

Mal de Meleda: A Case report in Eastern india

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Abstract:- Mal de Meleda is a rare (Prevalence 1:1,00,000) autosomal recessive, diffuse, transgradient, nonsyndromic, non-epidermolytic, palmoplantar keratoderma (PPK) with associated scleroatrophy, nail changes, pseudoainhum around digits & perioral erythema without a tendency of spontaneous resolution. We present here a case of Mal de Meleda. A 10 year old male student from rural area presented with keratoderma involving palms, soles, dorsum of foot & hand along with involvement of knee & elbow. Diagnosis was confirmed by histopathological examination which showed hyperkeratosis without epidermolysis. This type of PPK may be confused with others types (Greither's disease, Huriez syndrome, Olmsted's syndrome, Vohwinkel's syndrome, Papillon Lefevre syndrome) of PPKs & disorders of cornification.

Keywords:- Mal de Meleda, Transgradient, Non-epidermolytic, palmo-plantar keratoderma.

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I. Introduction

Mal de Meleda (A/k/a keratoderma palmoplantaris transgrediens of siemens, acroerythrokeratoderma, keratosis extremitatum hereditaria progradiens)^[1] is a rare (Prevalence 1:1,00,000) autosomal recessive, nonsyndromic, nonepidermolytic, diffuse, transgradient keratoderma named after the Croatian island of Meleda (Mljet) where it was first identified.^[2] Almost all cases occur out of consanguineous marriage.

This keratoderma begins in early childhood (Between birth to 3 years) & is characterized by transgradient keratoderma with sharp demarcation, erythema with associated scleroatrophy, nail changes, pseudoainhum and perioral erythema without a tendency for spontaneous resolution. The present report deals with a case of Mal de meleda syndrome with typical (clinical & histopathological) features of the syndrome.

II. Case Report

A 10-year old male from Bundu presented in Dermatology OPD of Skin, S.T.D. & Leprosy Department, RIMS, Ranchi, Jharkhand. He presented with erythematous dry, scaly, fissured thickening of the skin on palms and soles, extending towards dorsal surface of foot & sole & proximally towards distal forearm & leg. He also had psoriasiform plaques on knee & elbow, thickening of knuckle & dry erythematous thickening of lips. He had all lesions from 6 months of age.

First he had developed erythema & then gradually thickening of palm & sole. Then gradually it involved dorsal aspects of the palms and soles & distal parts of leg & forearm with increasing age. Gradually He also developed pigmented, rough, raised grouped lesions on the elbows and knees from 5 years of age, which progressed to form well defined erythematous scaly psoriasiform plaques. There was redness of lips since childhood, which developed into mild thickening and dryness recently. He was born of second-degree consanguineous marriage. Developmental milestones were normal and performance in school was above average.

There was no history of hearing impairment, eye problem or dental caries. Two other siblings, parents and relatives, did not have similar complaints. On examination, there was diffuse palmoplantar keratoderma involving bilateral palms and soles and extending proximally up to the wrists and the dorsa of hands and feet & distal parts of legs & forearms with well-defined margins (Figures 1,2,3,4). The skin on the palms and soles was thick, waxy and yellow with loss of dermatoglyphics. Thick, hyperkeratotic, scaly plaques were seen on proximal interphalangeal joints resembling knuckle pads. There were a few areas of erosions in the web spaces. Diffuse, well-defined, irregular shaped erythematous plaque with grayish white scales on both extensor aspects elbows & knees (Figures-2,4). Nails were thickened with hypercurvature, longitudinal ridges, hyperpigmentation

and prominent lunulae. The great toenail of both foots were thickened with subungual hyperkeratosis. Soles were dirty looking with fissuring. Lips were erythematous, dry with mild thickening (Figure 5).

Histopathology of palmar skin showed hyperkeratosis, hypergranulosis, and acanthosis. There was no feature of epidermolytic hyperkeratosis.

A diagnosis of Mal de Meleda was made based on the above-mentioned features.

Photographs



Figure 1: Plantar Keratoderma involving soles



Figure 2: Keratoderma involving dorsum of foot & distal parts of legs along with Psoriasiform plaque over knees.



Figure 3: Palmar Keratoderma



Figure 4: Keratoderma extending beyond wrist along with Psoriasiform plaque over elbows.



Figure 5: Perioral erythema with lip involvement

III. Discussion

The obligatory clinical features of Mal de Meleda are ^[3]

1. autosomal recessive inheritance
2. onset of diffuse keratoderma palmoplantaris soon after birth (or up to 3 years of life) ^[1]
3. transgressive and progressive nature of the keratoderma which tends to involve the dorsa of hands and fingers, feet and toes, flexor aspect of the wrist (“glove and stocking keratoderma”) with sharp margin. ^[1]
4. Hyperkeratotic plaques resembling knuckle pads may be seen on the interphalangeal joints. ^[1]

The facultative clinical features are ^[3]

1. palmoplantar hyperhidrosis
2. pitting in the keratoderma palmoplantaris
3. lichenoid polycyclic plaques on the elbows, knees, and groins,
4. subungual keratosis, koilonychia, dystrophy of the great toenail
5. progressive conical tapering of the fingertips which may lead to contractures of the fingers
6. perioral erythema
7. high arched palate
8. corneal lesions.
9. Digital constrictions (pseudoainhum) and angular cheilitis ^[2]

Other rare features include lingua plicata, syndactyly, left handedness and hair on palms and soles. ^[1] The involvement of the lips as was seen in our case is unique.

There is a great interindividual variation in the clinical manifestations of Mal de Meleda which should be differentiated from other similar palmoplantar keratoderma syndromes. ^[3] Greither’s disease has several clinical similarities with Mal de Meleda syndrome, but can be differentiated based on the following features. Greither’s disease is autosomal dominant, starts during 8-10 years of age with a gradual onset and a tendency to improve with age. ^[2] Mal de Meleda is autosomal recessive, appears early after birth and slowly progresses without any tendency for spontaneous involution. ^[4] Both have transgradient keratoderma and hyperkeratotic

plaques on the extensor aspects of the knees and elbows. However, perioral erythema is seen in Mal de Meleda syndrome.^[3]

In contrast to Mal de Meleda syndrome, the palms and soles may be spared in Greither's disease.^[4] Conical tapering of the fingertips seen in Mal de Meleda syndrome is not seen in Greither's disease.^[3] Constrictive bands and nail changes are frequent with Mal de Meleda, but unusual in Greither disease.^[5] Mal de Meleda syndrome also has typical nail changes.

Huriez syndrome (also referred to as "sclerolytosis") is an autosomal dominant genodermatosis, characterized by the triad of congenital diffuse scleroatrophy of the distal extremities, mild or lamellar keratoderma of the palms and to a lesser extent the soles, present from birth and ridging or hypoplastic nail changes.^[6,7] There is also an absence of dermatoglyphics.^[2] Atrophic parchment-like skin is present on the dorsal surface of the hands. Dense hyperkeratosis gives a pseudosclerodermatous appearance with nail atrophy.^[2] Nail changes include hypercurvature, longitudinal ridging, onychorrhexis, and koilonychias.^[2] Another prominent feature of Huriez syndrome is almost invariable family history.^[8]

Olmsted's syndrome is a severe, cicatrizing transgradient palmoplantar keratoderma with keratotic plaques around the mouth, nose and anus. Onset is in the first year of life, with symmetrical, sharply defined palmar and plantar keratoderma surrounded by erythema, flexion deformities, constriction or spontaneous amputation of the digits, onychodystrophy, follicular keratosis, congenital universal alopecia, hyperextensibility of the joints, absence of premolar teeth and keratosis of the oral mucosa.^[2,9]

Vohwinkel's syndrome is an autosomal dominant palmoplantar keratoderma characterized by honeycomblike keratoderma beginning early in life associated with stellate or "starfish" keratoses on the dorsa of hands and feet, and the formation of circumferential bands around digits ("pseudo-ainhum") and sensorineural deafness.^[2,10] Vohwinkel's syndrome was ruled out in our case by the absence of honey-comb keratoderma and sensorineural hearing loss. Camisa's syndrome is a variant of Vohwinkel's syndrome where sensorineural hearing impairment is absent, but these patients have mild ichthyosis^[11] which was not seen in our case.

Papillon Lefevre syndrome (PLS) is characterized by palmoplantar hyperkeratosis, severe periodontitis and premature loss of teeth. It manifests by 1-4 years of life and patients become edentulous by early teens.^[12] PLS was easily ruled out in our case by the presence of normal teeth and the absence of periodontitis. Here we present a case of Mal De Meleda which is a rare variant of palmoplantar keratoderma.

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