Ind J Dermatol Venereol Leprol 1994; 60 : 359-61

PALMOPLANTAR KERATODERMA - MAL DE MELEDA TYPE

BS Yadav, SN Sonawane, Rohini P Deshpande, Ashish A Risbud

We report the first Indian family of Mal De Meleda type of Palmoplantar Keratoderma (PPK). Three sisters born of a consanguinous marriage showed features of Mal De Meleda type of PPK. The diagnosis was confirmed clinically.

Key Words : Mal de Meleda, Palmoplantar keratoderma (PPK), Indian family

Introduction

Mal de Meleda (Mijet Disease) is a very rare syndrome of hereditary PPK. It is named so from the Dalmatian island of Meleda where its relative frequency is due to inbreeding. The mode of inheritance is autosomal recessive or variable autosomal dominant when it is called Greither's Disease.[1] The keratoderma usually begins between 0-3 years of age and is characterised by 'transgradiens' i.e. extension on the dorsal surface of hands and feet like gloves and socks. It may be associated with hyperkeratotic plaques on the dorsa of hands, feet, knees, elbows and medial malleoli, erythema of palms and soles, erythema of nose, cheek and perioral eczema, hyperhidrosis and slow progression. Fingers may be short and nails may show koilonychia or subungual hyperkeratosis. Other rare features include lingua plicata, syndactyly, high arched palate and left handedness, and hair on the palms and soles.[2]

Case Report

Three sisters (out of five), aged six, four & one year and born of a consanguinous marriage between first cousins, presented with thickening of palms and soles since infancy, which was slowly increasing, and simultaneous appearance of thickened skin-coloured plaques on dorsal aspect of hands, feet, knees & ankles. There was hyperhidrosis of palms and soles & a history of similar palmoplantar thickening in the paternal grandmother of the children. The pedigree was as shown in Fig 1.

On examination, the six year old and four year old sisters showed diffuse PPK extending on the dorsal surface with sharp margins (Figs 2 & 3). The fingers were short. Nails were normal. Circular hyperkeratotic knuckle-pad like thickenings were present on all proximal interphalangeal joints and some knuckles (Fig 4). Irregular shaped hyperkeratotic plaques and linear keratoses were present on flexor aspect of wrists (Fig 2), and on dorsal aspect of feet, ankles and knees (Fig 5).

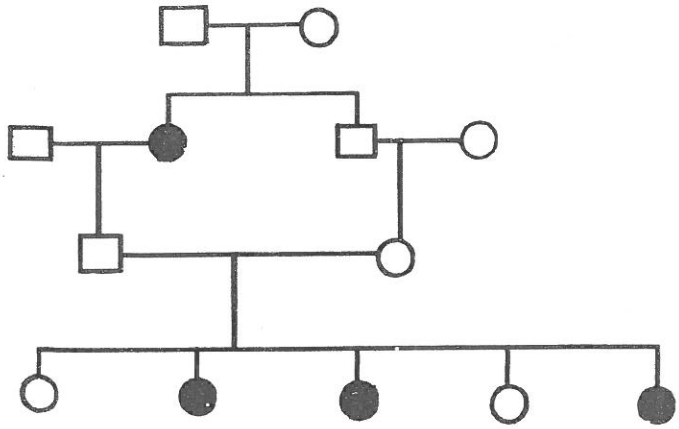

Fig. 1. Pedigree of the patients with Mal de Meleda.

From the Department of Dermatology & Venereology, Rajawadi Municipal General Hospital, Vidyavihar, Ghatkopar, Bombay - 400 077, India.

Address correspondence to : Dr BS Yadav, 11, Sheetal Dham, Plot No. 103, Garodia Nagar, Ghatkopar (East), Bombay - 400 077.

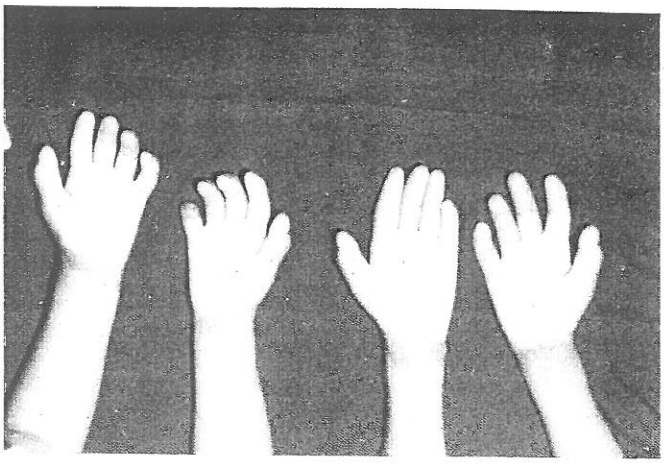

Fig. 2. Diffuse keratoderma of palms and soles with hyperkeratotic plaques on wrists ventrally.

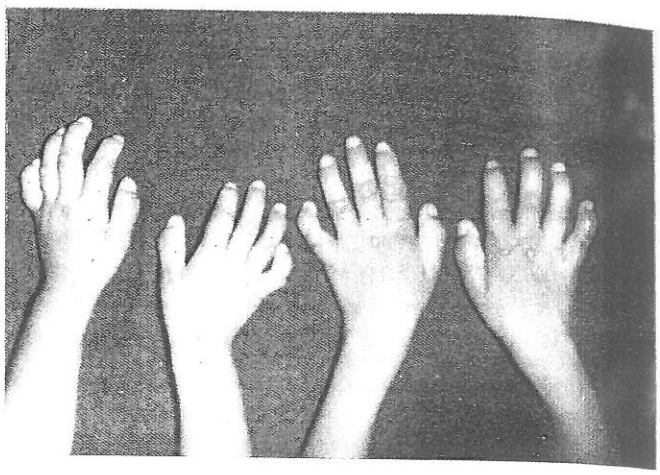

Fig. 3. Transgradiens with knuckle-pad like lesions.

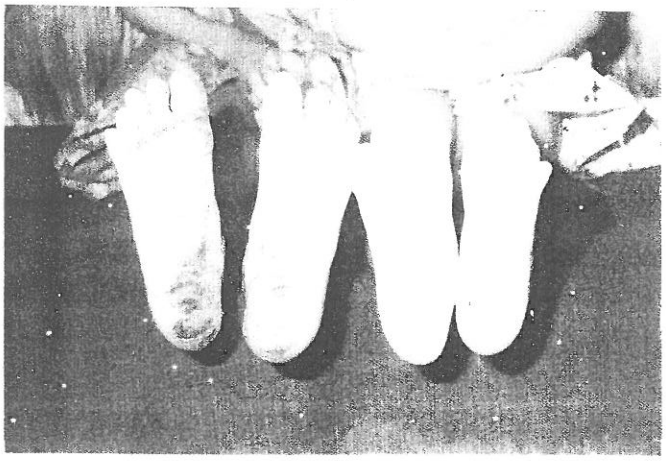

Fig. 4. Diffuse keratoderma of soles.

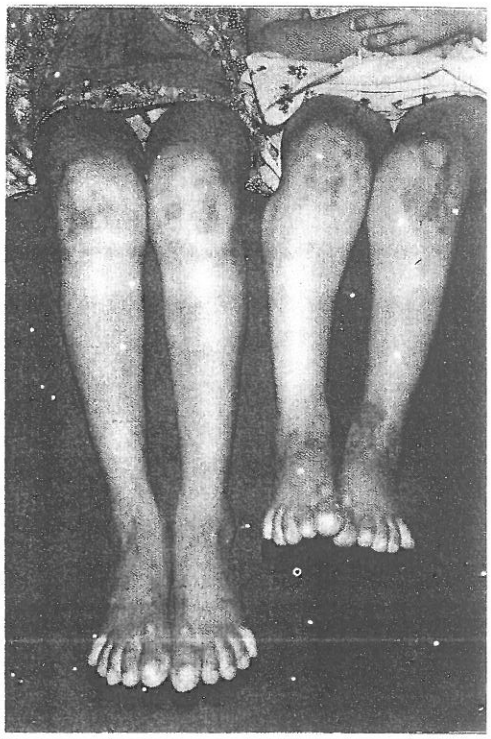

Fig. 5. Hyperkeratotic plaques on knees and ankles dorsally.

The youngest sister (one year old) showed mild keratoderma only on both palms but there was no hyperkeratotic plaque.

Histopathology revealed hyperkeratosis, hypergranulosis, acanthosis & chronic inflammatory dermal infiltrate.

General & systemic examination of all the sisters was normal. Eye & ENT examintion was normal. Intelligence testing of all sisters was normal.

The patients have been put on oral vitamin A[3] with vit. E and topical salicylic acid. They are under follow up at present.

Comments

Considering the consanguinous marriage of parents in our case, the possible mode of transmission is autosomal recessive. We made the diagnosis of Mal de Meleda based on the following features[1]

a. Autosomal recessive inheritance
b. Onset during the first year of life
c. Characteristic glove and socks like

hyperkeratosis with sharp margins.

d. Hyperkeratotic plaques on knees and ankles
e. Hyperhidrosis
f. Slow progression without remission.

The presence of hyperkeratotic plaques looking like knuckle pads were present on all the proximal interphalangel joints and some knuckles of both hands of the two elder sisters. Such knuckle pads have been described in one case of hereditary epidermolytic PPK (Vorner type).[4] Our cases are examples of knuckle pads associated with Mal de Meleda.

Rarely ainhum-like lesions are described in cases of Mal de Meleda.[5] But usually they appear relatively late in the course of the disease. Hence the follow up in cases of Mal de Meleda keratoderma should be maintained for many years, in case mutilations develop.

References

1. Jee Sh, et al. Report of a family with Mal de meleda in Taiwan, a clinical, histopathological and immunological study. Dermatologica 1985; 171: 30-7.
2. Salaman T, Berberovic L, Topic B et al. Mal de Meleda-Data & Remarks on a series. G Ital Dermatol Venereol 1988; 123: 649-55.
3. Brunner MJ, Fuhrman DL. Mal de Meleda-Report of a case and results of treatment with Vitamin A. Arch Dermatol Syphilol 1950; 61: 820-3.
4. Nogita T. et al. Hereditary epidermolytic PPK with knuckle pad like lesions over the finger joints. Br J Dermatol 1991; 125: 196.
5. Brambilla L, et al. Unusual cases of Meleda keratoderma treated with aromatic retinoid etretinate. Dermatologica 1984; 168: 283-6.