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Actinic Prurigo, a Disease Seen in Indigenous Population of the South American Mountains

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Abstract

Actinic Prurigo (AP) is an uncommon photodermatosis affecting in the majority of the cases, natives and half-breeds of South and Central America. AP represents an abnormal response to ultraviolet A and B, and it is associated with the Human Leukocyte Antigen system HLA-DRA (DRB1 0407 Y DRB1 0401). Multiple epidemiological, clinical geographical, and immunological factors are involved in the etiology of this entity. Its treatment represents a major challenge. Most frequently found in the female population starting during the first decade of life, compromising symmetrically sun-exposed skin areas, as well as the submucosa of the lips and the conjunctiva. Clinically, it presents with erythematous papules, nodules, lichenification, hyper or hypo post-inflammatory pigmentation and scarring, accompanied by an intense pruritus, deeply affecting the quality of life of the patient as we will present after working at 3,400 mts in the Andes Mountains in Argentina. The diagnosis is made primarily by the clinical aspect and both personal and family history of the affected patient. We attend a health center at 3,400 mts high in the Andes Mountains with indigenous patients affected by AP.

Keywords: Actinic prurigo; Prurigo; Solar prurigo; Actinic

Introduction

AP is a rare photodermatosis of chronic evolution that occurs in the first decade of life.[1] It is characterized by recurrent outbreaks of papules and nodules, localized mainly at the photo exposed areas. The process is benign, but it is accompanied by intense itching and excoriation which has a profound impact on the activities of daily life, social relationships and psychological status of the patient. Its pathogenesis is unclear and its treatment represents great challenges.

Case Presentation

Actinic prurigo is a very infrequent entity in the world population, with a female predominance (3:1.4 ratio), observed more frequently in certain populations of Central and South America, where there is evidence of an abnormal response to Ultraviolet Radiation (UV).[1] Several patients who live in the North of Argentina, in the Andes Mountains at 3,400 mts high, chronically exposed to sunlight, were evaluated by dermatologists of our

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institution during this year. Many of them presented clinical lesions of actinic prurigo, such as lichenification in photo exposed areas, erosions and hyperpigmented scars. It causes persistent and symmetric lesions in the skin, in the labial semi mucosa and at the ocular level. Skin lesions are varied: papules, nodules, xerosis, erosions and lichenification in figure exposed areas (Figure 1 and 2). Eventually pigmentation disorders (hypo and hyperpigmentation) and scars are observed (Figure 3). It is accompanied by intense itching and excoriation. Vesicles or blisters are only observed if there is a superinfection. The lesions settle on the face, the neckline area, the extensor areas of the upper limbs, the back of the hands, the nape of the neck and the lateral aspects of the neck, and less frequently in the buttocks or trunk. Residual lesions are post-inflammatory hypo or hyperpigmentation and scars (Figure 4) Erosions,[2-4] fissures and edema are frequently observed in lesions mainly the lower lip (Figure 5) face and earlobes (Figure 6).[5] In an Australian study led by Crouch in 2002, cheilitis was seen in only 24% of patients, compared to natives of Central and South America which is 65%.[6,7] At the ocular conjunctiva, it can be found hyperemia, photophobia and tearing in 45% of patients with AP. In severe cases, it can lead to hypertrophy of the papillae and the formation of a pseudopterygium.[2,3]

AP lesions occur at times of the year with increased sun exposure –spring and summer, in areas at altitudes greater than 1,000 m, such as in the Andes Mountains.[8] In Canadian Native Americans the prevalence is 0.1%; and in mestizo populations in Mexico, is 1.3% to 3.5%. In Argentina it is observed more frequently in the high-altitude regions of the north and northwest of the country, areas with decreased oxygen, low humidity (14% to 20%), wind with high speed and extreme daily temperatures, three times more exposure to UV rays comparing with populations at sea level.[9,10]

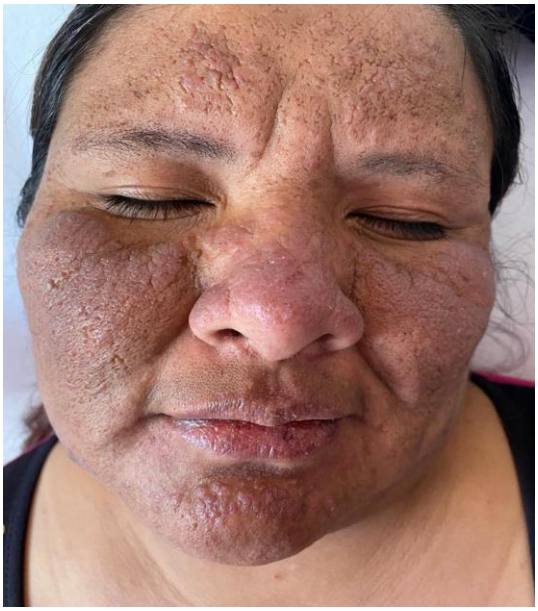

Figure 1: Lichenification on the face (compromising the front, nose, both cheeks and chin).

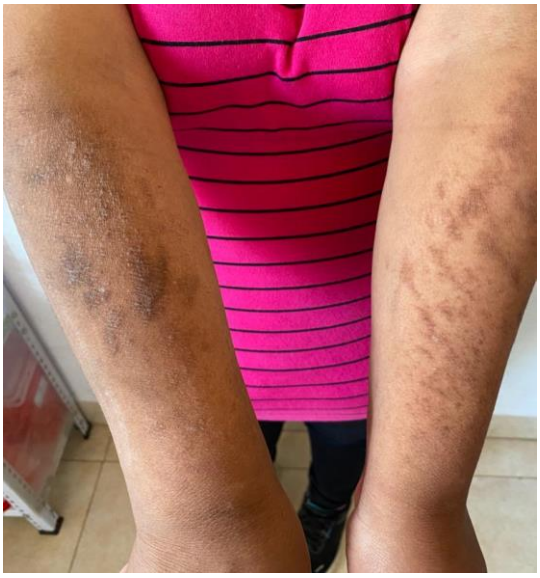

Figure 2: Lichenification on the forearms.

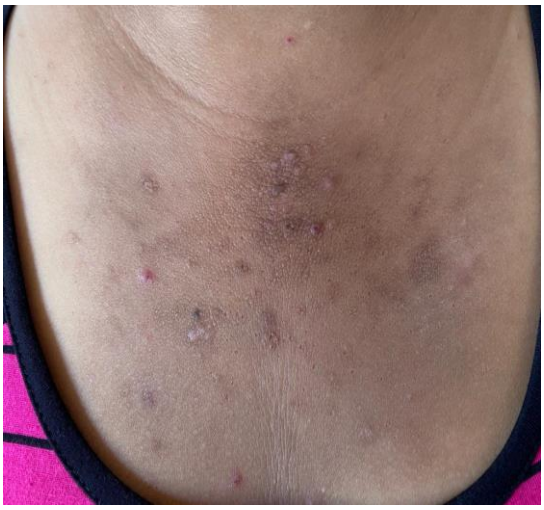

Figure 3: Papules, hyperpigmented lesions and scars on the chest.

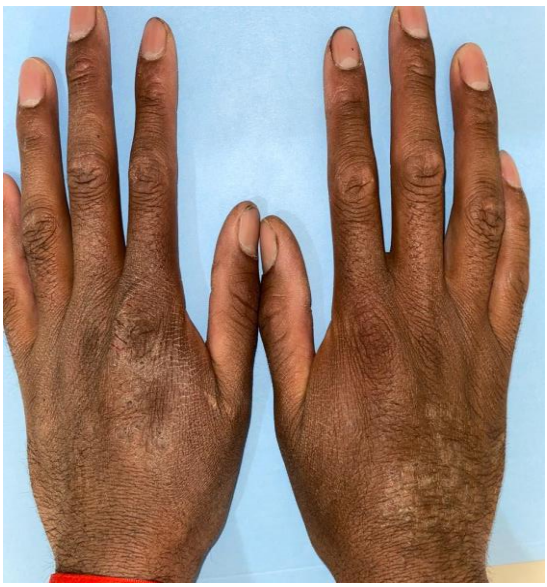

Figure 4: Lichenification and hyperpigmentation on the back of both hands.

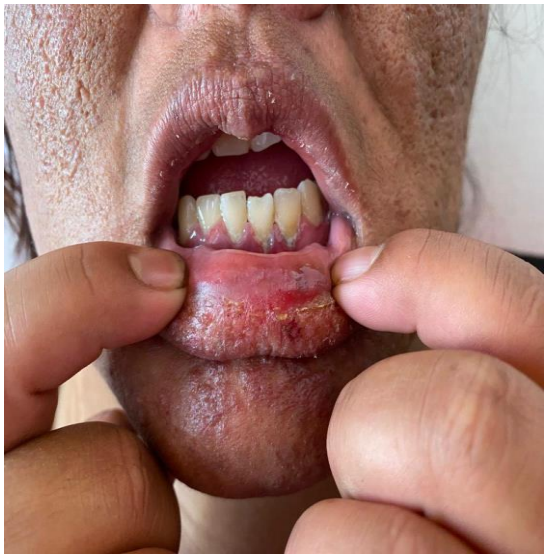

Figure 5: Cheilitis and erosions in the lower lip.

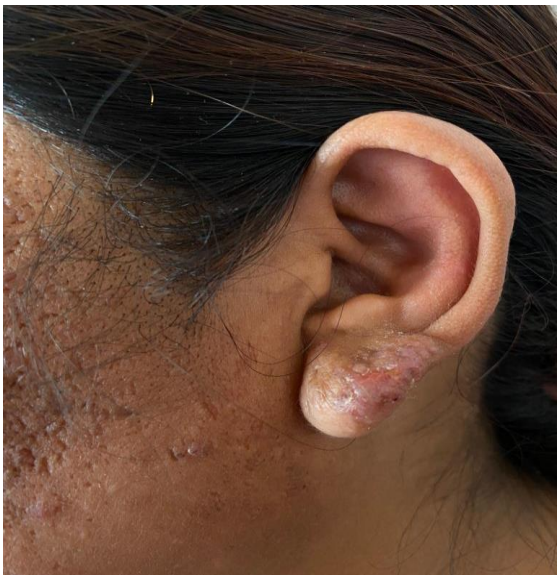

Figure 6: Erosions on the earlobe.

The usual age of onset is 10 years, although it can appear between 2 and 43 years. In native populations of South and Central America, the age may be even earlier (5 years) than in other areas (Figure 7). Spontaneous remission occurs in adolescence or early adulthood, but in certain patients it persists into late adulthood.[11,12] Particularly in Thailand, several cases of late onset have been reported and the average age of onset was 37 years old, with a male predominance (ratio 1.5:1).[13] Late-onset, has less florid clinical features, is more persistent and recalcitrant to treatment response than usual earlier onset.[14]

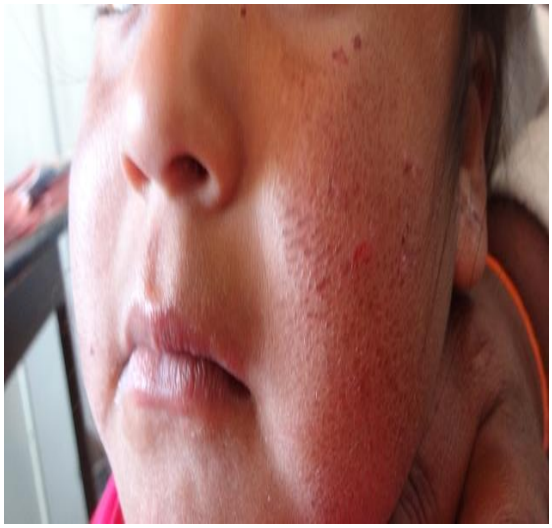

Figure 7: Xerosis, erosions and lichenification on the cheek.

AP may be hereditary, autosomal dominant mode of transmission as in cases of natives from America, or sporadic, as in non-native populations in Europe or Asia.[15]

Other photo dermatoses are; solar urticaria, solar hydroa polymorpha, acute actinic dermatitis, solar dermatosis, phototoxic dermatitis and photoallergic dermatitis. Some authors consider actinic prurigo as a variant of polymorphous actinic eruption (polymorphic light eruption PLE) as both entities have inflammatory characteristics, mediated by an abnormal T-cell immune response.[16,17]

AP is caused by the electromagnetic irradiation of Ultraviolet (UV) radiation, mainly UVA (315 nm to 400 nm) and to a lesser extent by UVB (280 nm to 315 nm). Photo provocation tests are positive in 2/3 of these individuals with the rapid appearance of typical PA lesions. The Minimal Erythema Dose (MED) is decreased in 60% of these patients with UVA and UVA-UVB radiation, demonstrating an abnormal response to UV rays caused by an alteration in the functioning of the immune system involving cellular and humoral immunity.[18-20] There is a delayed hypersensitivity reaction to autoantigens induced by ultraviolet rays in genetically susceptible people.[21] Langerhans cells, which normally decrease in number in the skin when they come into contact with UV, would be found to be increased in patients with AP and are responsible for the antigenic presentation of those antigens induced by UV, generating a greater inflammation.[22,23] It has been proposed that PA is an autoimmune disease, whose putative antigen would be a protein of the epidermis, which is transformed when it is irradiated by UVA rays.[4]

Studies led by Crouch, demonstrated a strong relationship between AP and HLA subtypes, and DRB1-0407 is the most frequently found in people with actinic prurigo (60% to 70% in AP *vs.* 4% in healthy controls), followed by DRB1-0401 (20% positive in PA).[24] There is an increase in Tumor Necrosis Factor alpha (TNF-α) after sun exposure in this population.[24]

Certain particularities are observed in the histopathology that must be taken into account, such as hyperkeratosis, parakeratosis, acanthosis and superficial and perivascular lymphocytic infiltrates. In addition, spongiosis, epidermal ulceration, dilatation of the superficial vessels of the dermis, presence of eosinophils and dermal edema can be seen.[25]

The most appropriate and effective measures to treat AP are physical protection with adequate clothing and avoiding sun exposure. It is advisable to wear long sleeves and pants, gloves, a wide-brimmed hat, sunglasses,

UVA-UVB sun protection factor greater than 30 and lip balm should be worn. The most effective systemic treatment is thalidomide,[25,26] due to its suppressive effect on TNF-α produced by keratinocytes, responsible for the marked lymphocytic infiltrate at the level of the upper dermis with a predominance of Th1.[27] The loading dose for adults would be 100 mg/day to 200 mg/day, and for pediatric populations 50 mg/d to 100 mg/d. For maintenance, a dose of 50 mg per week is recommended.[27,28]

Cyclosporin A reduces itching and improves skin lesions. It has also been used in eye drops (0.05% to 2%) for the treatment of conjunctival involvement.[29,30] Oral corticosteroids are reserved in case of great severity and its therapeutic duration would be short courses with methyl-prednisolone initially at 20 mg per day.[24] The use of PUVA (UVA plus psoralens; 8-MOP or 5-MOP) or NBUVB (narrow band UVB) would be useful, but at discontinuation of treatment it may relapse. Antihistamines, topical corticosteroids and emollients are reserved for the control of pruritus or a small number of lesions. Topical calcineurin inhibitors can also be used.[19] Hydroxychloroquine is an option at doses of 200 mg/day to 400 mg/day.

Conclusion

The diagnosis of actinic prurigo we made at the Andes Mountain is based on an exhaustive clinical history, including place of origin and residence, personal and family history, morphology of the rash and pathological anatomy study in certain cases. It is advisable to request a photo proof and photopatch test. The photo test would evaluate the cutaneous response to UVA, UVB and visible light, looking specifically at the minimum dose of erythema for UV and the urticarial response to visible light, while the photopatch test would be requested in case of suspected photo contact with an allergen.[31] The response to thalidomide can also be considered a diagnostic factor. It is essential to remember that this pathology leads to a significant decrease in the quality of life.

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