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Case Report

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Lichenoid Sarcoidosis Simulating Pseudofolliculitis Barbae

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Introduction

Lichenoid sarcoidosis is an extremely rare cutaneous variant of sarcoidosis, occurring in approximately 1-2% of all cutaneous presentations [1]. It is classically described in Asian children and usually affects the eyes and joints, sparing respiratory involvement [2] .However, case reports in adults and the elderly are frequent [3-7].

Clinically, lesions are characterized by erythematous or violaceous papules measuring 1-3 mm which may be singular or numerous and present with mild desquamation. Distribution is primarily on the face, trunk, and limbs [8].

Reports indicate that cutaneous sarcoidosis is frequently misdiagnosed as other skin diseases, highlighting the importance of follow up by the dermatologist. The severity of systemic involvement and prognosis may vary according to the type of cutaneous manifestation [9].

Case Report

A 30-year-old male patient presented with a 6-year history of violaceous, desquamative, and pruritic papules starting on the right beard region as a single lesion, which then progressed symmetrically leftwards after seven months and was also present in the scrotal region. The lesions appeared and regressed spontaneously in the three described regions during this seven-month period.

Patient's personal history included allergic rhinitis, controlled asthma, and gastroesophageal reflux disease. Patient was taking Relvar Ellipta, Aerolin and Omeprazole. The patient denied a previous history of tuberculosis. Regarding epidemiological history: the patient lived in Ireland in 2016, where he worked as a cleaner and was exposed to mold and cleaning products.

On examination, he presented multiple brownish and purplish papules, some confluent in plaque with an atrophic center, and desquamation in the beard region, including the mentum and the body of the jaw. The same aspect was noted in the scrotal region. He presented no adenopathy visceromegaly, or pain. (Figure 1A, Figure 1B, Figure 1C, and Figure 1D)

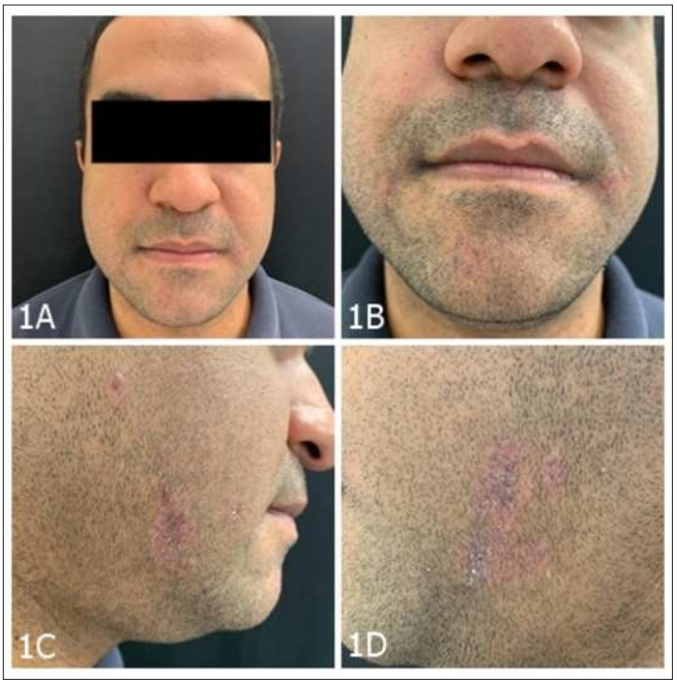

Figure 1A: Dermatological examination of the face, global view
Figure 1B: Clinical examination at higher magnification, showing erythematous-violaceous and infiltrated lesions in the beard region
Figure 1C: Multiple erythematous-violaceous papules, confluent into a plaque with an atrophic center and infiltrated borders, topped by thin desquamation
Figure 1D: Atrophic erythematous-violaceous plaque infiltrated

in the left mandibular region with irregular borders

Dermoscopy of the lesions showed brownish, homogeneous, rounded, and well-defined papules. We did not note the presence of Wickham's streaks. (Figure 2)

Figure 2: Dermoscopy showed an apple-jelly appearance, and the presence of Wickham's striae was absent

We started treatment for a month with Clindoxyl and Sébium Foaming Gel. As these treatments were unsuccessful, we then started treatment for Tinea barbae with topical ketoconazole for 40 days. In the follow-up consultation the patient presented the same lesions with no improvement. Thus, we opted for biopsy, which evidenced the diagnosis of lichenoid sarcoidosis. (Figure 3A, Figure 3B, Figure, 3C and Figure 3D)

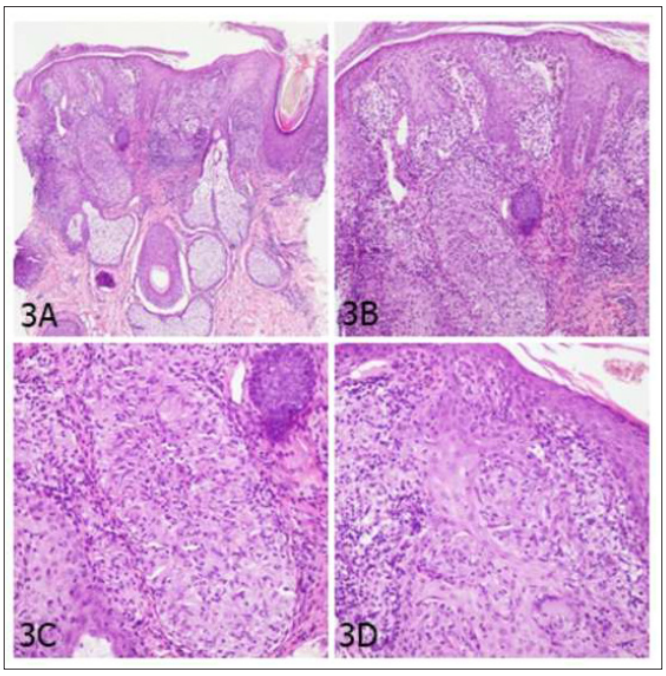

Figure 3A: Panoramic view of the anatomopathological examination, including in the material epidermis, dermis, subcutaneous cellular tissue, and skin annexes

Figure 3B: A 20-fold magnification, showing in the papillary and reticular dermis, infiltration of epithelioid histiocytes forming well-defined nodules of varying sizes, with some multinucleated giant Langhans-like cells. Lymphocytes are also visible in the permeation and around the blood vessels of the superficial vascular plexus. At this level, the proliferation of fibroblasts and thickened collagen fibers are also noted with melanophages. The infiltrate approaches the epidermis in a large part of the fragment, where vacuolar degeneration of the basal layer is seen. In the deep dermis, the infiltrate consists of sparse, isolated nodules. The epidermis shows irregular acanthosis with hypo- or agranulosis and continuous parakeratosis crust. The hair follicles are preserved.

Figure 3C: A 40x magnification, showing granuloma composed of epithelioid histiocytes, with some multinucleated giant Langhans cells; absence of caseous necrosis is noted

Figure 3D: A 40x magnification, showing lymphocytes distributed preferentially around the blood vessels of the superficial vascular plexus. At this level, the proliferation of fibroblasts and thickened collagen fibers are also noted with melanophages. The infiltrate approaches the epidermis in a large part of the fragment, where vacuolar degeneration of the basal layer is seen. The epidermis exhibits irregular acanthosis with hypo- or agranulosis and continuous parakeratosis crust. The hair follicles are preserved. Diagnosis is compatible with granulomatous dermatitis of the lichenoid tuberculoid type (sarcoid granuloma), compatible with lichenoid sarcoidosis.

The chest CT scan presented mediastinal and hilar lymphadenopathy with "icing sugar" images, compatible with systemic sarcoidosis. Spirometry showed an obstructive pattern, which was reversible after bronchodilator testing, consistent with his history of asthma. (Figure 4A and Figure 4B) Ophthalmology evaluation ruled out ocular involvement of sarcoidosis.

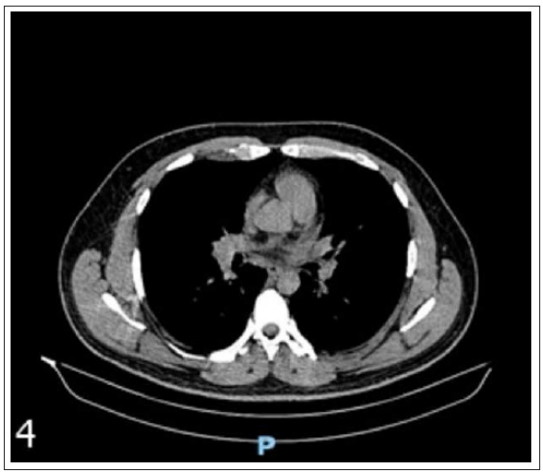

Figure 4: Chest computed tomography scan showing discrete lymphadenomegaly in the various mediastinal and hilar chains bilaterally, concomitant with the presence of pulmonary nodules and sparse bilateral regular contours, most of them in subpleural or peribronchovascular topography, up to 1.0 cm in diameter; changes related to the clinical hypothesis of sarcoidosis.

With guidance from the pulmonologist, we started treatment with oral prednisone 50 mg a day for 30 days followed by a progressive medication withdrawal. After initiating this treatment, the patient saw improvement in the condition of lesions and systemic involvement, confirming that lichenoid sarcoidosis was the accurate diagnosis.

Discussion

This article describes a case of lichenoid sarcoidosis in an adult patient mimicking a common disorder: beard folliculitis. This is a differential diagnosis, distinct from those typically described for

lichenoid sarcoidosis: lichen planus, subacute cutaneous lupus, cutaneous tuberculosis, leprosy, and cutaneous leishmaniasis [3].

The diagnosis of sarcoidosis remains challenging and time-consuming. The dermatoscopic findings of homogeneous brownish plaques or yellow dots are suggestive of granulomatous diseases [3]. but do not define the diagnosis of lichenoid sarcoidosis, requiring exclusion of infection and anatomopathological study. Biopsy enables prompt differentiation from lichen planus in the absence of Wickham's striae [6].

Previous articles have described lichenoid sarcoidosis clinically mimicking lichen planus and erythroderma due to drugs or malignancies [4,7]. In these cases, the patients were first treated for other conditions. Lichenoid sarcoidosis was diagnosed after treatment failure and anatomopathological study, thereby delaying the onset of specific treatment. In our patient this interval was four months.

Due to scarcity of literature, clinical experience determines the course of treatment for sarcoidosis and there is a lack of standardization [9]. In asymptomatic or cosmetically insignificant cases, treatment is conservative as the disease may remain stable or regress spontaneously [2]. Treatment with topical or intralesional corticosteroids may be indicated in mild or moderate disease. Immediate systemic corticosteroid or corticosteroid-sparing therapy is suggested in cases where the disease is disseminated, the patient has disfiguring lesions, or in cases of lupus pernio. If the aforementioned treatments are unsuccessful, anti-TNF alpha, laser, or surgery should be used [2, 7].

Data regarding therapeutic options remain conflicting. Isolated reports have already shown success with systemic application of Tacrolimus in the remission phase; however, the lesions returned after 4 months thus requiring the use of methylprednisolone to increase remission duration [8]. Conversely, Tacrolimus has been shown to increase remission duration of lichenoid sarcoidosis as compared to systemic and topical corticotherapy [3, 7].

More recently, topical pimecrolimus 1% was applied after topical clobetasol failed to treat lichenoid sarcoidosis lesions on the right knee; after 12 months, the patient remained relapse-free [5].

In this report, the patient was treated with systemic corticosteroids at a dose of 50 mg/d for 30 days due to presentation of pruritus, aesthetic impairment, and systemic involvement, evolving with significant improvement of symptoms and lesions. Although lichenoid sarcoidosis has previously been observed to be refractory to oral corticotherapy our patient showed rapid resolution of the condition with administration of this treatment with the possibility of weaning and no need for maintenance medication to prevent relapses [7]. This resolution is consistent with the findings of Sanchez-Lopez [4].

Anti-TNF agents are indicated in refractory cases of cutaneous sarcoidosis to inhibit the formation of granulomas, in which TNF plays a central role [2]. However, there have been reports of the occurrence of lichenoid sarcoidosis induced by certolizumab pegol (an Anti-TNF agent) in patients with rheumatoid arthritis [10].

Among the options reviewed in this article, we conclude that corticosteroids are still the most widely accepted therapy worldwide and guarantee more extended periods of remission. However, due to scarcity of literature clinical experience is the primary determinant of treatment for sarcoidosis. More case reports with successful treatment need to be published and randomized clinical trials should be performed to increase the scientific basis of treatment.

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