Journal of Clinical & Experimental Dermatology Research

Omar et al., J Clin Exp Dermatol Res 2015, 6:4
http://dx.doi.org/10.4172/2155-9554.10000287

Case Report Open Access

# Familial Psoriasis: Report of Three Generations Affected and Literature Review

**Graciano-Machuca Omar[1], Salazar-Torres Francisco Javier[2], Zepeda-Nuño Sergio[3], Villanueva-Quintero Delfina Guadalupe[2], Alvarado-Navarro Anabell[4], Ramírez-Dueñas María Guadalupe[1], Sánchez-Hernández Pedro Ernesto[1*]**

[1]*Immunology Laboratory, Department of Physiology, University Center for Health Sciences, University of Guadalajara, Guadalajara, Jalisco, México*

[2]*Dermatological Institute of Jalisco "Dr. José Barba Rubio"*

[3]*Institute of Dental Research, University Center for Health Sciences, University of Guadalajara, Guadalajara, Jalisco, México*

[4]*Research Center for Immunology and Dermatology, Department of Physiology, University Center for Health Sciences, University of Guadalajara, Guadalajara, Jalisco, México*

[*]**Corresponding author:** Pedro Ernesto Sanchez Hernandez, Immunology Laboratory, Department of Physiology, University Center for Health Sciences, University of Guadalajara, Guadalajara, Jalisco, México, Tel: +523310585200; E-mail: pedro.e.sanchez@gmail.com

**Received date:** March 28, 2015, **Accepted date:** June 22, 2015, **Published date:** June 30, 2015



**Abstract**

Psoriasis vulgaris (PsV) is a chronic inflammatory skin disease associated to genetic and environmental factors. We report two cases of a family with three generations affected with this pathology. The *propositus* (a three-year-old boy), his father and his grand-father present confluent erythematous plaques with scales and their diagnostic of PsV was made based on the clinical characteristics and supported by histopathological study. More than 50% of the body is covered by lesions. The *propositus* received treatment as hydrocortisone and emollients reflected a slight positive response; however his father has had multiple treatments without significant improvement.

**Keywords:** Psoriasis vulgaris; Plaques

## Introduction

Psoriasis vulgaris is a chronic and inflammatory skin disease that affects 3% of world population. The prevalence rate for pediatric psoriasis is less than 1% [1-3]. The pathogenesis is complex and depends on environmental, genetic and autoimmune factors. Lesions are the result of abnormal epidermal proliferation, vascular changes and immunological disturbances in combination with genetic predisposition [4].

The pathology presents a wide clinical variety and evolution, the most common form of psoriasis is psoriasis vulgaris, which is characterized by well defined patches of red raised skin that can appear on any area of skin, although the knees, elbows, scalp, trunk and nails are the most common locations [4].

### Onset psoriasis

Psoriasis is classified into two types according to the age of onset the initial presentation (at psoriasis onset):

Type I, early age of onset form, the diseases´ onset is before the 40 years of age: It is correlated with HLA-Cw6, -B13 and –B57. The clinical course is severe, and often the patient has a family background affected by psoriasis [2,5].

Type II, late age of onset form is after 40 years old: It is associated with HLA-Cw2 and –B27 [2].

### Genetics of psoriasis

36 susceptibility *loci* have been identified as a result of Genome-wide association studies (GWAS) and a meta-analysis of GWAS of psoriasis in European population [6].

The major psoriasis susceptibility gene named PSORS1 is located in the MHC region, and the most strongly associated allele is *HLA-Cw*0602* [5,7].

A non-Mendelian mode of inheritance has been proposed for this disease, although according to a variety of pedigree and population surveys, different genetic segregation models have been considered for psoriasis. Only a few cases could be explained through them, so, a multifactorial model cannot be rule out [5,7].

One third of psoriatic patients have an affected first-degree relative. Studies in monozygotic twins show a concordance of psoriasis ranging from 35% to 73% [7].

When psoriasis does not affect parents or sibling, the risk is around 2%, but on the other hand, if both parents were affected there is a is a threefold increased risk of psoriasis in 41%, by the other side if one of them were affected the risk would be around can be as low as 14% and just a 6% when a sibling is affected [7].

## Case Reports

### Case III 13

The *propositus* is a three-year-old boy who was born in Guadalajara, Mexico. Patient was presented at Instituto Dermatológico de Jalisco "Dr José Barba Rubio" dermatology clinic with a symmetric and generalized dermatosis. Physical examination showed mostly confluent erythematous plaques with scales. A 70% of body surface area (BSA) is covered by lesions, affecting head, arms, trunk and legs (Figure 1). No dystrophic or abnormal nail changes were observed. Moreover, propositus' parents mention presence of occasional irritability with pruritus on the patient.

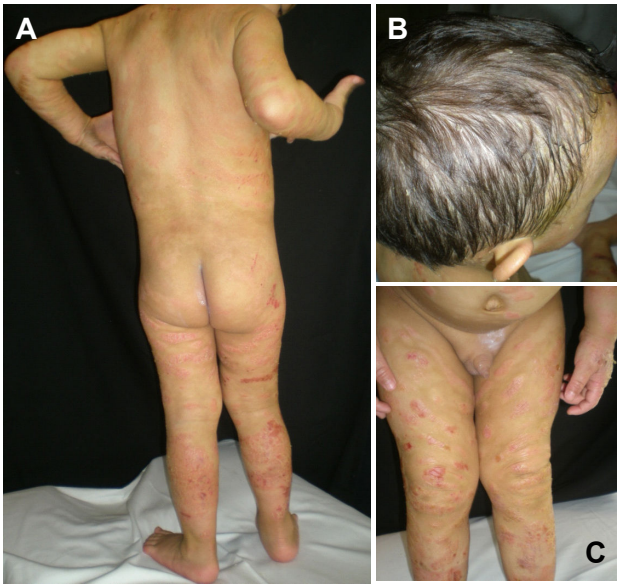


**Figure 1: Erythematous plaques covered 70% of body surface area of index case (III 13). A)** Posterior region of trunk and lower extremities; **B)** Erythematous scaly patches on the scalp; **C)** Anterior legs and genital area.

The onset of the disease in the purpose was 4 months old with erythematous macules on the trunk, which subsequently developed scales. Lesions gradually spread to form plates of different diameters. Fathers denied prior infection to the appearance of lesions; however, he had family history of psoriasis: father (II 11), grand-father (I-1), and two uncles (II 1 and II 5) (Figure 2).

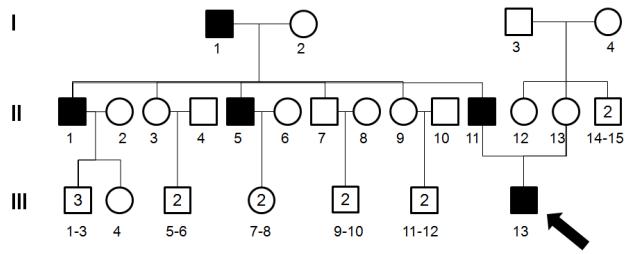


**Figure 2: Three generation pedigree of a Mexican Mestizo family affected by PsV.** Subjects I 1, II 1, II 11 and III 13 were diagnosed and treated for PsV. Subject II-5 has presented some sporadic plaques however has not been diagnosed. Lesions' age-onset referred in their medical records of individuals: I 1, II 1, II 5, II 11, III 13 were 44, 41, 33, 4 years old and 4 months, respectively.

A histopathological study was requested. The results of skin biopsy were positively associated with psoriasis (Figure 3a). According to clinical characteristics, histopathological study and family history of psoriasis, the child was diagnosed with PsV. The patient received hydrocortisone and emollients treatment with a slight positive response.

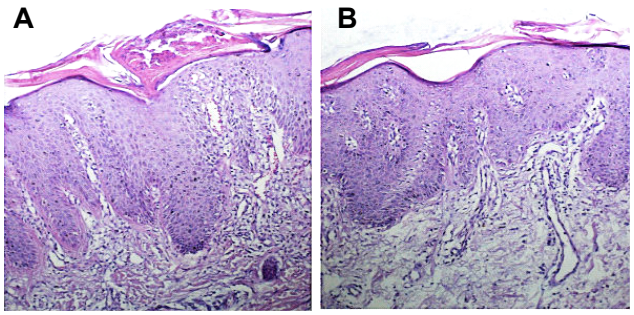


**Figure 3: Biopsy specimen from plaques of the purpose (case 1, A), and purpose's father (case 2, B).** Both Photomicrographs showing an epidermal hyperplasia with elongation of the rete ridges in a regular pattern, loss of granular cell layer, and overlying parakeratosis. Also present are blood vessels dilated and tourtuous, perivascular lymphocytic infiltrate, edema in the superficial dermis and presence of lymphocytes into the epidermis. Further, figure a presents intracorneal microabscesses and similar neutrophilic pustules in the spinosum stratum. In both cases findings are highly consistent with psoriasis. (Hematoxylin and eosin stain, original magnification x10).

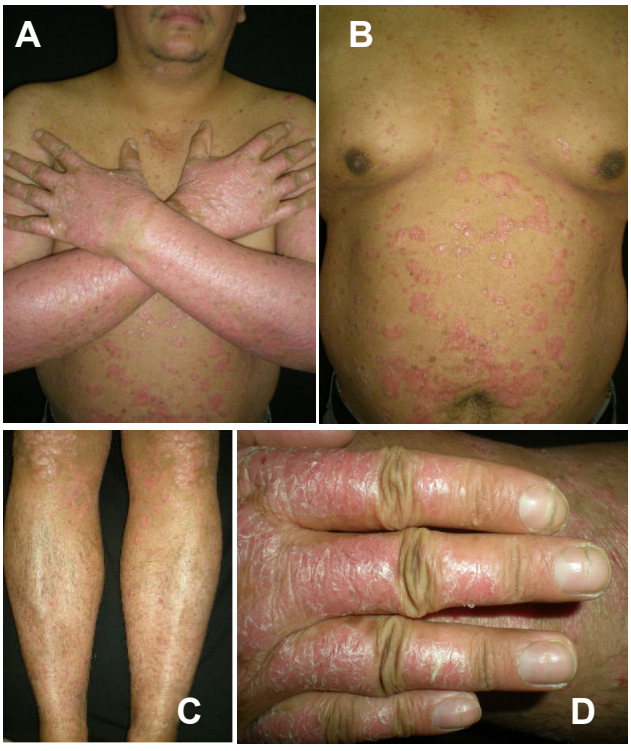


**Figure 4: Psoriatic lesions are present in 60% of the body surface area of case 2 (II 11). A)** Forearm with palmar region; **B)** anterior chest and abdominal region; **C)** previous lower extremity D) bilateral digital.

### Case II 11

The *propositus'* father, 36 years old, is a native of Los Mochis, Mexico. He works as an engineer. On physical examination he had the same morphology and topography of lesions that were observed in *propositus*. He referred that the onset of symptoms was at 4 years old. Plaques cover a 60% BSA. Further, he presents Beau lines on nails suggesting a previous period of exacerbation of the disease or some stronger event that impact on the nail process (Figure 4). Unlike his son, he mentions that his injuries were asymptomatic. Clinical characteristics were consistent with PsV, and histopathological result served as a confirmatory study to diagnose this pathology (Figure 3b).

The patient has been involved on multiple treatments including methotrexate, topical steroids and emollients without significant improvement.

### Case I 1

The *propositus*' grand-father, 69 years old, is a native of Los Mochis, Mexico. He works as farmer. Physical examination showed confluent erythematous plaques with few scales. A 55% of BSA is covered by lesions predominantly on head, arms, and legs, and scantily on trunk. Previously, he had been diagnosed with psoriasis with clinical and histopathological evidence obtained from other health institution. He mentioned that the onset age of the lesions was at 44 years old with recurrence of exacerbation periods.

## Discussion

This report shows a family that has three consecutive generations affected by PsV. The subjects I 1, II 11 and III 13 had a severe manifestation of the disease because they have more than 50% of the body surface covered by plaques. Subject II 1 has a mild PsV affecting mainly elbows and lumbar region, with periods of disease clearance. Subject II 5 reported having presented some erythematous plaques on the elbows, however never has been diagnosed or treated.

Environmental (infections, stress, traumas) and genetic (PSORS genes and different single nucleotide polymorphisms (SNPs) factors have been postulated to trigger and aggravate PsV [2,4]. This family clearly has an advance on the age of the onset of the disease; this fact could suggest the involvement of a specific genetic event, something similar to the event calling "anticipation", speaking of the tendency in certain genetic disorders for individuals in successive generations to present at an early-onset and / or with more severe manifestations [8].

Males and females are equally affected by psoriasis [7]. However, previous research suggests there is an increased risk for PsV in the patient when he has an affected father rather than an affected mother [9]. Despite the phenomenon of anticipation in psoriasis seems to be a relevant condition, also affected family members are only from male gender. They are events that need more research to deepen and understand heritability of psoriasis.

## Conclusion

Because of the complex network of genetic and environmental factors involved in the development of this disease is difficult to create a genetic test for diagnosis. In this report, the family history of psoriasis facilitated the diagnosis in the index case and it was supporting by physical examination and histopathology. A correct diagnosis and an appropriate treatment are necessary for patients to continue with a good quality of life.

## References

1. Busch AL, Landau JM, Moody MN, Goldberg LH (2012) Pediatric psoriasis. Skin Therapy Lett 17: 5-7.
2. Gupta R, Debbaneh MG2, Liao W1 (2014) Genetic Epidemiology of Psoriasis. Curr Dermatol Rep 3: 61-78.
3. Mahé E, Gnossike P2, Sigal ML2 (2014) [Childhood psoriasis]. Arch Pediatr 21: 778-786.
4. Ammar M, Souissi-Bouchlaka C2, Gati A2, Zaraa I3, Bouhaha R2, et al. (2014) [Psoriasis: physiopathology and immunogenetics]. Pathol Biol (Paris) 62: 10-23.
5. Chandra A, Ray A, Senapati S2, Chatterjee R3 (2015) Genetic and epigenetic basis of psoriasis pathogenesis. Mol Immunol 64: 313-323.
6. Tsoi LC, Spain SL, Knight J, Ellinghaus E, Stuart PE, et al. (2012) Identification of 15 new psoriasis susceptibility loci highlights the role of innate immunity. Nat Genet 44: 1341-1348.
7. Gudjonsson JE, Elder JT (2007) Psoriasis: epidemiology. Clin Dermatol 25: 535-546.
8. Friedman JE1 (2011) Anticipation in hereditary disease: the history of a biomedical concept. Hum Genet 130: 705-714.
9. Burden AD, Javed S, Bailey M, Hodgins M, Connor M, et al. (1998) Genetics of psoriasis: paternal inheritance and a locus on chromosome 6p. J Invest Dermatol 110: 958-960.