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## **Palmo-Plantar Keratoderma**

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Palmo-Plantar Keratoderma is a hereditary disorder, in which there is localized or diffuse thickening of the stratum corneum which primarily involves soles and palms but it may be a part of generalized disorder. We admitted such a case which we are reporting as under.

### **Case Report**

A thin lean man, H.S. age 51-years from Moh: Zangi Khel, Village Dargai, District Swabi, N.W.F.P. came to skin O.P.D. on 24-8-91 with a complaint of thickening and peeling of skin of palms and soles for the last 51-years i.e. since birth. The thickening and peeling of the skin of soles and palms started when he was 40- days old. The peeling starts now with the formation of vesicles on the adjacent sides which rupture and make the thickened skin to peel of. Itching was also a complaint, rarely when vesicular formation occurred.

He has three brothers and two sisters. Parents are alive. Two brothers alongwith him and one sister are the effectees. The eldest sister is spared and the brother, who was 2nd: junior to him, is also free from the problem. The rest all had the disorder within first six months of their lives. Now the rest, all living (except him) are crippled and they live bed-ridden life, but the parents along with all blood relatives are living normal life.

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Bilateral diffuse hyper-keratosis  
of the palms and soles.





Contracture of the metacarpo-phalangeal joints and circumscribed lesions showing hyper-keratosis

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**EXAMINATION:** On examination there was bilateral diffuse hyperkeratosis of the palms and soles involving the volar aspects of the digits. There was contracture of the metacarpo-phalangeal joints. Nails showed dystrophy (Picture-No: 1-2). Hair of the scalp were normal along with teeth. Circumscribed lesions showing hyper-keratosis of the knees and elbows were also noted. (Picture No: 3).

### **Differential Diagnosis of Hyperkeratosis**

1. **PSORIASIS:** It is genetically determined erythro-squamous disorder of the skin characterized by lesions: (a) which are sharply demarcated with clear-cut borders; (b) surface consists of easily peelable silvery scales; (c) under the scale, skin has glossy homogeneous erythema; and (d) they exhibit Auspitz sign which is specific diagnostic feature of erythro-squamous lesion of psoriasis in which small blood droplets on the erythematous surface appear within a few second after mechanical removal of the scales.

2. **ECZEMA:** It is a distinctive reaction of the epidermis to a variety of agents both endogenous and exogenous: characterized clinically by eruption of itchy erythematous plaques and vesicles leading to weeping and/or lichenification and histological evidence of spongiosis.

3. **LICHEN PLANUS:** Dermatitis of unknown aetiology characterized by typical violaceous papular eruption on characteristic site and white mucosal lesions. Papules are characteristically

- a) Plane topped,
- b) Purple coloured,
- c) Pruritic and
- d) Polygonal.

Sites are wrists, lumber region, lower legs and ankles.

4. **SYPHILIS:** It is chronic contagious, principally sexually transmitted disease, in which primary chancre occurs on the glans of the penis and female genitalia.

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5. **PITYRIASIS RUBRA PILARIS:** It is a chronic disorder characterized by reddish orange scaling plaques and keratotic follicular papules.

Types are familial and acquired. Familial type has an autosomal dominant mode of inheritance. Redness and scaling of the face and scalp are often seen first and are followed by redness and thickening of stratum corneum of palms and soles. The follicular papules are characteristic of the disease, which soon appear as a keratotic plug of the follicles surrounded by erythema. They are most commonly seen on dorsal aspect of proximal phalanges, elbows and wrists. A more wide-spread eruption consisting of scaling orange plaques is observed on the trunk and extremities. The lesions have sharp borders and island of normal skin within them are characteristic. The lesions may coalesce and expand and eventually cover the entire body.

6. **TINEA PEDIS:** Dermatophytic infection of feet: toes and interdigital clefts. Interdigital peeling, maceration and fissuring are common. Itching and hyperhidrosis are often associated. Occasionally there is vesico-bullous reaction on the palms and soles; and rarely hyperkeratosis, scaling and fissuring of the soles and heels. The nails are often infected.

Other common disorders associated with palmer and plantar hyperkeratosis are:

- a) Yaws.
- b) Reiter disease.
- c) Hidrotic ectodermal dysplasia.
- d) Franceschetti-jadassohon syndrome.
- e) Epidermolytic Hyperkerotisis.
- f) Dyskeratosis cogneita.
- g) Epidermal nevus.

### **Discussion**

Palmo-plantar keratoderma is a term which refers to a condition in which thickening of stratum corneum occurs, localized or diffuse, primarily involving the palms and soles but may be a part of generalized disorders. Many of these disorders are hereditary and can be classified on the basis

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of morphology of the lesion

**Clinical Manifestations:**

1. Keratosis Palmaris et - Plantaris (Unna Thost disease).
2. Howel Evans Syndrome.
3. Mal-de-Meleda.
4. Papillon Lefevre.
5. Keratosis Punctata.
6. Richner Hanhart Syndrome.
7. Pachyonychia-congenita.
8. Acrokerato Elastoidosis.
9. Keratosis Striata.
10. Keratoma Hereditarium Mutilans.
11. Keratoderma Climactericum

**KERATOSIS PALMARIS ET - PLANTARIS:** (Unna-Thost disease): Keratosis palmaris plantaris is a rare, dominantly inherited disorder, first described by Thost, with variable involvement of the palms and soles. Other forms of inheritance rarely have been reported. The disorder usually appears in infancy but a number of cases have first occurred in childhood. The involvement is generally bilateral and symmetrical with either circumscribed or diffuse lesions. The lesions may extend into contiguous skin; hyperkeratosis of the knees and elbows has also been observed, to occur with keratosis pilaris. The palms may have a smooth, waxy appearance or a dry surface with fissures. While the borders may be surrounded by an erythematous halo. Involvement may be different in affected members of the same family. Hyperhidrosis and bromhidrosis can be a major problem. There have been reports of alopecia and thickened nails but these are not common or consistent features. Teeth are normal.

**HOWEL EVANS SYNDROME:** The Howel Evans syndrome has been observed in two English families perhaps related, as a dominantly inherited diffuse keratoderma of the palms and soles which appears between the ages of 5 and 15 years. Carcinoma of the oesophagus is seen in most but not all affected members of the family.

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**MAL-DE-MELEDA:** Mal de Meleda is a rare, recessively inherited disorder described in individuals living on the adriatic island of Meleda. The condition is evident at birth or shortly there after and persists throughout life. There is diffuse involvement of the palms and soles, starting with redness and then progressing to heperkeratosis which spreads onto the dorsa of the hands and feet; patients may also develop circumscribed lesions of the extremities and erythematous plaques in the groin and axilla which resemble psoriasis. The typical palmar and plantar lesions show accentuation of the creases with darkening of the surface, sometimes accompanied by deep fissuring. No specific defects of teeth, hair, or eyes have been described; however, nail changes are common.

**PAPILLON-LEFEVRE:** Papillon-Lefevre is an autosomal recessive disorder starting within the first 6-months of life, which is manifested as redness and diffuse but sometimes localized heperkeratosis of the palms and soles, gingivitis and periosteal changes of the alveolar bone, and calcification of the falx and tentorium. Extension of the disorder on the dorsa of the hands and feet and psoriasiform changes of the elbows and knees. There is loss of the permanent as well as the deciduous teeth. Hair is usually normal.

**KERATOSIS PUNCTATA:** Keratosis Punctata is an autosomal dominant disorder developing gradually in the first decade and presenting with discrete keratosis of the palms and soles. There may be more than 200 lesions on each extremity. The lesions are usually not more than several millimetres in diameter, yellowish to dark brown in colour, and raised upto 2mm. The central keratin plug may be lost, leaving a pit with a hyper-keratotic base. Pain may be present in the feet from pressure during walking, hyperhidrosis is usually not observed.

**RICHNER HANHART SYNDROME:** It is an autosomal recessive disorder consisting of corneal dystrophy, brady-phalangia, oligo-phrenia and prepatellar and palmo-plantar keratosis. The patients have tyrosinemia and when placed on a low tyrosine diet, the cutaneous and ocular manifestations disappear.

**PACHYONYCHIA CONGENITA:** It is a dominantly inherited disorder which is manifested primarily by hyperkeratotic lesions of the nail beds. In

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addition, patients have hyperkeratosis of the plantar skin in areas of pressure as well as bullae of that skin.

**ACROKERATO ELASTOIDOSIS:** It is a disorder most commonly seen in adult women in which skin-colored to yellow papules are seen primarily on the plantar surfaces of the hands and feet, but some lesions are present on the dorsal surfaces as well.

**KERATOSIS STRIATA:** It is an autosomal dominant disorder that most commonly presents as linear keratosis radiating from the palm onto the volar aspects of the digital creases. The process usually starts in the first decade and may show varying types of lesions as well as involvement of the soles.

**KERATOMA HEREDITARIUM MUTILANS:** It is usually the disorder of childhood, autosomal recessive in nature, occurring as keratotic lesions of the palms or soles, extending onto the digits. Star-shaped lesions develop on the dorsa of the digits. The borders of the lesions are red and compress the underlying tissues as the keratosis extend, resulting in constriction and eventual amputation of the digit.

**KERATODERMA CLIMACTERICUM:** It appears in women at the time of menopause and manifests itself as circumscribed hyperkeratosis of the palms and soles, often associated with obesity. The lesions vary in size and are prone to occur at the site of trauma.

**DIAGNOSIS:** The patient, we are discussing, had the problem of palmo-plantar keratoderma among the other keratotic disorders of the palms and soles occurring as a part of generalized disorders already mentioned.

Among the various types of palmo-plantar keratoderma, the patient we admitted belonged to the Papillon Lefevre type on the basis of: (a) history, (b) clinical features and (c) biopsy

(a) History: History of the patient clearly revealed that patient is suffering from autosomal recessive type of the palmo-plantar keratoderma.



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(b) Clinical Features showed that the disorder was Papillon Lefevre type. As the examination revealed that there was bilateral diffuse hyperkeratosis of the palms and soles involving the volar aspects of the digits; there was contracture of the metacarpo-phalangeal joints along with atrophy of interossei and lumbrical muscles. Nails showed dystrophy. Hair were normal. Circumscribed lesions showing hyperkeratosis of the extremities i.e. knees and elbows were observed with psoriasis appearance. There was loss of permanent teeth except the upper two incisors. The gums were swollen, reddish black in colour. X-Ray skull showed periosteal changes of the alveolar bone.

(c) Biopsy: Biopsy of the skin was taken from the mole of left foot, according to which there was marked acanthosis with irregular type of hyperkeratosis with some prominent perivascular infiltrate of lymphocytes and histiocytes. Thus confirming the diagnosis.

**TREATMENT:** We treated the patient with Seproderm ointment (a salicylic acid preparation) along with vitamin A. Salicylic acid is a keratolytic agent which helps in removing stratum corneum. Management of hyperkeratosis is done on conservative lines. Space shoes which allow an even distribution of the pressure are helpful in relieving the symptoms. The lesions may be pared with a blade or rubbed with a pumice stone, which is done when the parts are wet. Lubrication with ointments and cream after soaking in water is helpful in softening the skin. Bromhidrosis is most readily controlled by painting the soles with 10% glutaraldehyde solution. Isotretinoin in doses of 2mg/kg per day has been shown to markedly reduce the hyperkeratosis, but must be given continuously or the disorder recurs.

**PROGNOSIS/FOLLOW-UP:** The prognosis of palmo-plantar keratoderma is very distressing for the patient as it is a genetically determined disorder and continues throughout the life of the patient. The symptoms can be relieved by the conservative treatment which has to be continued through out the life.

We gave the patient the salicylic acid preparation (Seproderm ointment). The patient was admitted for 3 weeks and was relieved partly by the above mentioned treatment. We asked the patient to come back after 15 days but he did not reappear, most probably due to poverty and due

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to relief from the symptoms.

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