin biopsy showed changes typical of verruca plana (Fig. 2).

Fig. 2

Photomicrograph of a lesion from the back of the patient

Therapeutic trial with sulphaphenazole (Orisul), 250 mg twice daily for four weeks did not show any improvement. Dapsone 50 mg twice daily for six weeks proved failure and later on Thiosemicarbazone 100 mg twice a day for six weeks also did not prove of any help to the patient. Since these empirical measures failed, the patient was put on conventional local treatment with Keralolytic i. e. 5% salicylic acid ointment. In three weeks time patient showed remarkable improvement but the lesions reappeared on stopping the local treatment.

Discussion

In the beginning the condition was thought to be genodermatosis transmitted by autosomal recessive (Lewandowsky and Lutz 1922). Familial incidence often reported (Midana, 1949; Jablonska et al. 1966) was not present in our case.

Later on the possibility that it represented an unusual response to wart virus was supported by successful autoinoculation and heteroinoculation studies (Lutz, 1946; Jablonska and Milewski, 1957; Jablonska and Formosa, 1959). In 1957 Lutz himself agreed that it is an unusual body response to wart virus. However, doubts were cleared by the demonstration of intranuclear virus particles by electron-microscopy (Jablonska et al. 1966) and the present thought is that epidermodysplasia verruciforms is a dermatosis in which the lesions of the verruca plana occur on the skin with certain anatomical or functional abnormalities, which may be inherited in susceptible individuals.

Treatment of this condition is still exceedingly unsatisfactory. In this patient empirical use of Sulphaphenazole, Dapsone and Thiosemicarbazone were not effective and local treatment with keratolytic has given a temporary benefit.

Malignant changes often described in 20% of cases (Mashkilleison, 1931, Jablonska et al. 1966) usually occur at younger age but no such change was seen in this patient during the period of 2 years follow-up.

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True

So far all the reports of Wiskott – Aldrich syndrome has been in boys. The thrombocytopoenia and bleeding are almost invariably found. However a very similar clinical picture was reported in a girl who had no thrombocytopoenia. This may be a possible female variant of the Wiskott – Aldrich syndrome.

Reference: Arch. Dis. Childhood 45 : 527, '70.