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FAMILIAL MULTIPLE KERATOACANTHOMAS

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A man aged 54 years developed multiple keratoacanthomas with typical histological features over the scalp, face, trunk, upper and lower limbs. History of similar lesions being present in the patient's father and also his son led to the diagnosis of familial multiple keratoacanthomas. Response to oral methotrexate was encouraging.

Key Word : Keratoacanthoma

Introduction

Familial multiple keratoacanthomas are often referred to as Ferguson-Smith type of multiple self-healing squamous epitheliomas of the skin. The inheritance is of the autosomal dominant type, with majority of lesions occurring on the exposed surfaces, appearing in early adult life. Histologically pseudoepitheliomatous hyperplasia of epidermis with keratin masses and absence of significant atypia differenciates it from squamous carcinoma. The number of lesions may vary and in this case their large number and familial incidence were interesting features.

Case Report

A 54-year-old man came with multiple pruritic nodules on the skin over the scalp, trunk, upper and lower limbs of 1 year duration. History of lesions developing at the site of injury like minor trauma, scratching and also after taking injections was given. Starting as a small papule the lesions attained various sizes leaving behind pigmented depressed scars after spontaneous involution.More than 100 lesions were present on both lower limbs (Fig. 1) followed by upper limbs and trunk. Few papulonodules were seen on the scalp and no lesions were observed on the lips or subungual regions.Papules were firm,

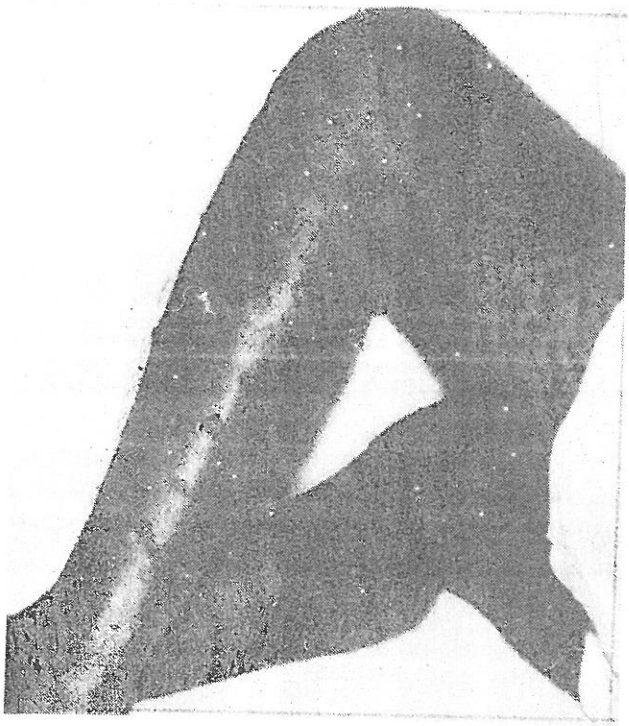

Fig. 1. Numerous keratoacanthomas on the lower limbs

pigmented with the centre containing a horny plug and over-hanging edges. On removing the horny plug the base had irregular and puckered surface with a thick epidermal

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edge. Areas of depressed pigmented scars were spread between papules. Systemic examination showed a monoplegia of the left upper limb of 15 years duration.

The patient was a known diabetic for past 3 years. Biopsy revealed pseudoepitheliomatous hyperplasia of epidermis with keratin masses without significant atypical cells, central keratin-filled crater and epidermal proliferation, extending into the dermis with chronic inflammatory cell infiltration in the dermis.

The patient was put on weekly oral methotrexate calculated on weight basis. There was history of similar lesions being present in the patient's son, and father, who had expired 3 years back due to unknown reasons.

Comments

Cases of multiple keratoaconthoma reported earlier[1,2] do not highlight the familial tendency as seen in this case.The presence of large number of lesions was not observed in any other reports. Cutaneous squamous cell carcinoma was excluded by the familial incidence, large number of lesions, and absence of induration and regional lymphadenitis.

Topical 5 fluorouracil was not tried as the area of involvement was extensive, The initial response to oral methotrexate was encouraging.

References

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