

WHAT IS YOUR DIAGNOSIS?

Case for diagnosis

Caso para diagnóstico

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HISTORY OF THE DISEASE

A thirteen-year-old black boy, native of Sao Paulo, complained of recurring pruritic lesions on his forearms associated with edema of the lower lip, photophobia, and red eyes that had developed two years before. Physical examination revealed conjunctival enanthema associated with pterygium (Figure 1). The lower lip was everted, swollen, exulcerated with vegetant lesions all over its extension (Figure 2). Erythematous, scaly lesions in the nose (Figure 1); excoriating papules and nodules in the forearms were observed in association with residual hyperchromic maculae (Figure 3). The patient presented low cognitive development without previous diseases. He denied similar cases of the disease in his family.

Skin biopsy of the lower lip showed acanthosis and spongiosis with dermal perivascular mononuclear cell infiltration composed by lymphocytes, plasma cells, and eosinophils (Figure 4). Eye examination revealed enanthema, Trantas dots, and pterygium. After three months of thalidomide use (100mg/day), remission of lesions was observed, including that of cheilitis. The patient suspended the medication and symptoms recrudesced. Treatment with thalidomide 100mg/day for six months was reestablished with rapid improvement; the dose was then reduced to 50mg/day and lesions were controlled for two more years. The patient's low cognitive development prevented the progressive reduction of medication.



FIGURE 1: Conjunctival enanthema, erythematous, scaly lesions in the nose and cheilitis



FIGURE 3: Papular, exulcerated, exudative, and excoriating lesions on the extensor region of the forearms



FIGURE 2: Exulcerated lower lip with vegetant lesions

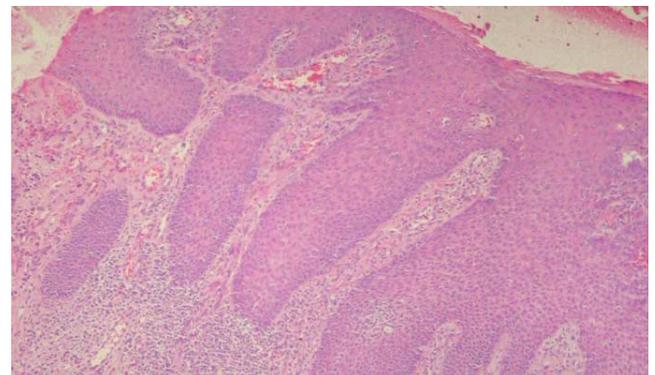


FIGURE 4: Acanthotic epidermis, parakeratosis and serous exudate, inflammatory infiltrate composed by lymphocytes and eosinophils in the papillary dermis

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COMMENTARIES

The case was diagnosed as actinic prurigo (AP), a rare acquired photodermatosis, characterized by pruritic, excoriated papules and nodules in photoexposed areas.¹ It is believed that the disease is caused by a late hypersensitivity reaction against autoantigens induced by ultraviolet radiation in genetically predisposed individuals.² However, these chromophores have not been identified.¹⁻³ Ultraviolet A and B rays are involved in the process.³

The disease was described among North-American Indians and Central and South America mestizos, with very few reports in the Asian and Caucasian population.³ Actinic prurigo is strongly associated with HLA-DR4, particularly the DRB1*0407 subtype.³ The electronic search of the *Anais Brasileiros de Dermatologia* did not yield other reports; a single review article from 1984 was accessed.⁴

Symptoms develop at approximately 10 years of age, affecting mostly women. The disease has a chronic course with exacerbations and remissions.⁵ In temperate climates, it improves in the winter, whereas in tropical climates it stays the same throughout the year.⁶ Skin lesions develop in photoexposed areas of the forearms, hands, neckline, face, ears, legs and feet.⁽¹⁻³⁾ Covered areas, such as the gluteal region, may

also be involved.³ Mucosas are frequently affected, especially the lips (30-60%) and conjunctiva (about 30%).⁷ The most relevant eye alterations are photophobia, pterygium, pinguecula, enanthema, Trantas' dots, hyaline exudates, and follicles, that is, predominantly conjunctival alterations.⁷

Histopathologic examination reveals discreet acanthosis, spongiosis, and perivascular lymphocytic infiltrate.⁵ Eosinophils are also found.^{6,7}

Treatment of AP includes photoprotection, topical and systemic corticosteroids, antihistamines, anti-malarial drugs, low doses of phototherapy (PUVA and UVB), and thalidomide. The last two appear to have a prophylactic effect.^{1,5,6} Thalidomide reduces polymorphonuclear leukocyte phagocytosis, the production of tumoral necrosis factor-alpha (TNF-alpha), lymphocyte production, inhibits angiogenesis and converts the T-helper 1 cell response to T helper 2.⁵ It is not clear how these effects modulate the activity of actinic prurigo.⁵ The use of thalidomide (50 to 100 mg/day) leads to the rapid lightening of skin lesions with a slower response of mucous lesions.^{1,5,6} When treatment is suspended, symptoms return. Therefore, low maintenance doses are recommended with control of adverse reactions.^{3,6} There is a report of control of the disease with the use of thalidomide for 23 years.¹ □

Abstract: A 13-year-old black boy had pruritic papular and nodular lesions on his forearms associated to edema of the lower lip, photophobia, conjunctivitis and pterygium. Skin biopsy of the lower lip revealed acanthosis, spongiosis with dermal perivascular mononuclear cell infiltration composed by lymphocytes, plasma cells and eosinophils consistent with actinic prurigo. Lesions improved considerably with the use of thalidomide 100mg/ day.

Keywords: Cheilitis; Photosensitivity disorders; Prurigo; Thalidomide; Ultraviolet rays

Resumo: Paciente do sexo masculino, negro, 13 anos, apresenta há dois anos lesões pruriginosas, pápulo-nodulares nos antebraços, associadas a edema do lábio inferior, fotofobia, conjuntivite e pterígio. O exame histopatológico do lábio inferior revelou acantose, espongiose e infiltrado inflamatório perivascular superficial, composto por linfócitos, plasmócitos e eosinófilos, compatível com o diagnóstico de prurigo actínico. As lesões regrediram com o uso de talidomida 100 mg/dia.

Palavras-chave: Prurigo; Queilite; Raios ultravioleta; Talidomida; Transtornos de fotossensibilidade

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