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MUTILATING ACRAL KERATODERMA

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A 15-year-old male, born of non-consanguineous marriage started developing thickening of skin of sole of the both feet at the age of 7 which progressed to involve whole of both feet by 11th year. Patient later developed painful autoamputation of little toes of both feet due to pseudoainhum. Involvement of the palms was limited to a localised area over palmar aspect of right middle finger and was non-progressive. No other family member was affected. This may represent a variant of Vohwinkel's syndrome or the rarer form of acral keratoderma.

Key Words : Keratoderma, Mutilating, Palmoplantar

Introduction

The term palmoplantar keratoderma is used to describe the thickening of skin over palms and soles. The condition may be inherited or acquired. They are classificed by morphology and distribution of thickening, genetic transmission, presence of skin lesions on areas other than palms and soles and age of onset.[1]

Case Report

A 15-year-old male patient born of non-consanguineous marriage, apparently normal at birth with normal milestones presented with complaints of thickening of skin over palms and soles and loss of little toe of both feet. Patient first noted thickening of skin of sole of right foot at the age of 7 years over the heel. It gradually spread to involve the whole of the right sole in 1 year. The process later affected sole of the left foot. At 11 years of age, he developed constriction at the base of the little toe over both feet. Constriction progressed to produce painful autoamputation of right little toe in 2 years time and left little toe in 3 years time. At 13 years of age patient developed thickening of skin over palmar aspect of finger of the right hand. Since then patient has not develped any new lesions. There was no family history of similar illness.

On examination, patient was thin with diffuse thickening of skin over soles of both feet. Little toe was missing on both feet. There was localised thickening of skin over palmar aspect of right middle finger. Left hand was completely normal. Hyperhidrosis of palms and soles was present. The thickening lacked an inflammatory halo. There was no thickening of skin over knees, elbows, wrists or dorsa of hands. Patient did not show any features of myopathy or spastic paraplegia. Nails, hair and mucous membranes were normal.

Haemogram was within normal limits. Barium swallow did not show any abnormality. X-ray feet showed absence of little toe on both sides. Other bony structures were normal. X-ray hands did not reveal any abnormality. Audiometry was normal. Biopsy taken from plantar skin showed massive hyperkeratosis, hypergranulosis, acanthosis and sparse inflammatory infiltrate around the blood vessels and appendages consistent with the histology of palmoplantar keratoderma.[2] There were no features suggestive of epidermolytic hyperkeratosis or any other form of dyskeratosis.

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The foot lesions were excised by plastic surgeons and patient was treated with topical keratolytics and retinoic acid 0.05%. But the patient again developed diffuse thickening of skin over soles of both feet. But the palmar lesion has remained the same for past 2 years.

Discussion

Thickening of skin over palms and soles with constricting bands over the digits as in our case has been described in Vohwinkel's syndrome[3] and acral keratoderma.[4]

Vohwinkel reported a 24-year-old woman with rapidly progressive keratoderma affecting the acral areas.[3] The palmo-plantar keratoderma had a reddish blue border. Patient later developed pseudoainhum of several digits. It was inherited in an autosomal dominant fashion. Since then there have been reports of Vohwinkel's syndrome with star shaped lesions over knees and knuckles,[5] diffuse hair loss, high frequency neural hearing loss, spastic paraplegia, myopathy and ichthyosiform dermatoses.[6]

Acral keratoderma which is similar to Vohwinkel's syndrome differs from it by the absence of inflammatory halo and star fish keratosis over extensor surface of hands, presence of striate hyperkeratosis of palms in addition to diffuse keratoderma, presence of linear hyperkeratosis over acral areas like Achilles tendon, and pebbly appearance over dorsum of the finger. Histology shows a distinct type of focal dyskeratosis most striking in the malphigian layer. These patients also develop constriction bands over the digits. It was thought to be inherited in an autosomal recessive manner.

Our patient had diffuse thickening of soles of both the feet of late onset with auto-amputation of little toes. But he lacked the inflammatory halo around palmo-plantar keratoderma and star fish keratosis over extensor surface of hands characteristic of Vohwinkel's syndrome. Involvement of palms was limited to small area of hyperkeratosis over palmar aspect of middle finger of right hand and was non-progressive as described in acral keratoderma[4] but lacked other features. Thus our case may represnt a variant of Vohwinkel's syndrome or the rarer form of acral keratoderma.

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