A rare case of Mal de meleda keratoderma with pseudoainhum

Dr Geetika Chhabra
Department of Dermatology
Vardhman Mahavir Medical College & Safdarjung hospital, New Delhi, India
Poster ID 26

Introduction

Mal de Meleda is a rare form of autosomal recessive keratoderma and is named after Mljet Island in Croatia.

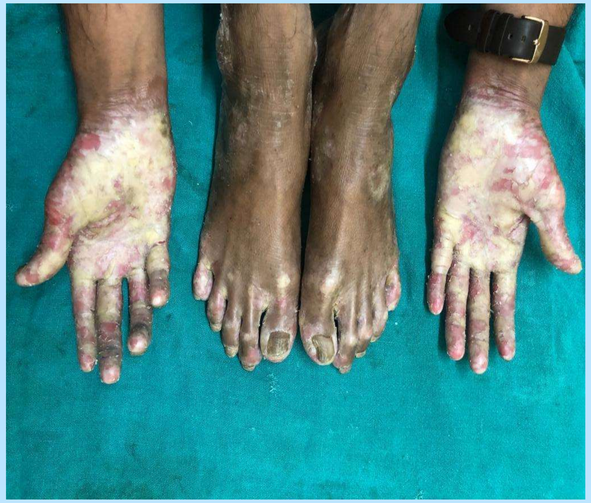
Mal de Meleda has an estimated prevalence of 1:100,000.

Pseudoainhum may complicate the course of palmoplantar keratodermas and has rarely been described in Mal de Meleda keratoderma.

Case report

- A 25-year-old man with progressive thickening of the skin over palms and soles along with nail changes since 6 months of age. (Fig 1)
- parents had a consanguineous marriage
- He had 3 siblings; one sister being affected by the same condition; parents were not affected though.
- Examination revealed diffuse waxy, thickened and macerated skin of palms and soles along with transgrediens.
- Constriction bands, pseudoainhum with flexion deformity due to contracture was observed distal to proximal interphalangeal joints of little finger of both hands.
- prominent knuckle pads on dorsum of both metacarpophayngeal joints, and all finger and toenails were dystrophic.
- Rest of the mucocutaneous examination was normal.

Fig 1



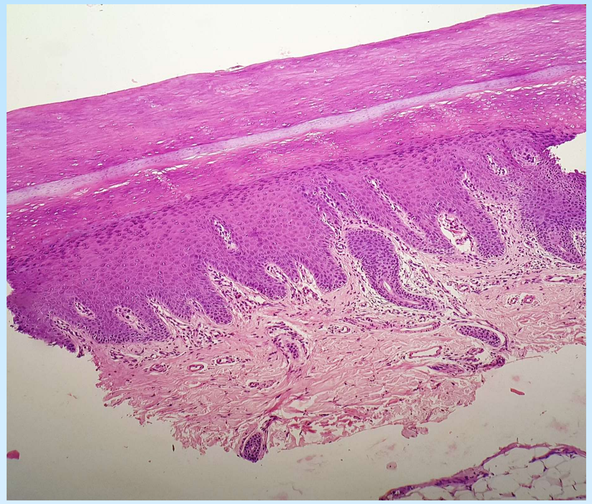


Fig 2

Histopathology

High power view showing a greatly thickened stratum corneum, an accentuated normal basket weave pattern of orthokeratosis producing a vacuolar appearance in the stratum corneum, prominent keratohyalin granules and acanthotic epidermis without epidermolysis. (Fig 2) (H&E, 200X)

Discussion

[2]

Mal de Meleda clinically, the onset of the disease is typically soon after birth and is characterised by transgrediens and progrediens of hyperkeratosis of the palms and soles.

Pseudoainhum describes constriction band around digit as a result of an associated disease process, which can lead to autoamputation. Pseudoainhum can be further classified as either primary, whereby it presents at birth or early life, or secondary when presenting later in life.

Pseudoainhum is an infrequent complication in Mal de Meleda secondary to severe hyperkeratosis. Treatment of pseudoainhum involves surgical release and/or oral acitretin.

Reference

- Bakija-Konsuo A, Zitinski M, Fatovic-Ferencic S. Mal de Meleda: A great imitator. Clin Dermatol. 2019; 37: 175-181.
- Perez C, Khachemoune A. Mal de Meleda: A Focused Review. Am J Clin Dermatol. 2016; 17: 63-70.