OLMSTED SYNDROME

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A 20-year-old Sikh man had palmoplantar keratoderma, flexion deformity of digits, universal alopecia, keratotic plaques at the angles of mouth, gluteal cleft, knees and dorsal aspects of the metacarpophalangeal joints of the left hand; features of Olmsted syndrome. He had normal nails, teeth, oral mucosa and normal joint movements. Treatment with acitretin, 25mg/day for three and a half months, followed by 25mg once daily alternating with 50 mg once daily for 3 months resulted in significant improvement.

Key words : Olmsted syndrome, Genodermatosis, Palmo-plantar keratoderma

Introduction

Olmsted syndrome is a rare disorder characterised by palmoplantar keratoderma and keratotic plaques around the mouth, nose and anus. There is linear extension of the keratoderma onto the flexual aspects of the wrist and flexon deformity of the digits. It is often associated with constriction and autoamputation of the digits, onychodystrophy, follicular keratosis, congenital universal alopecia, hyperextensibility of the joints, absence of premolar teeth and keratosis of the oral mucosa. Fewer than ten cases[1-6] have been reported in the world literature and there has been one previous report of an Indian patient.[6]

Case Report

A 20-year-old man developed palmoplantar keratoderma at the age of 4 years. It began as a focal area of erythema and scaling and gradually progressed to cover the entire sole (Fig.1).The keratoderma was severe, surrounded by a rim of erythema and showed pits on the surface. Keratoderma remained confined to the feet till he was 16 years of age. At this time he developed thickening of the palms which extended linearly on to the flexual aspects of both wrists (Fig.2). Progression of the keratoderma had produced flexion deformities of the feet and digits of both hands over the last 4 years. He also had

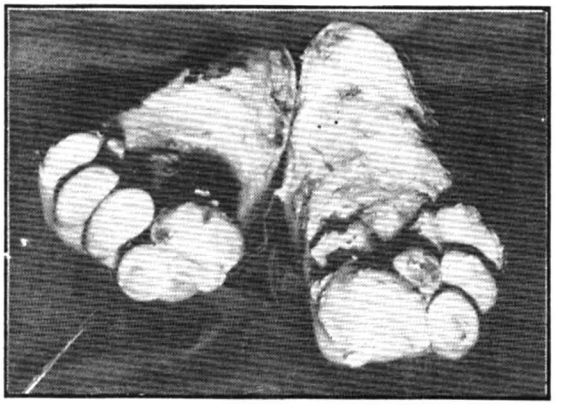

Fig.1. Massive keratoderma on the soles with flexion deformities of toes

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keratotic plaques over the gluteal cleft and the angle of mouth, both the knees and the dorsal aspects of the metacarpophalangeal joints of the left hand. He had no growth of hair on the scalp (Fig.3) or body since birth. The nail, teeth, and oral mucosa were normal. There was no hyperextensibility of the joints. No other member of the family had a similiar illness. He had been shaving the thick keratoderma with a safety blade and was using 40% urea under occlusion. At presentation, there was maceration, blackening and foul smell from the keratoderma on the feet. The haemogram, liver function test, kidney function test and lipid profile were within normal limits.The skin biopsy showed features of palmoplantar keratoderma. He was treated with hydrogen peroxide washes for the feet and oral metronidazole 400mg, thrice daily. This led to improvement in the foul smell and black, macerated areas on the sole resolved. He was given acitretin 25mg daily and asked to continue 40% urea cream under occlusion. On review, three and a half months later, there was slight improvement in the keratoderma. There were no side effects due to acitretin. The dose of acitretin was then increased to daily 50mg daily alternating with 25 mg daily. He was reviewed after 3 months and found to have a significant reduction in the keratoderma on the hands and feet.

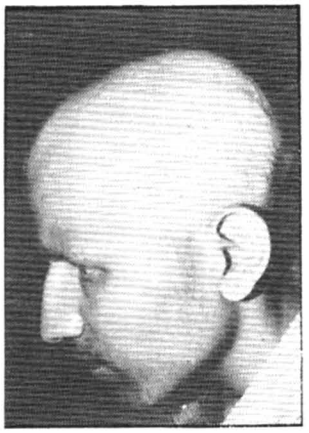

Fig.2 Congenital absence of hair on scalp

Discussion

In 1927, Olmsted[1] first described a patient who had well circumscribed palmoplantar keratoderma and sharply marginated hyperkeratosis around the mouth, nose and anus. In addition, the patient had flexion deformity of the digits, spontaneous amputation of the terminal phalanges and grossly dystrophic nails with subungual hyperkeratosis. Later on, scaly plaques developed over the dorsal aspects of the fingers and around the umbilicus. Subsequently, Costa in 1962[2] reported a patient who had palmoplantar keratoderma with thick keratotic plaques around the nose, mouth, anus and inner aspects of thigh. There was linear extension of keratoderma on to the flexural aspects of both wrists, flexion deformity of the fingers and grossly dystrophic nails. In addition, the patient had follicular keratosis of the extremities and leukokeratosis of the tongue. Poulin et al[3] in 1984, described a case of congenital sharply marginated keratoderma of palm and sole, periorificial keratosis, constriction of the digits, onychodystrophy, congenital universal alopecia, leukokeratosis of the tongue, hyperextensibility of the joints and absence of premolar teeth. Atherton et al[4] in 1990, and Cambiaghi et al[5] in 1995 reported familial cases of the disease. From India, Dogra et al[6] described a 8-year-old boy who had massive palmo-plantar keratoderma, keratotic plaques at the angles of mouth, elbows and knees, scanty hair showing defects

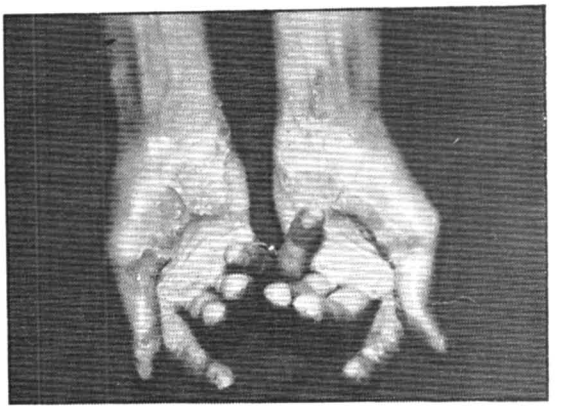

Fig.3. Linear extension of keratoderma on to the wrists. The patient had pared the keratoderma on the palms.

in the shaft, keratotic follicular papules and leukokeratosis.

Our case had massive palmoplantar keratoderma, with linear extension on the flexural aspects of both the wrists, flexion deformity of the digits, keratotic plaque at the angle of mouth, gluteal cleft, dorsum of the metacarpophalangeal joints of left hand, knee and alopecia universalis. All these findings are consistent with Olmsted syndrome. Our patient had no abnormalities of the nails, oral mucosa, teeth or joints.

The multilating keratoderma of Olmsted syndrome may be confused with Vohwinkels keratoderma or Mal de Meleda. However, the latter conditions lack the characteristic periorificial plaques. Acrodermatitis enteropathica may be misdiagnosed because of the keratotic plaques around the orifice. However, the condition does not respond to therapy with zinc. If available, estimation of serum zinc level can also be used to differentiate these conditions.

Treatment of Olmsted's syndrome is difficult. The keratoderma may be controlled by regular paring. Etretinate has been reported to partially relieve symptoms in 2 previous reports.[4,5] In our patient, acitretin produced significant improvement of the keratoderma after the dose was increased to 50 mg daily alternating with 25 mg daily.

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