O’Brien’s Actinic Granuloma: an unusually extensive presentation

Granuloma actínico de O’Brien: apresentação clínica exuberante

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Abstract: O’Brien’s actinic granuloma is a rare skin disease. Controversy continues over whether it should be considered a specific condition or a form of granuloma annulare located in sun-exposed areas. Its pathogenesis is unknown; however, the most widely accepted hypothesis suggests that solar radiation is the triggering factor. This paper describes the case of a 78-year old, fair-skinned male, who presented with a 10-year history of an infiltrate of annular erythematous papules on his forehead and left malar region. The diagnosis of O’Brien actinic granuloma was established from histopathology, since the clinical condition of the patient was extensive, unlike cases reported in the literature.

Keywords: Giant cells; Granuloma; Granuloma, giant cell

Resumo: O granuloma actínico de O’Brien é uma doença cutânea rara. Há muitas controvérsias na sua caracterização, podendo representar uma entidade nosológica distinta ou uma forma de granuloma anular em áreas expostas. A patogênese é desconhecida; no entanto, acredita-se que a radiação solar seja um fator desencadeante. Os autores relatam o caso de um paciente de 78 anos, branco, que apresentava há 10 anos pálulas eritematosas e infiltradas na fronte e hemiface à esquerda. O diagnóstico de granuloma actínico de O’Brien foi estabelecido a partir do exame histopatológico, uma vez que o quadro clínico exuberante apresentado pelo paciente diferia do de casos anteriormente observados.

Palavras-chave: Células gigantes; Granuloma; Granuloma de células gigantes

INTRODUCTION

Actinic granuloma, annular elastolytic giant-cell granuloma or O’Brien granuloma is a rare dermatosis that was first described in 1975. 1 Few cases have been reported in the literature and classification of this condition as a specific disease is questioned, since it is considered by some authors as a form of granuloma annulare that affects sun-exposed areas of the skin. 2 Clinically, it is characterized by erythematous papular lesions grouped together to form asymptomatic annular lesions with raised borders and atrophic centers. The areas most commonly affected are the head, neck and hands. 3

Histological analysis reveals skin granulomas with multinucleated giant cells containing degenerating elastic fibers, a process known as elastophagocytosis. 4

This paper reports the case of a patient with a diagnosis of O’Brien’s actinic granuloma in which the clinical manifestation was unusually extensive. He responded well to therapy.
CASE REPORT

A 78-year old white man, originally from Hungary, with erythematous, infiltrated lesions on his forehead and left malar region for the past ten years, extending progressively to his face, scalp and ear lobes. The condition became worse following sun exposure. He had no pruritus or burning sensation.

Dermatological examination identified the presence of multiple erythematous papules grouped together to form well-delimited, annular-like lesions with signs of atrophy at the center. There was a diffuse edema in the left malar region, including areas of ulceration with a granular base. There was evidence of hair loss on the areas of the scalp affected by the lesions (Figure 1).

Supplementary tests performed consisted of: full blood count, kidney and liver function, glucose and urinalysis, tuberculin skin test (Mantoux test), serology for syphilis, microscopy for acid-fast bacilli, Mitsuda reaction, pilocarpine test and x-ray of the chest and hands. All were negative or within the normal range.

Biopsy of the lesion showed a healing epidermis and the presence of an inflammatory process in the dermis characterized by an epithelioid histiocytic reaction, forming granulomatous clusters with multinucleated giant cells in addition to a dense lymphomononuclear infiltrate. The process was affecting adnexal structures. Microscopy for acid-fast bacilli using Ziehl-Neelsen staining was negative. Specimens stained using Verhoeff’s method showed the presence of degenerating elastic fiber inside some giant cells (Figures 2, 3 and 4).

A diagnosis of O’Brien’s actinic granuloma was based on histopathology, since the unusually extensive clinical presentation in this case was different from other previously reported cases.

Treatment was initiated with 25 mg/day of acitretin and continued for nine months. There were signs of improvement by the third month of treatment (Figure 5). An increase in cholesterol and triglyceride levels to more than twice the upper limit of normal led to suspension of the medication in this elderly patient. Maintenance therapy was then initiated with the use of sunscreen and the condition remains inactive to this date.
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DISCUSSION

O’Brien’s actinic granuloma is a chronic disease, although spontaneous remission has been reported in some cases. The condition affects middle-aged individuals of either sex who have a history of intense sun exposure. Clinically, it is characterized by the presence of erythematous papules on sun-exposed areas that group together to form asymptomatic annular plaques with mild central atrophy.

Although several of the clinical findings in the present case resembled those described in the literature, classification of the condition was hampered because of the unusual extent of skin involvement and because of the areas of hair loss. A case in which there was an association with alopecia was described in the literature; however, in this case, histopathology failed to confirm that the granulomatous inflammation had directly affected the hair follicle.

The pathogenesis of the condition is unknown; however, solar elastosis is believed to be a triggering factor. In this hypothesis, the degenerating elastic tissue is believed to become a target for an immunological reaction.

Since the condition was first described, there has been much controversy regarding whether O’Brien’s actinic granuloma is a separate nosological entity, since some authors consider it to be a form of granuloma annulare that affects sun-exposed areas of the skin. Although the two dermatoses are clinically similar, certain histopathological characteristics classify actinic granuloma as a separate condition.

In the case of actinic granuloma, multinucleated giant cells with degenerating elastic fibers are found in its interior, and there is an absence of mucin and a sarcoid-like granuloma confined to the superficial dermis at solar elastosis level. On the other hand, in granuloma annulare, an inflammatory infiltrate in a palisade pattern surrounds degenerating collagen tissue in the superficial and deep dermis. Furthermore, mucin is present. An interstitial granulomatous infiltrate is found in both conditions.

The mechanism of this disease has been investigated by immunohistochemistry and suggests a cell-mediated immune response against the degenerating elastic tissue. In actinic granuloma, the presence of lysozyme is found in the giant cells and there is a predominance of T-helper cells in the lymphocytic infiltrate. This predominance is also found in other dermatoses in which there is an abnormal response to ultraviolet radiation such as polymorphic light eruption and photosensitivity to drugs.

Some authors have reported lesions resembling actinic granuloma in unexposed areas, i.e. areas with no solar damage. In these cases, the term annular elastolytic giant cell granuloma has been suggested.

O’Brien’s actinic granuloma may be associated with temporal arteritis, polymyalgia rheumatica, relapsing polychondritis and pseudoxanthoma elasticum, none of which were seen in this patient.

Several different forms of treatment have been proposed with varying results: chloroquine, intraleisional steroids, cyclosporine, pentoxifylline, methotrexate and cryotherapy. Case reports show positive results with the use of isotretinoin, acitretin and retinoid psoralen + UVA (RePUVA). The mechanism of action of systemic retinoids is not yet completely understood; however, it is believed to be associated with their effect on photo-aging and actinic elastosis, both of which are present in the etiology of actinic granuloma.

The patient responded well to treatment with...
O'Brien’s actinic granuloma. The lack of cases reported in the literature makes the appropriate classification of this condition even more of a challenge.

REFERENCES

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