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(CASE REPORT)

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Lichenoid Sarcoidosis: A rare form of cutaneous sarcoidosis

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Abstract

Background: Sarcoidosis, also known as Besnier-Boeck-Schaumann disease, is a systemic granulomatous disease of unknown origin that can affect various organs. Cutaneous manifestations are diverse, presenting initially in 80% of cases and being the sole sign of the disease in 25% of patients. Among these manifestations, lichenoid sarcoidosis is a rare form, accounting for only 1 to 2% of cutaneous cases. First described in 1899, it manifests as slightly scaly papules, primarily located on the trunk and face. Although often benign, this form can be associated with ocular and joint complications, especially in children. This article presents a case of lichenoid sarcoidosis in a 44-year-old woman.

Methods: we present the case of A 44-year-old woman presented with asymptomatic skin lesions on the trunk and face, evolving over three months. Clinical examination revealed flesh-colored papules grouped on the face, neck, and back.

Results: Histological analysis showed non-caseating epithelioid granulomas. Laboratory tests were normal except for elevated ACE levels. Based on clinical and histological findings, a diagnosis of lichenoid sarcoidosis was made, and treatment with hydroxychloroquine and topical corticosteroids was initiated.

Discussion: Sarcoidosis is a multisystemic disease of unknown etiology, primarily affecting young adults, with a female predominance. Cutaneous manifestations, present in 20–25% of cases, may be nonspecific (such as erythema nodosum) or specific, featuring non-caseating granulomas. The lichenoid form, first described in 1899, presents as slightly scaly papules, mainly on the trunk and face. Though often benign, it may be associated with ocular and joint complications, particularly in children. In this case, the patient had no pulmonary or ocular involvement, and histological analysis confirmed the characteristics of lichenoid sarcoidosis. The main differential diagnoses include lichen scrofulosorum and other lichenoid dermatoses.

Conclusion: This article reports a case of lichenoid sarcoidosis in a woman, illustrating this rare form of cutaneous sarcoidosis, diagnosed based on clinical observation and histopathological analysis.

Keywords: Sarcoidosis; Lichenoid Sarcoidosis; Granulomas; Besnier-Boeck-Schaumann disease

1. Introduction

Sarcoidosis, or Besnier-Boeck-Schaumann disease, is a systemic granulomatous condition of unknown etiology that can affect any organ. Cutaneous manifestations are multiple and polymorphic, occurring as the initial symptom in 80% of cases and representing the sole manifestation in 25%.

Lichenoid sarcoidosis is an extremely rare cutaneous variant, accounting for approximately 1–2% of all cutaneous presentations. Clinically, it is characterized by asymptomatic, erythematous or flesh-colored papules, most often

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localized on the trunk and face [1–4]. This entity has been rarely described in the literature; we report here a case of lichenoid sarcoidosis in a 44-year-old woman.

2. Material and methods

A 44-year-old female patient, with no known medical history, presented with isolated, asymptomatic skin lesions located on the trunk and face, evolving over the past 3 months. Clinical examination revealed monomorphic, firm, smooth-surfaced, flesh-colored papules grouped on the face, nape, and back (Figure 1 and 2). The rest of the physical examination was normal, with no palpable peripheral lymphadenopathy.

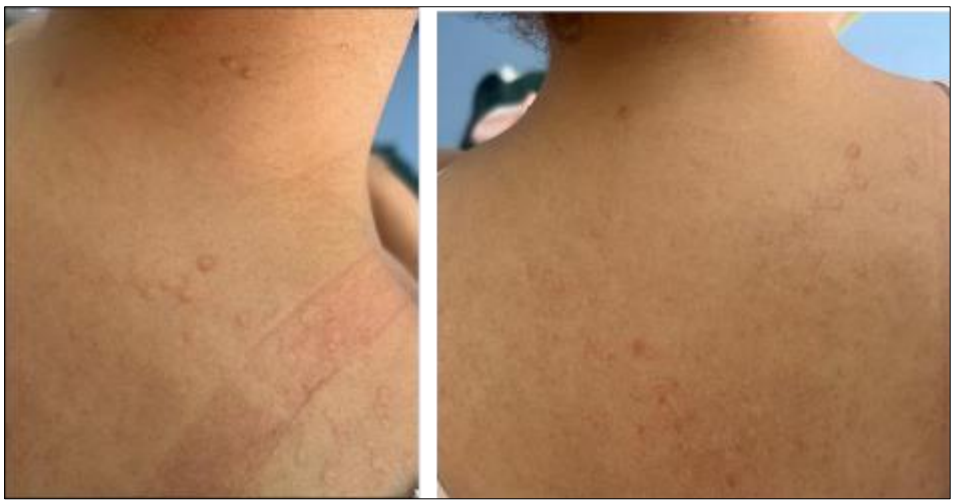

Figure 1 and 2 Flesh-colored papules grouped on the face, nape, and back

3. Results

A skin biopsy revealed numerous variably sized, focally confluent granulomas composed mainly of epithelioid and giant cells, without caseating necrosis. Blood work, including liver and kidney function tests, were normal. However, the serum angiotensin-converting enzyme (ACE) level was elevated at 260 IU/L. Given the clinical and histological findings, a diagnosis of lichenoid sarcoidosis was established. The patient was started on hydroxychloroquine and topical corticosteroids.

4. Discussion

Sarcoidosis is a multisystemic granulomatous disease of undetermined etiology. It is a ubiquitous condition that primarily affects young adults aged 20–40 years, with a female predominance [1].

Cutaneous manifestations vary greatly, occurring in 20–25% of cases according to literature, usually after intrathoracic or lymph node involvement. They are classified into non-specific lesions (mainly erythema nodosum) and specific sarcoid lesions, which histologically present as non-caseating tuberculoid granulomas [2,6].

Lichenoid sarcoidosis was first described in 1899 by Boeck [3]. Clinically, it presents as multiple dome-shaped, erythematous or flesh-colored, slightly scaly maculopapules. They can be isolated or grouped, mainly located on the trunk, limbs, and face. Dermoscopic examination typically reveals homogeneous yellow-brown macules and the "apple jelly" sign; however, Wickham striae are absent.

This form is particularly noted in children under 6 years old and is often associated with ocular and joint complications, though respiratory involvement is absent [5,7]. While skin lesions can regress spontaneously, ocular lesions may progress and cause blindness in about 22% of cases [7]. These may be present at diagnosis or appear years later, as described by Hafner and Vogel in a series of 12 children who developed polyarthritis and uveitis during long-term follow-up (5–30 years) [5,7]. In adults, lymph node and pulmonary involvement are the most common extracutaneous localizations.

In our case, the patient had no extracutaneous involvement. Ophthalmologic and pulmonary assessments were normal. Histopathology typically reveals epithelioid granulomas in the upper dermis, sometimes in a follicular pattern; multinucleated giant cells and lymphocytes surround the granulomas, without caseating necrosis. Only one case of lichenoid distribution of epithelioid granulomas at the dermoepidermal junction has been described by Garrido-Ruiz and Enguita-Valls, supporting the diagnosis of lichenoid sarcoidosis both clinically and histologically [2].

The main differential diagnosis is lichen scrofulosorum, which may resemble lichenoid sarcoidosis both clinically and histologically. However, in lichen scrofulosorum, the tuberculin skin test is strongly positive, and pulmonary parenchymal involvement is often present. Other differential diagnoses include lichen nitidus, lichenoid drug eruptions, lichen amyloidosis, and papular mucinosis.

5. Conclusion

In conclusion, we report the case of a young woman with lichenoid sarcoidosis, a rare form of cutaneous sarcoidosis. The diagnosis was suggested by the atypical clinical presentation of lichenoid papules distributed on the trunk and limbs and confirmed by histopathological examination.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

All the procedures were carried out after the agreement of all individual participants included in the study.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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