Mal de Meleda: A report of two siblings in one family

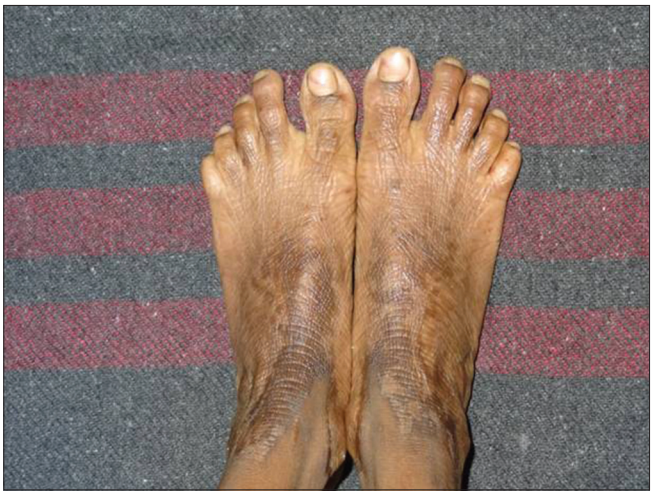
Sir,

Two siblings female and male of age 12 and 10 years born out of a second degree consanguineous marriage presented with a small patch of asymptomatic peeling of the skin over the palms since birth. The patch started increasing at the age of nine months to involve both palms and soles and spread to the dorsal aspect of hands, feet, and extensor knees and elbows. Developmental milestones were normal and performance in school was above average. There was no history of hearing impairment, eye problem, or dental caries.

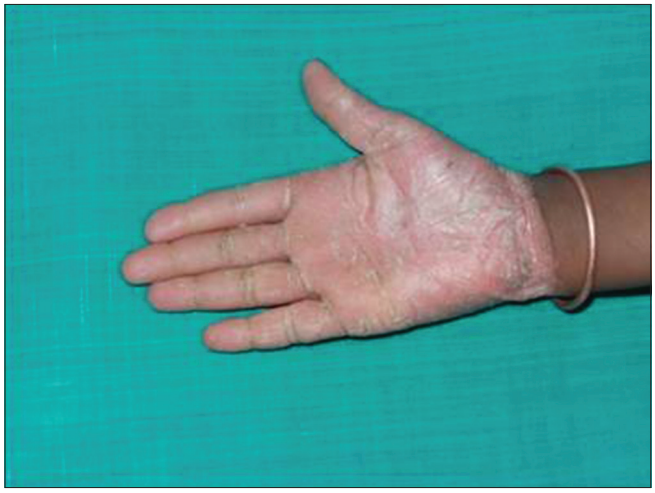
On examination, both the siblings showed diffuse palmoplantar keratoderma (PPK) extending proximally up to the wrist and dorsum of hands and feet with well-defined margins, in a transgrediens pattern [Figures 1-4]. The skin over the palms and soles was thick and erythematous with loss of dermatoglyphics and fissures. The severity of skin lesions was more in the male sibling. Hyperhidrosis of palms and soles was also noted in the male. Well-defined erythematous plaques were present over the extensor aspect of the elbow and knee joint. The nails were normal. Histopathology of skin lesions showed hyperkeratosis, hypergranulosis and acanthosis and a perivascular mononuclear cell infiltrate in the upper dermis.

The PPKs are a heterogenous group of disorders, characterized by thickening of the skin of the palms and soles. They can be categorized as inherited and acquired, or subdivided based on clinical patterns. Mal de Meleda (MDM), one of the autosomal recessive forms of PPK, is clinically characterized

Figures 1: Transgradient palmoplantar keratoderma with sharp margins in the girl sibling



Figures 2: Transgradient palmoplantar keratoderma with sharp margins in the girl sibling



Figures 3: Transgradient palmoplantar keratoderma with sharp margins in the boy

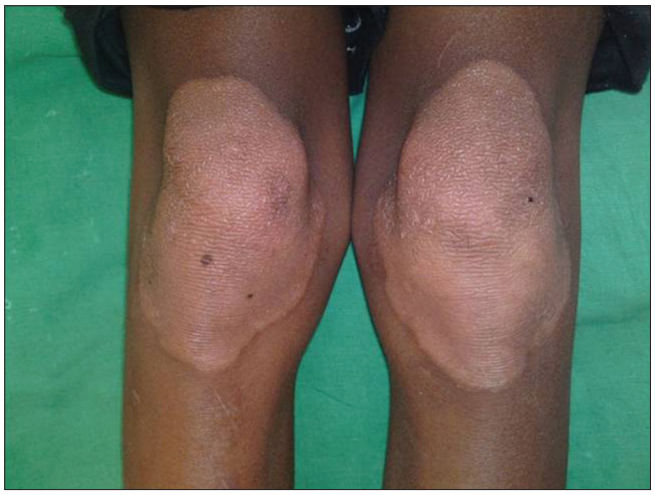


Figure 4: Transgradient palmoplantar keratoderma with sharp margins on both knees in the boy

Table 1: Differential diagnosis of MDM

Syndromes	Clinical features
Gamborg-Neilsen variant of MDM	Has less severe hyperkeratosis and no distant keratosis
Greither's syndrome[2]	Autosomal dominanat inheritance, progressive evolution and presents with epidermolysis
Papillon-Lefèvre syndrome[3]	Autosomal recessive inheritance, diffuse keratoderma with gingivitis, early loss of teeth, periosteal changes and intracranial calcification
Richner-Hanhart syndrome	Keratoderma, mental retardation, raised tyrosine and tyrosine metabolite levels
Vohwinkel's syndrome[4]	Autosomal dominant inheritance, mutilating palmoplantar keratoderma, alopecia, icthyosis and deafness
Unna Thost syndrome[5]	Autosomal dominant inheritance, diffuse non-transgressive palmoplantar keratoderma, mutilation in keratin 9 gene
Naxos disease	Autosomal dominant inheritance, deletion in plakoglobin gene, non-transgressive palmoplantar keratoderma, arrhythmogenic right ventricular cardiomyopathy and wolly hair
Huriez syndrome	Autosomal dominant inheritance, diffuse palmoplantar keratoderma, sclerodacttyly, atrophy and koilonychia

MDM: Mal de meleda

by a well-demarcated erythema and hyperkeratosis of the palms and soles that usually occurs soon after birth. The hyperkeratosis spreads slowly to the dorsal aspects of the hands and feet, which is referred to as transgrediens. Many of the patients will experience bothersome pain due to fissures. Hyperhidrosis with maceration is feature of MDM and is often accompanied by malodour. In addition, patients may develop keratotic plaques over joints, nail abnormalities, brachydactyly, pseudoanihum and perioral erythema.[1] In 2001, mutations in the gene encoding secreted Ly-6/uPAR-related protein 1 (SLURP-1) located on the chromosome 8q24.3 were found to be the cause of MDM. It is currently identified that SLURP-1 is an epidermal secreted neuromodulator that influences both epidermal homeostasis and inhibition of tumor necrosis factor-alpha release by the macrophages during the wound healing process. Such roles would explain the hyperproliferative and inflammatory clinical characteristics of MDM. Both our cases showed the following compatible clinical features: "glove-and-socks" distribution of the keratoderma with a sharp demarcation that appeared after birth and progressively extended to the dorsa of the hands and feet and palmoplantar hyperhidrosis. Both siblings were treated with acitretin 10 mg and improvement was noted within one month.

MDM must be differentiated from other syndromes as shown in Table 1.

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