Lymphocytoma cutis - Case report

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Abstract: We describe a clinical case involving a 62-year-old white male, diagnosed with lymphocytoma cutis (Spiegler-Fendt sarcoid) in the cephalic segment. The diagnosis was carried out by pathological study and confirmed by immunohistochemical panel: evidence of polyclonality. Phototherapy sessions were suggested as treatment (13 PUVA sessions, with an accumulated dose of 58.65 J/cm²). The improvement was partial. Thus, infiltration of triamcinolone was opted for (one intralesional infiltration every 3 weeks). After 5 sessions, satisfactory improvement was observed: regression of nearly all the lesions.

Keywords: Immunohistochemistry; Lymphoma; Pseudolymphoma; Triamcinolone acetonide

INTRODUCTION

Lymphocytoma cutis, or Spiegler-Fendt sarcoïd, is classed as one of the pseudolymphomas,1 referring to inflammatory disorders in which the accumulation of lymphocytes on the skin resemble, clinically and histopathologically, cutaneous lymphomas.1-3

In order to obtain an accurate diagnosis, careful clinical evaluation is needed, as well as histopathological and immunohistochemical exams.1-3

Here, we describe a case involving a patient over 60, with lesions distributed across upper parts of the head, whose final diagnosis was lymphocytoma cutis.

Despite the improvement in the lesions following corticoid infiltrations, the patient remains under clinical observation, since, in some cases, the condition can evolve into cutaneous lymphoma.

CASE REPORT

White male, 62, suffering for a year from lesions in the skin in the frontal, zygomatic lateral and temporal anterior (both bilaterally), regions. There was no itching but he experienced occasional pain to local palpation. He sought help from doctors, who prescribed ketoconazole shampoo and antihistamines, among other medications, but to no avail.

The patient denied being a smoker or a heavy drinker, and was taking omeprazole to control the reflux disease.

The physical exam revealed macules, papules, nodules and plaques. The lesions presented in an erythematous-violaceous color, and others in erythematous-pink. There was no scaling (Figures 1 and 2).

Further diagnoses included sarcoidosis, pseudolymphoma and cutaneous lymphoma. Other
more unlikely hypotheses were: luminal eruption, rosacea and erythematous lupus, borreliosis.

Serum exams revealed no alterations. The serum calcium dosage was applied, as a hypercalcemia suggested the diagnosis of sarcoidosis.

The ANF was non-reactive, suggesting, in principle, that the condition was not autoimmune.

There were non-reactive IgM and IgG antiborreli antibodies, eliminating the possibility of borreliosis.

A chest x-ray revealed no pulmonary alterations (which thus reduced the chances of sarcoidosis). An ultrasound of the abdomen showed the presence of light hepatic steatosis.

The skin biopsy revealed hydropic degeneration of the basal layer. In the dermis, there was chronic and nonspecific inflammatory infiltrate, with a predominance of lymphocytes in the reticular dermis and the formation of lymphoid follicles: germinative centers (Figure 3).

The immunohistocemistry confirmed polyclonal lymphoid infiltrate: lymphocytes B-CD20 and lymphocytes T-CD3 (Figures 4 and 5).

The immunohistochemical analysis was also complemented by the BCL2, CD4, CD7 and CD8, markers; thus reaffirming the polyclonality.

The initial treatment suggested was phototherapy: 13 PUVA sessions were carried out in a period of 2 months, with a total accumulated dose of 58.65 J/cm² (8-MOP administered twice a week with a dose of 0.65mg/kg). There was some improvement, though the patient could not continue with this treatment.

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Hence, the chosen treatment was infiltrations of triamcinolone acetonide 40mg/ml, every 10-15 days. Clinical improvement was noted during the 5th session. (Figure 6)

DISCUSSION

Lymphocytoma cutis, or Spiegler-Fendt sarcoid, can be understood as a lymphoreticular and hyperplasic reaction occurring in the dermis.¹⁻²,⁴ It is a pseudolymphoma of the B cells.⁴⁻⁵ Many possible triggering factors have been identified: acupuncture, earrings, piercings, tattoos, phenytoin, vaccinations, infections from the herpes simplex virus and zoster, insect bites, and infection by the bacterium *Borrelia burgdorferi*.¹⁻²,⁵ However, in most cases, the trigger seems to be idiopathic.²,⁵

It is a rare disease, generally affecting young adults (under the age of 40), with a predilection for females (3:1).²⁻⁵ However, cases involving different age groups have been reported.¹ It affects predominantly caucasians.⁵

Clinically, lesions present as papules and/or nodules that can be shiny.¹⁻⁸ They are not scaly and can be skin-colored, erythematous, brownish or violaceous.¹⁻⁴,⁵ Itching can occur. In the literature, it is common to find descriptions of lesions with softened consistency.¹⁻⁵ In the case under discussion, the consistency of the lesions was similar to that of non-affected skin.

The localized form accounts for 70% of cases, occurring predominantly on the face or upper limbs. The disseminated form is rarer, found mostly in elderly people, affecting the midsection and extremities, in addition to the face.⁵⁻⁶ The clinical condition in which there is dissemination of lesions is termed miliary lymphocytoma cutis.⁵⁻⁶

Differential diagnoses include: cutaneous lymphoma, sarcoidosis, erythematous lupus, angiolymphoid hyperplasia, luminal eruption and rosacea.¹⁻³,⁵

Histopathologically, it is characterized by foci of lymphocytes in the dermis, permeated by histiocytes, bringing about a follicular arrangement that is translated by B cells bordered by T cells, that is to say, the germinative centers. With lymphomas, there are no germinative centers, except in cases of malignant lymphomas of the giant follicular type.³

The dermis can be separated by a thin area of collagen.²⁻⁵

Immunohistochemistry reveals the polyclonality (presence of B lymphocytes and T lymphocytes).³⁻⁵

Therapeutics options include systemic corticoids, corticoid infiltrations, superficial radiotherapy, PUVA, cryosurgery, hydroxychloroquine, surgery, photodynamic therapy.¹⁻²,⁴⁻⁷,⁹⁻¹⁰ When the lymphocytoma cutis is associated with borreliosis, treatment is tailored to this condition.

Follow-up of patients is crucial, as the condition can evolve into cutaneous lymphoma.²⁻⁴

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