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Palmo Plantar Keratoderma in Iraqi Patients A Clinical Study

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Abstract

Palmo plantar keratoderma is a term that means marked thickening of the skin of the palms of the hands and soles of the feet. It may be either diffuse affect most of the palms and soles or focal mainly affect pressure areas. Keratoderma may be inherited (hereditary) or more commonly acquired. Palmoplantar keratoderma is not an uncommon disease. The aim of this work was to shed light on causes and clinical presentation of this condition in Iraqi patients at Hilla city.

A total of 60 patients with palmoplantar, palmar and plantar keratodermas were seen. Their ages ranged between 5–70 years (mean ± SD, 24.9 ± 18 years), while their ages of onset averaged from since birth or few months after birth to 68 years (mean ± SD, 17.9 ± 18.5 years). The duration of the disease ranged from 1-32 years (mean ± SD, 6 ± 7.2 years). There were 40 males (66.6٪) and 20 females (33.4%) with a sex ratio of 2 : 1.

Acquired causes of palmoplantar keratoderma were seen in 83.3٪ included the followings: psoriasis (66.6٪), cumulative irritant contact dermatitis (5٪), occupational (5٪), lichen planus (3.3٪), pityriasis rubra pilaris (1.6٪) and Reiter syndrome/ keratoderma blenorrhagicum (1.6٪). Hereditary causes were seen in 16.7 ٪ of cases and included the followings: Papillon–lefevr'e syndrome (3.3٪), congenital diffuse non–epidermolytic palmoplantar keratoderma/Unna–Thost(3.3٪), X–linked ichthyosis (1.6٪), Lamellar ichthyosis (1.6٪), Ichthyosis vulgaris (1.6٪), Striate keratoderma (1.6٪), Acrodermatitis entro pathica/primary zinc defiency (1.6٪) and palmoplantar keratoderma areata (1.6 ٪).

Psoriasis was the most common acquired cause of palmoplantar keratoderma in this study (66.6٪). Papillon – lefevre syndrome and Unna – Thost were the main causes of hereditary palmoplantar keratoderma (3.3٪). From a hereditary causes, Striate keratoderma, a focal palmoplantar keratoderma, autosomal dominant, as a part of epidermal nevus syndrome was the first reported case that show an association of epidermal nevus with rhabdomyosarcoma of the neck and eye proptosis . Family history was positive in 21.6 ٪ .

No previous study had evaluated palmoplantar keratoderma in our country, so this report was a first step towards shedding light on this subject .

الخلاصة

تم دراسة 60 حالة تقرن جلدي تشمل باطن اليدين والقدمين كليهما أو باطن اليدين فقط أو باطن القدمين فقط. تراوحت أعمار المرضى ما بين 5 – 70 سنة ومدة الاصابة تراوحت ما بين 1– 32 سنة . بلغ عدد الذكور40 (66.6 ٪) وعدد الاناث 20 (33.4 ٪) ونسبة الذكور إلى الاناث بلغت 2 : 1. مثلت الاسباب المكتسبة لمرض التقرن الجلدي نسبة 83.3 ٪ وشكل مرض الصدفية الجزء الاكبر (66.٪). أما الاسباب الوراثية لمرض التقرن الجلدي فشكلت نسبة (16.7 ٪) وشكلت متلازمة بابلون لفقري ومتلازمة أنّا – ثوست نسبة (3.3 ٪) لكل واحدة منهما .

Introduction

Keratoderma is a term that means marked thickening of the skin. Palmoplantar refers to the palms of the hands and soles of the feet ; these are the areas, keratoderma affects most often. Palmoplantar keratoderma is also sometimes known as keratosis palmaris et plantaris (Kimyai ,2002; Odom *et al.*, 2006). Classification of keratodermas depends on whether or not it is inherited and it's clinical features :

1. Diffuse keratodermas affect most of the palms and soles .
2. Focal keratodermas mainly affect pressure areas.
3. Punctate – type keratodermas result in tiny bumps on palms and soles(Stevens and Leigh 1999; Sybert, 1988).

Most often the abnormal skin involves only the palms and soles (non – transgradient) but sometimes it extends on to the top of the hands and feet as well (transgradient). In some rare froms of keratoderma, other organs in the body may be affected in addition to the skin and the keratoderma can be a marker of this internal

abnormality (Griffiths *et al.,* 1998). Keratoderma may be inherited (hereditary), or more commonly acquired. In hereditary keratoderma, the condition runs in families and is passed down from one or both parents to their children (Griffiths *et al.,* 1998). In acquired keratoderma, the condition is not inherited and occurs as a result of a change in the health or the environment of the affected person (Griffiths *et al.,* 1998).

The hereditary keratodermas are caused by a gene abnormality that results in abnormal skin protein (keratin). They may be inherited either from one affected parent (Autosomal dominant inheritance) or from both parents who are generally unaffected (Autosomal recessive inheritance). Several family members may be affected. Some types of keratoderma are associated with abnormalities of internal organs. The affected gene has been specifically identified for many of these keratodermas (Griffiths *et al.,* 1998; Stevens and Leigh 1999;Odom *et al.,* 2006).

Diffuse hereditary palmoplantar keratodermas (PPKs) present in early childhood with redness of the palms and soles which gradually become thicker and develop a yellowish waxy appearance. There is a clear cut – off between affected and unaffected skin and the edge of the thickening is often red. This is usually obvious by the age of 3–4 years (Griffiths *et al.,* 1998; Stevens and Leigh 1999;Odom *et al.,* 2006). The non–transgradient diffuse inherited keratodermas are known as Vorner's disease and Unna–Thost disease (Kanitakis *et al.,* 1987; Kuster & Becker ,1992). They are inherited as autosomal dominant conditions. There are no associated abnormalities. Transgradient diffuse PPKs include Olmsted syndrome (AR), Mal-de Meleda (RA) and Huriez syndrome (AD) (Griffiths *et al.,* 1998). Papillon–lefevre syndrome is an autosomal recessive keratoderma associated with inflammation of the gums and loss of teeth, sparse hair and recurrent bactcrial infection of the skin and internal organs (Haneke , 1979; AL – Khenaizan *et al.,* 2002).

Types of focal hereditary PPKs include : ppk striata/areata, hereditary painful callosities, Richner–Hanhart syndrome and pachyo–anychia congcnita (Griffiths *et al.,* 1998). In Howell–Evans syndrome: autosomal dominant inheritance, childhood onset of focal keratoderma, oesophageal cancer in middle age, white areas may be seen inside the mouth (Leukokeratosis) (Howell – Evans *et al.,* 1950).

Acquired keratodermas are keratodermas that are not inherited as a primary genetic condition. They may occur as part of a generalized skin condition (some of which may be inherited) or as a result of another illness(Samanta *et al.,*1976). Acquired ppk is more likely to present in adulthood (compared with inherited keratoderma which usually present in childhood). It presents with thickening of skin of the palms and/or soles which may be diffuse (involving most of the palms and soles) or focal (localized mainly to pressure areas) (Samanta *et al.,*1976). Some causes of acquired keratodermas (Samanta *et al.,*1976; Griffiths *et al.,* 1998; Stevens & Leigh, 1999; Odom *et al.,* 2006).

Inflammatory skin conditions :

Psoriasis, Dermatitis (eczema), Lupus erythematosus, Lichen planus, Erythrokeratoderma .

A. Infections :
   Reiter syndrome, Dermatophyte fungal infection (tinea), Syphilis , Crusted scabies , Extensive viral warts (usually in immuno suppressed patients).
C. Circulatory problems (lymphoedema)
D. Medical ions and toxins :
   Iodine, Lithium, Glucan, Arsenic, Dioxine , Chemotherapeutic agents used in cancer .
E. Internal illness :

Myxoedoma (thyroid disease), internal malignancy (cancer) has been noted with the development of acquired keratoderma.

F. Miscellaneous: Keratoderma climacterum which is a keratoderma that develops usually in middle age women. It has been suggested that this is related to the menopause .

Aim of study

Palmo plantar keratoderma is not an uncommon disease. The aim of this work was to shed some light on causes and clinical presentations of this condition in Iraqi patients at Hilla city.

Patients and methods :

A total of 60 patients with palmo – palnter, palmar or plantar keratoderma were studied as an out patients in Department of dermatology at Merjan teaching hospital during the period from January 2006 to January 2008.

A clinical history was taken from each patient including name, sex, age at presentation, age of onset and duration of disease. Symptoms related to the disease like itching, pain and sweating … Family history of similar condition in relatives of patients was asked about.

Physical examination was performed to determine the site of involvement weather both palms and soles together, palms only or soles only. Also the type of involvement wether diffuse affects most of the palms and soles or focal areas involved. Also determine the extent of involvement weather confined to palms and soles only (non–transgradient) or extends to the dorsum of hands and feet (transgradient). Lesions on other parts of the body and any other associated dermatoses or systemic disease were sought for.

Through history and physical examination, the causes of palmo – plantar keratoderma was determined wether acquired or hereditary.

Results

A total of 60 patients with ppk was seen. Their ages ranged between 5 – 70 years with a mean ± standard deviation (SD) of 24.9 ± 18 years, while their ages of onset averaged from since birth or few month after birth to 68 years with a mean ± SD of 17.9 ± 18.5. The duration of the disease ranged from 1– 32 years with a mean ± SD of 6 ± 7.2 years. There were 40 males (66.6 ٪) and 20 females (33.4 ٪) with a sex ratio of 2 : 1 .

Regarding the age of presentation, the majority of patients were in the first 3 decades of life forming about 75٪ of cases (Figure I). In regarding to the age of onset, half of cases started in the first decade of life (figure II).

Acquired causes of keratodermas were seen in 50 patients (83.3٪) (table 1). Hereditary causes were seen in 10 patients (16.7 ٪) (table 2).

Psoriasis was the commonest cause of acquired ppk (66.6٪). Twenty - six cases of psoriasis presented as diffuse keratoderma **figures (1)&(2)** and 14 cases as focal keratoderma **figures (3) & (4)**. Clinically, psoriatic keratoderma presented either as diffuse thickening, fissuring and hyperkeratosis on palms and soles, or localized fissuring and hyperkeratosis of the nnars and hypothennars of palms, palmar surface of finger tips of hands, heels, insteps of soles and plantar surfaces of big toes and forefeet. Nail involvement was seen in 80٪ of patients with psoriasis. In some patients, psoriasis may involve other parts of body like scalp, elbows, knees , trunk and extremities.

Other causes of acquired ppks were as follows :

Cumulative irritant contact dermatitis (5٪) presented as a diffuse thick skin with fissuring and increase skin markings of palms from chronic contact with soaps,

detergents and water in 2 housewives and one restaurant worker male patient. Chronic paronychia in some fingers was seen in one house wife patient.

Occupational keratoderma was seen in 3 farmers (5٪), two males palmoplantar and plantar and one female patient, palmar, especially when working in a field and walking barefeet, presented as a diffuse thickening with increasing skin markings, hyperpigmentation and fissuring **figure (5)**.

Lichen palnus keratoderma was seen in 2 patients (3.3٪). One male patient presented as a diffuse thickened yellowish verrucous plaques on palms and soles associated with itching, hyperpigmentation on back and abdomen, scarring alopecia of scalp with nail dystrophy of all fingers and toes nails (20 nail dystrophy syndrome). Another female patient presented as a focal localized yellowish thick plague at the center of both palms. Flexor surfaces of wrists involved with violacious itchy papules. The dorsal surface of tongue and buccal mucosa of oral cavity involved with fixed violet patches, lace – like .

A 12 years old female patient had diffuse palmoplantar keratoderma presented as fissured hyperkeratosis on palms and soles extends to sides of soles (sandal–like) due to pityriasis rubra pilaris, classical type. The patient also had generalized symmetrical involvement of trunk by keratotic follicular scaly papules with islands of normal skin within affected area in addition to scaly scalp. There was thickening of fingers nails.

A 25 years old male patient presented with diffuse plantar keratoderma as thick crusted hyperkeratotic lesion on soles (keratoderma blenorrhagicum). This Reiter's syndrome patient also had reactive arthritis with bilateral knee swellings, knee and ankle joints arthralgia, conjunctivitis and urethritis with subungual hyperkeratosis in some fingers nails .

Hereditary causes of ppk included :

Two patient with papillon– Lefevre syndrome, autosomal recessive, (33%). One male patient 33 years old presented with well demarcated psoriasiform diffuse ppk since 3 years of age. The dorsum of hands, elbows and knees were also involved in addition to loss of teeth at 2 years of age. Another patient was 5 years old male presented also with diffuse psoriasiform thickening of palms and soles **figure (6)** and loss of teeth since 2 years of age. Family history was positive in both cases .

Two patients with congenital diffuse non – epidermolytic ppk (Unna–Thost). Autosomal dominant (3.3 ٪) . One female patient 9 years old presented since 3 years of age with diffuse yellowish thickening of palms and soles, bilateral symmetrical. The dorsum of both hands also involved with thick papules and plagues in a cobble – stone pattern over the knucles especially. Other associated features : hyperhidrosis, chronic paronyhia in some nail folds of hands and thickening of nails. Another 22 years old male patient presented since 1 year of age with diffuse thickened yellowish skin on palms and soles with linear extension over dorsum of hands to cubital area and over dorsum of feet.

A 9 years old female patient with X – linked ichthyosis, carrier, presented since birth with a diffuse fissuring and hyperkeratosis of palms and soles with fish – like scales all over the body, trunk and extremities. Another 23 years old male patient with lamellar ichthyosis, autosomal recessive, presented since birth with diffuse fissured hyperkeratosis in palms and soles **figure (7)** with large fish scales on the trunk and limbs associated with ectropion of both lower eyelids. A 12 years old female patient with Ichlthyosis vulgaris, autosomal dominant, presented since 3 months of age with diffuse dry thickened fissured palms and soles. The dorsum of hands and feet were also involved. There was fish – like scales on the extensor surfaces of legs and upper limbs sparing the flexures.

An 18 years old female patient presented since birth with a localized warty – like thickening of palms and soles with involvement of dorsum of hands and feet in a linear fashion (Striate keratoderma , autosomal dominant) **Figure (8)**. It is a part of epidermal nevus syndrome, there was similar lesions in abdomen around umbilicus in transverse manner. Other involved areas were flexures of elbows , knees, axillae , inguinal regions and around the neck. Also this patient had swelling in left side of the neck below the mandible. A biops from this swelling was shown a rhabdomy osarcoma. Also left optic never involvement (proptosis) with left eye blindness.

A 13 years old female patient presented since one year of age with diffuse fissured psoriasiform thickening of palms and soles associated with hypotrichosis, cheilitis and dystrophy of some fingers nails. This is due to a primary zinc defiency (Acrodermatitis enteropathica, autosomal recessive).

Discussion

Palmoplantar keratoderma is one of the common disorders of keratinzation. It is characterized by focal thickening of the stratum corneum of palms and soles. It can be caused by a host of disorders, hereditary or acquired. It may be the only change or a part of a more wide spread manifestation of an internal malady [11].

The present work represents the first study that deals with this subject in Iraq. When the 60 cases were studied for the incidence in different age and sex groups, ppk was found to be more common is males (66.6٪) with a male to female ratio of about 2 : 1. The highest incidence was between the ages of 20–30 years (25٪). In 28 patients (46.6٪), the disease started between the age of 0–10 years. These findings were nearly comparable to what had been reported in an Indian study of 82 patients with ppk in which the incidence was more in males (64.6٪). The highest number of cases was between 15–30 years (32.9٪) and in 40 patients (48.7٪), the disease started between the ages of 0 – 15 years [12].

Psoriasis was the most common of acquired causes of ppk in this study (66.6٪). Papillon–Lefevre syndrome and congenital diffuse non–epidermolytic ppk (Unna–Thost) were the main causes of hereditary ppk (3.3٪) for each. This contradicts to findings in Indian study in which Unna–Thost was the main cause of hereditary ppk (28.05٪) whereas psoriasis among acquired causes formed about (17.07 ٪) of cases(Mahajan *et al.,* 1994).

In order to determine the causes of ppk, it is important to examine other parts of the body other than palms and soles like nails, scalp, elbows, knees, especially in psoriasis. Teeth examination is important in cases of hereditary Papillon–Lefevre syndrome in which there is loss of teeth and inflammation of gums of earlier age about 2 or 3 years in our patients, especially those presented with psoriasiform thickening of palms and soles. So teeth involvement can differentiate this syndrome from psoriasis hence ppk clinically was the same in both conditions and knees and elbows which are the sites of predilection in psoriasis can be involved in patients with Papillon–Lefevre syndrome .

From a hereditary causes, Striate keratoderma, a focal ppk as a part of epidermal nevus syndrome, to the best of our knowledge, was the first reported case that show an association of epidermal nevus with rhabdomyosarcoma of the neck and eye proptosis (AL–Alawachi *et al.,* 2006).

No previous study had evaluated ppk in our country, so this report was a first step towards shedding light on this subject .

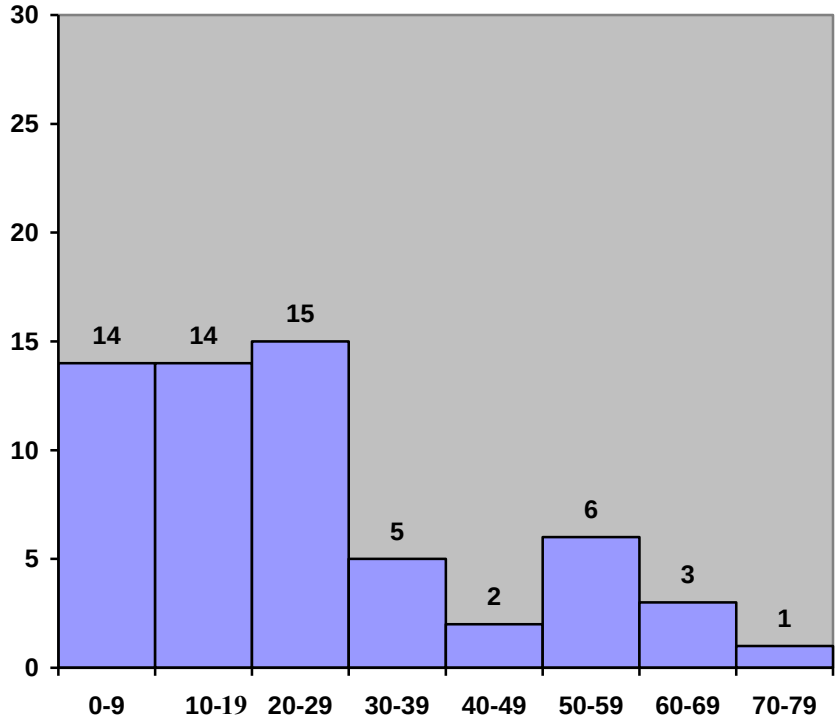

Figure I : Age of presentation for patients with ppks :

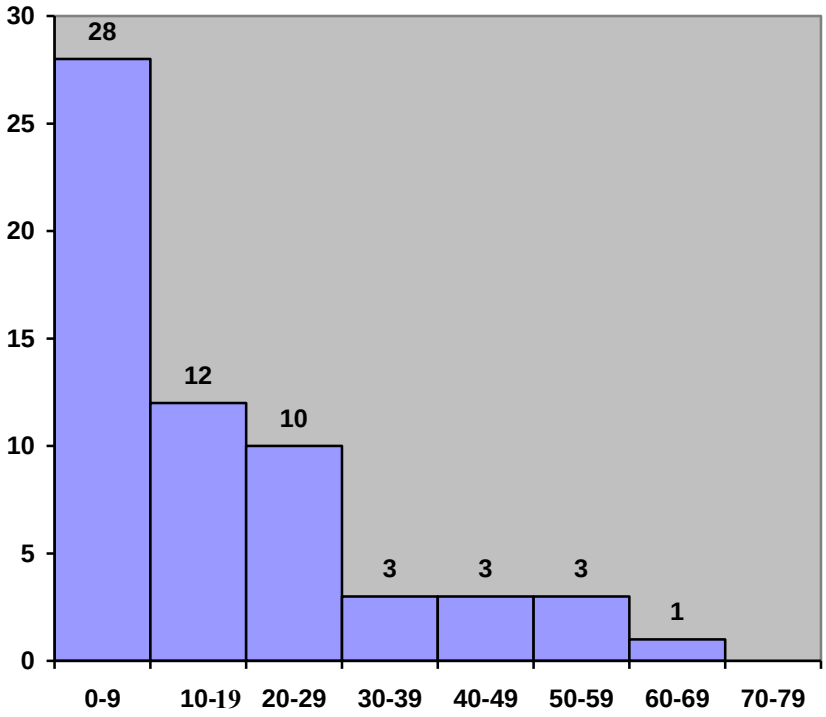

Figure II : Age of onset for patients with ppks :

Table (1) : Acquired causes of palmoplantar keratoderma :

Acquired cause of ppk	No. of cases	Percentage
Psoriasis	40	66.6 ٪
Cumulative irritant contact dermatitis	3	5 ٪
Occupational	3	5 ٪
Lichen planus	2	3.3 ٪
Pityriasis rubra pilaris	1	1.6 ٪
Reiter syndrome (keratoderma belnorrhagicum)	1	1.6 ٪

Table (2): Hereditary causes of palmoplantar keratoderma:

Hereditary cause of ppk	No. of cases	Percentage
Papillon – Lefevre syndrome (Autosormal R.)	2	3.3 ٪
Congenital diffuse non – epidermolytic ppk (Unna – Thost , Autosormal D.)	2	3.3 ٪
X – liked Ichthyosis (x – liked R.)	1	1.6 ٪
Lamellar Ichthyosis (AR)	1	1.6 ٪
Ichthyosis vulgaris (AD)	1	1.6 ٪
Striate keratoderma (AD)	1	1.6 ٪
Acrodermatitis enteropathica (primary zinc defiency, AR)	1	1.6 ٪
Palmoplantar keratoderma areata (AD)	1	1.6 ٪

Table (3) : Sites of palmo plantar keratoderms :

Site of keratoderma	No. of cases	Percentage
Palmo – plantar	46	76.6 ٪
Palmar	4	6.8 ٪
Plantar	10	16.6 ٪

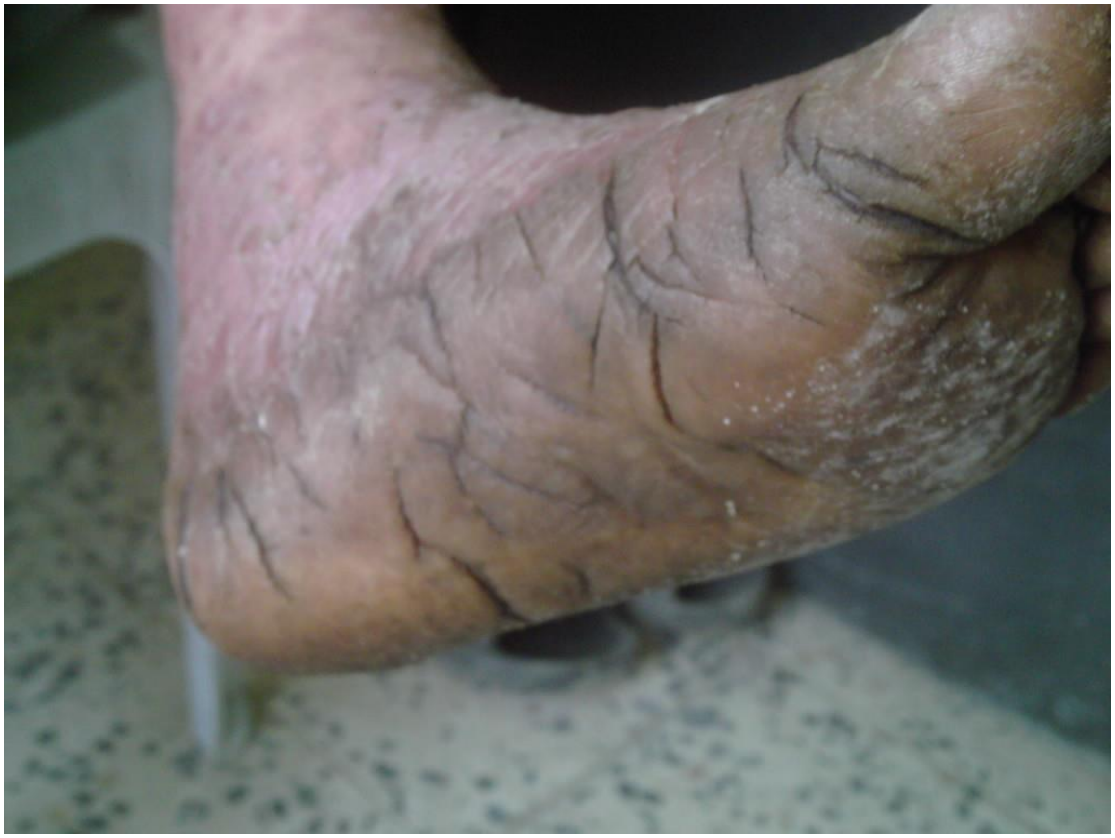

Figure (1): Diffuse plantar keratoderma with fissuring (psoriasis)

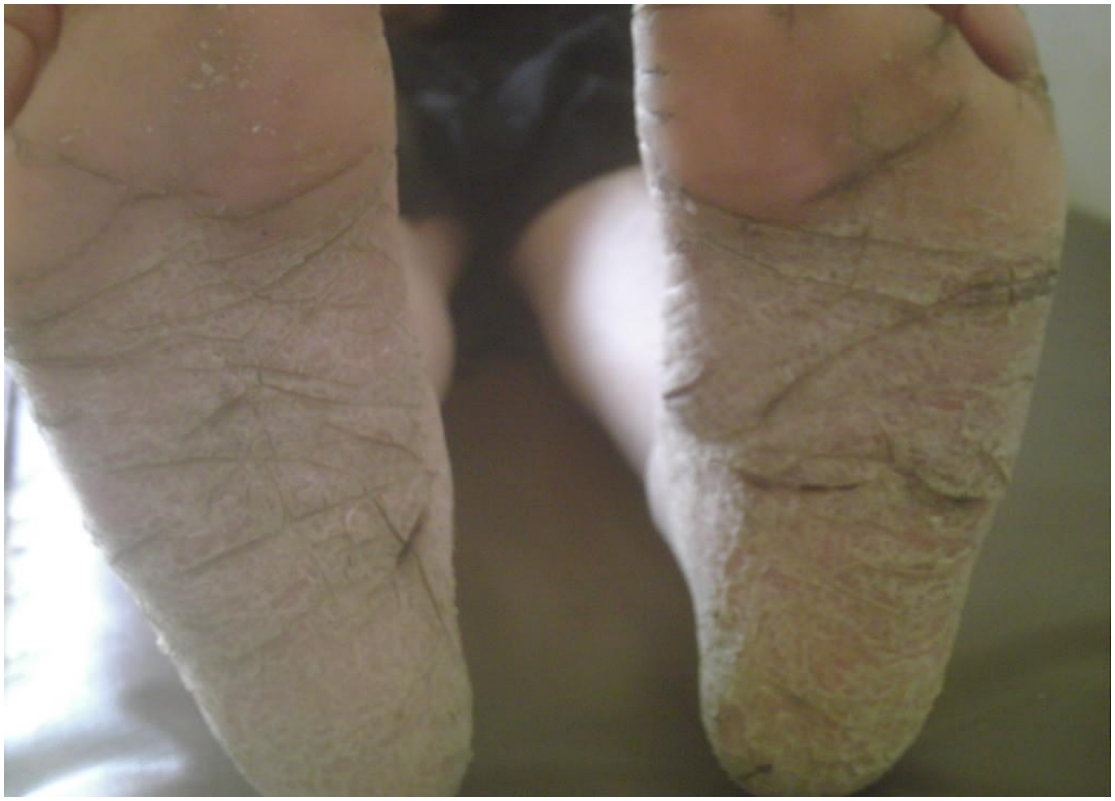

Figure (2): Diffuse plantar keratoderma (psoriasis)

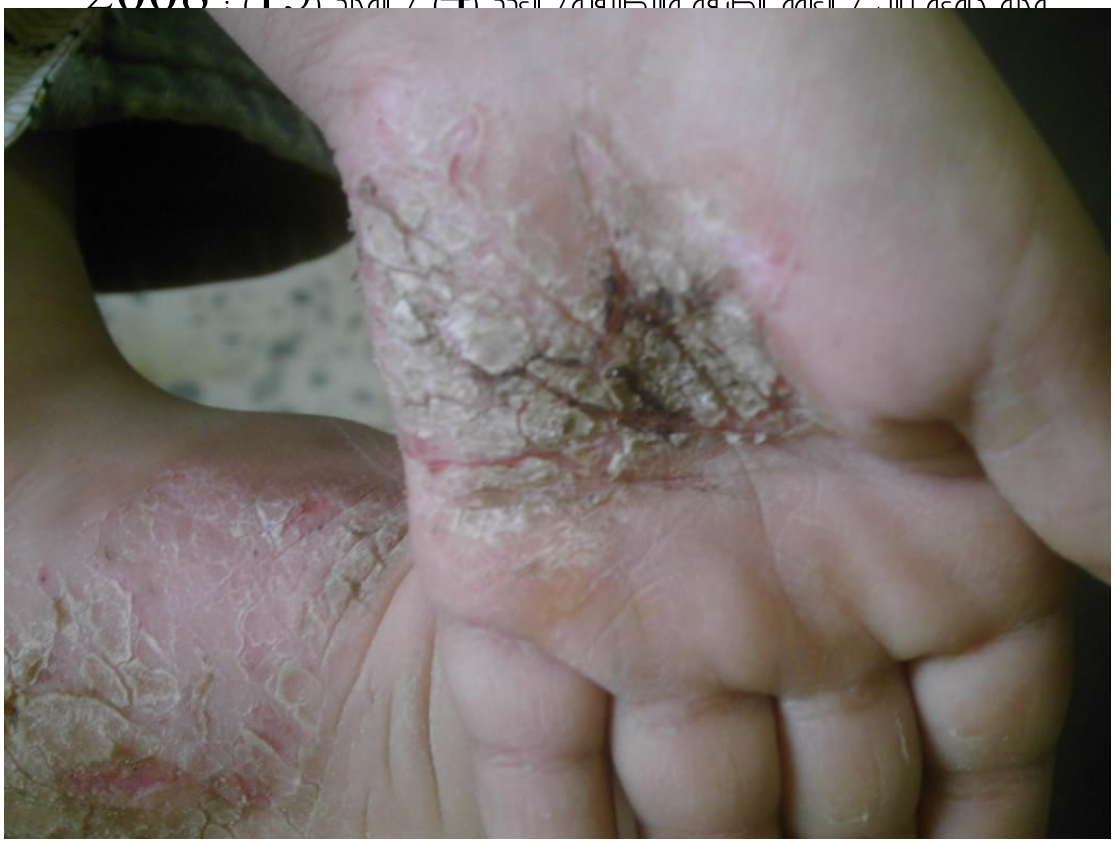

Figure (3): Focal palmar keratoderma (psoriasis)

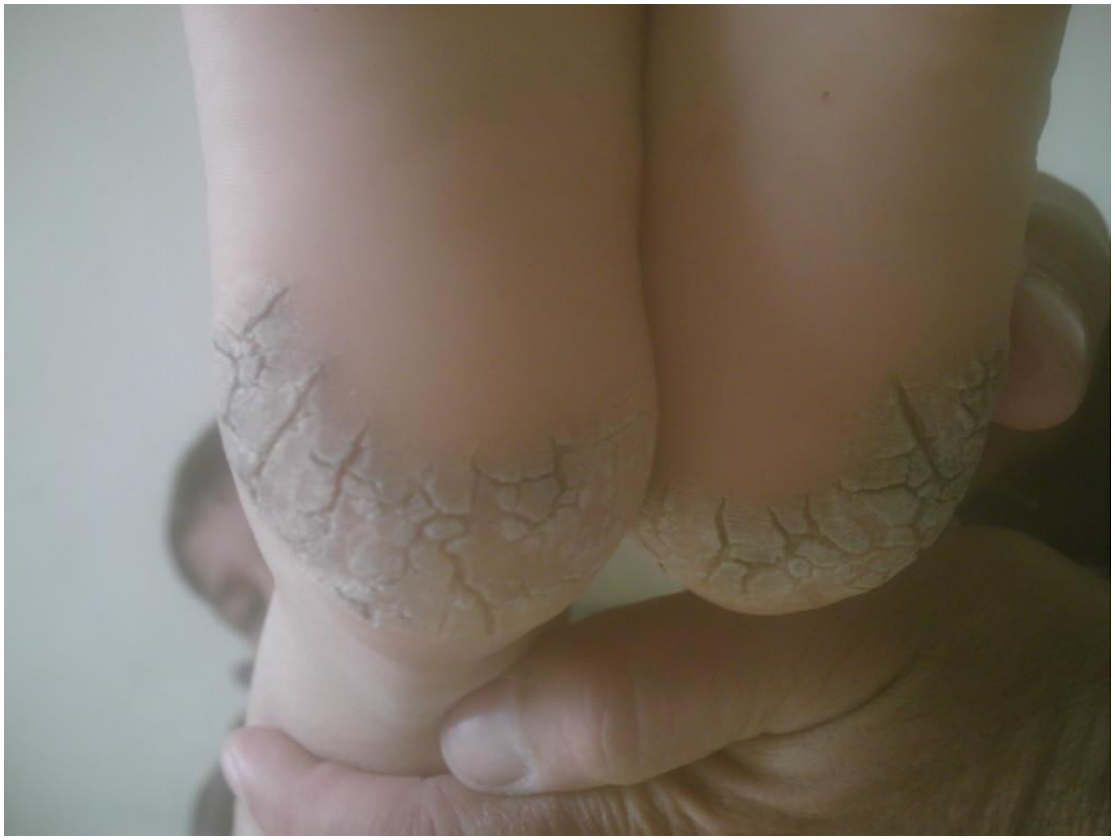

Figure (4): Focal plantar keratoderma of heels with fissuring (psoriasis)

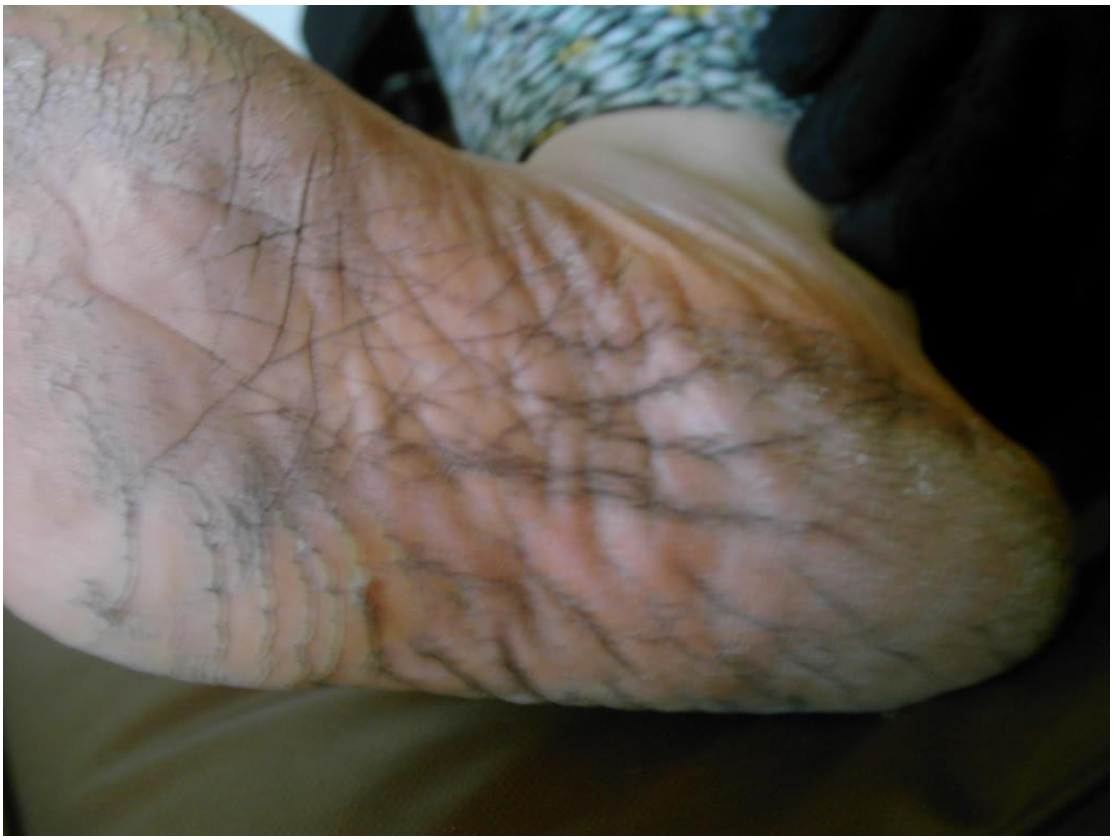

Figure (5): Diffuse plantar keratoderma (occupational)

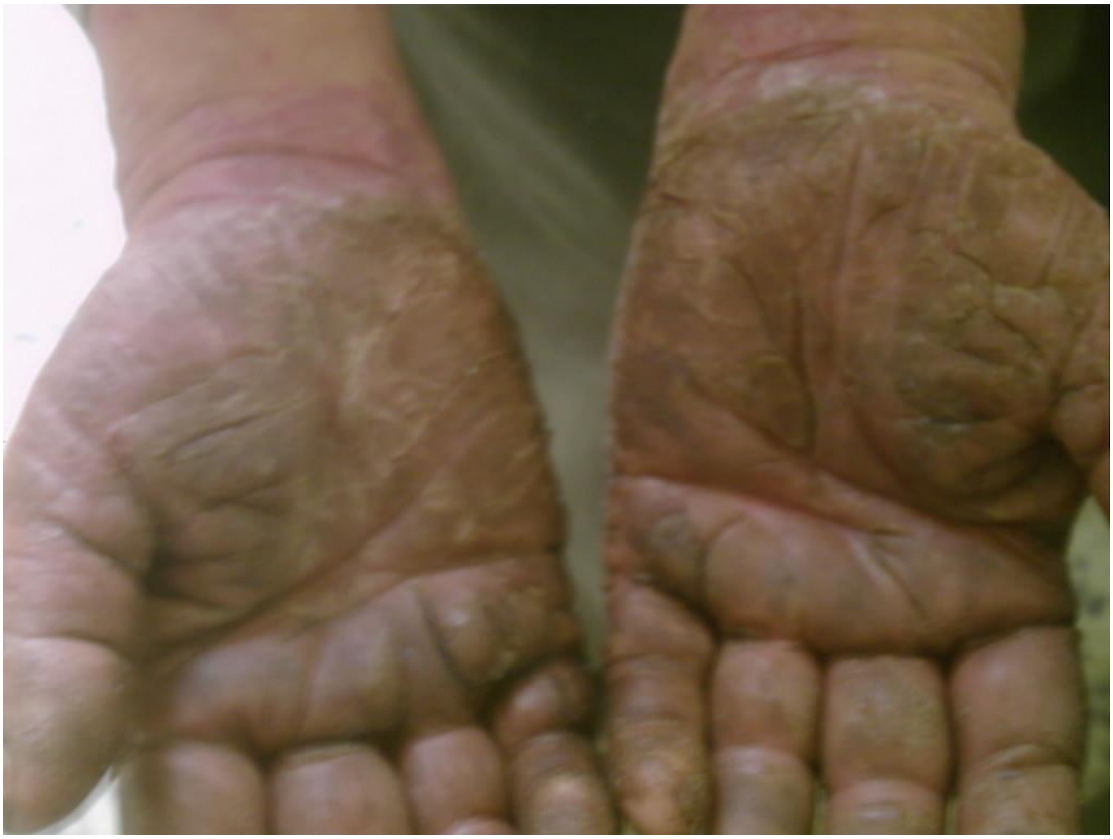

Figure (6): Diffuse palmar keratoderma (papillon – Lefevre syndrome)

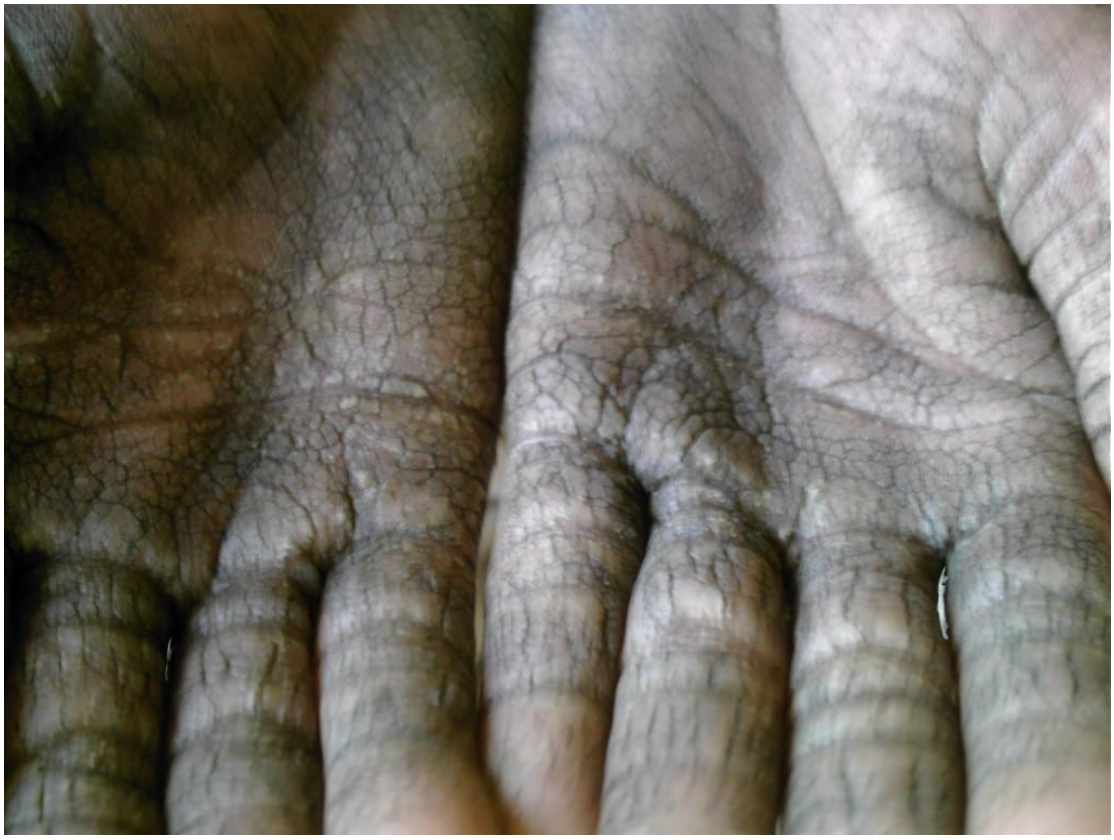

Figure (7): Diffuse palmar keratodermas (lamellar ichthyosis)

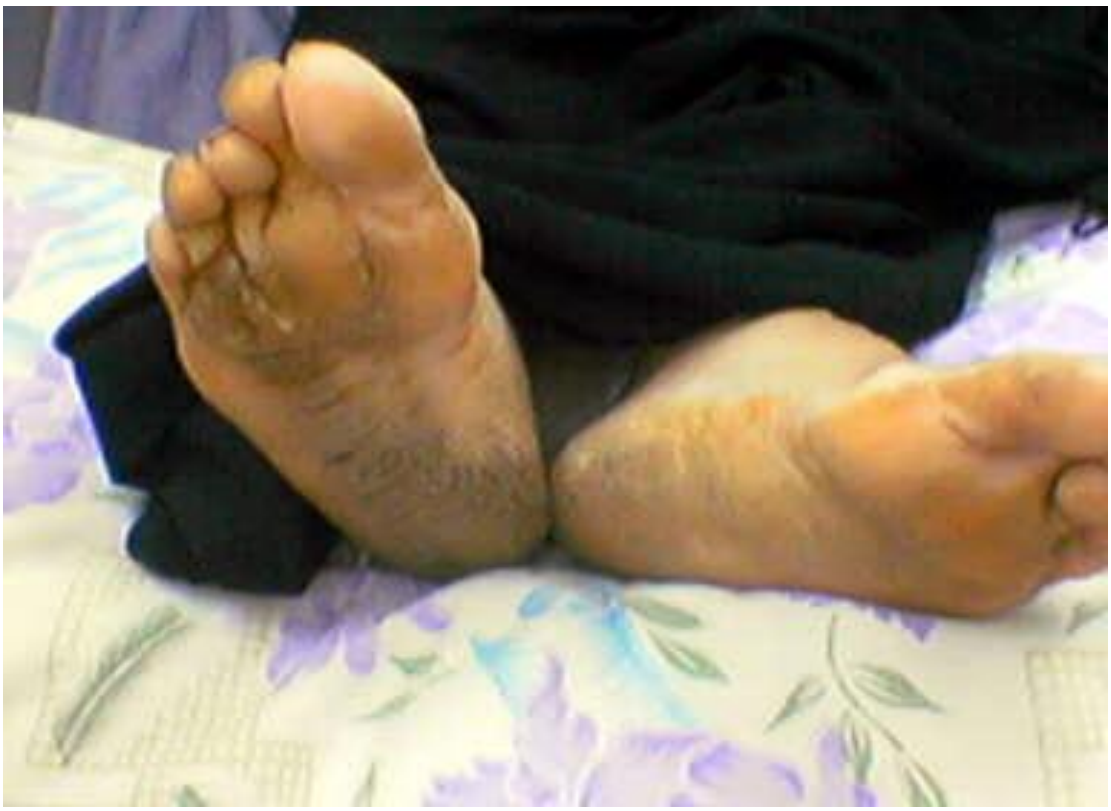

Figure (8): Focal linear striate plantar keratoderma

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