ACROKERATO-ELASTOIDOSIS LINEARIS MANUS

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A variant of acrokerato-elastoidosis characterized by linear, scaly, hyperkeratotic, fissured and transluscent papules on sides of thumbs and index fingers, was seen in 55 male patients, above the age of fifty years. Actinic damage and repeated trauma were not evident. The term acrokerato-elastoidosis linearis manus is suggested for this variant.

Key words : Acrokerato-elastoidosis, Linear.

Degenerative collagenous plaques of hands and feet or acrokerato-elastoidosis (AKE) is an uncommon distinctive form of elastosis seen in outdoor workers in sunny climates.[1] It is believed to be an autosomal dominantly inherited condition characterized by firm, shiny papules at the periphery of the palms and soles with some extension to the dorsa of feet.[2-4] The present communication describes a series of patients seen by us in which the lesions of AKE were confined to the sides of index fingers and thumbs only.

Case Report

A 52-year-old male taxi driver developed gradually increasing lesions on both hands for the past 3 years. Apart from a cosmetic disfigurement, the lesions were asymptomatic. There was no excessive sweating or family history of similar lesions.

Examination revealed scaly, fissured, hyperkeratotic and translucent papules at the junction between the palmar and the dorsal skin especially along the contiguous borders of the thumbs to the tip of the index finger. The papules were confluent at places. There was thickening of the skin of palms and soles. The rest of the cutaneous examination did not reveal any abnormality. Routine laboratory investigations were normal.

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Skin biopsy from one of the papules on the finger revealed hyperkeratosis and mild irregular acanthosis. The dermal collagen was unremarkable with haemotoxylin and eosin staining. However, van Gieson's stain for elastic tissue showed thickening and fragmentation of elastic fibres in the mid-dermis.

During the past 3 years, we have come across 55 patients, including the above case, with identical skin lesions at similar sites. All these patients were males and above the age of 50 years (range 50.5 to 70 years). None of them had a positive family history of similar lesions or hyperhidrosis. They have been engaged in different professions namely doctors, engineers, clerks, executives and manual workers. Only 3 had lesions suggestive of actinic damage on the face. Twenty three had diabetes mellitus and/or hypertension.

Comments

Review of literature suggests that cases nearly similar to our patients have been described by only a few workers earlier.[5,6] Burks et al,[5] reported 5 patients under the title degenerative collagenous plaques of hands. The patients had linear, firm plaques on the medial and lateral aspects of the hands. Four of the patients were over 60 years in age and also had actinic skin changes. Skin biopsy revealed reticular dermal elastic degeneration as well as evidence of solar damage in the papillary layer. Ritchie and Williams,[6] reported 6 patients with similar lesions and on the basis of histopathologic features concluded that it does not represent a separate clinical entity since

histopathology was similar to that of senile degeneration. Again, all patients had actinic changes on the sun-exposed areas. Kocsard,[7] reported 15 patients under the heading keratoelastoidosis marginalis of the hands. All these were manual workers and he suggested that occupational trauma might be a factor in addition to actinic damage. Recently, Highet et al,[8] reported that there appears to be two clinical syndromes of AKE;(1) an idiopathic, sometimes familial, form without associated actinic change with onset usually in youth, and (2) lesions developing later in patients with existing actinic damage, and in whom repeated trauma may also be important.

In our cases however, lesions start late in life, are not usually associated with actinic damage and repeated trauma is not a factor in its causation. The histopathological features are diagnostic of AKE.

Therefore we suggest that in addition to the 2 clinical syndromes of AKE as mentioned by Highet et al,[8] a third variant of AKE also needs to the recognised, and called acrokerato-elastoidosis linearis manus.

References

1. Cunliffe WJ : Disorders of connective tissue; in : Text Book of Dermatology; 3rd ed, Vol II, Editors, Rook A, Wilkinson DS and Ebling FJG : Blackwell Scientific Publications, Oxford, 1979; p 1648.
2. Lever WF and Schaumberg Lever G : Congenital diseases (genodermatoses), in : Histopathology of the Skin, 6th ed, JB Lippincott, New York, 1983; p 61.
3. Costa OG : Acrokeratoelastoidosis, Arch Dermatol, 1954; 70 : 228-231.
4. Jung EG, Beil FV and Anton-Lamprecht I et al : Acrokeratoelastoidosis, Hautarzt, 1974; 25 : 127-133.
5. Burks JW, Wise LJ and Clark WH Jr : Degenerative collagenous plaques of the hands, Arch Dermatol, 1960; 82 : 362-364.
6. Ritchie EB and Williams HM Jr : Degenerative collagenous plaques of the hands, Arch Dermatol, 1966; 93 : 202-203.
7. Kocsard E : Keratoelastoidosis marginalis of the hands, Dermatologica, 1964; 131 : 169-175.
8. Highet AS, Rook A and Anderson JR : Acrokeratoelastoidosis, Brit J Dermatol, 1982; 106 : 337-344.