CASE REPORT

Lymphocytoma cutis - Case report*

Linfocitoma cutis - Relato de caso

Ederson Valei Lopes de Oliveira[1] Giovana Bachega Badiale[2] Margarida M. F. S. Moraes[3]

DOI: http://dx.doi.org/10.1590/abd1806-4841.20132320

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

Resumo: Relatamos um caso de um paciente de 62 anos, branco e com diagnóstico de linfocitoma cutis (Sarcoide de Spigler-Fendt) em segmento cefálico. O diagnóstico foi confirmado pelo histopatológico e reafirmado pelo painel imuno-histoquímico: evidência de policlonalidade. Sessões de fototerapia foram propostas como tratamento: 13 sessões de PUVA cuja dose acumulada foi de 58.65 J/cm2); a melhora foi parcial. Optado, então, por infiltrações de triancinolona (uma infiltração intralesional a cada 3 semanas). Na quinta sessão, satisfatória melhora já podia ser evidenciada: regressão de quase todas as lesões.
Palavras-chave: Imunoistoquímica; Linfoma; Pseudolinfoma; Triancinolona acetonida

INTRODUCTION

Lymphocytoma cutis, or Spiegler-Fendt sarcoid, is classed as one of the pseudolymphonas,[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

Received on 28.11.2012.
Approved by the Advisory Board and accepted for publication on 17.01.2013.
* Study carried out at the Clínica Endoderm – Ribeirão Preto (SP), Brazil.
Conflict of interest: None
Financial funding: None

[1] Dermatologist. Private clinic – Ribeirão Preto (SP), Brazil.
[2] Pathology specialist, qualified at the Medical School of the University of de São Paulo, Ribeirão Preto (FMRP USP). Ribeirão Preto (SP), Brazil.
[3] Pathologist. Master's in pathological anatomy. Training Doctor in the Pathological Anatomy department of the Medical School of the University of São Paulo, Ribeirão Preto (FMRP USP). Ribeirão Preto (SP), Brazil.

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 antiborrelia 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).

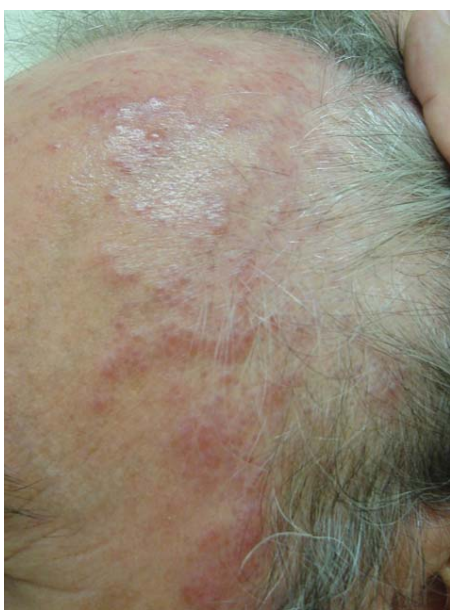

Figure 1: Presence of erythematous-violaceous and erythematous-pink macules, papules and nodules

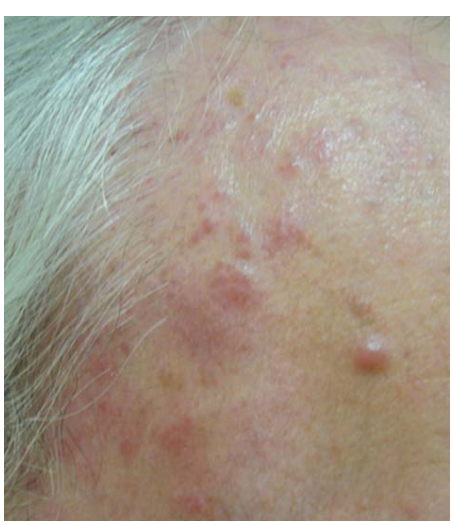

Figure 2: Macules, papules and plaques. Some are erythematous-violaceous and others are erythematous-pink. There are no scaly lesions

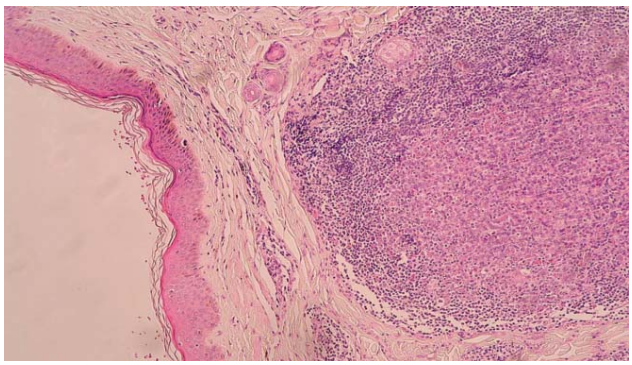

Figure 3: Skin fragment showing inflammatory infiltrate in the dermis. Predominance of lymphocytes in the reticular dermis. Formation of lymphoid follicles: germinative centers. (HE, 100x)

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/cm^2 (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.

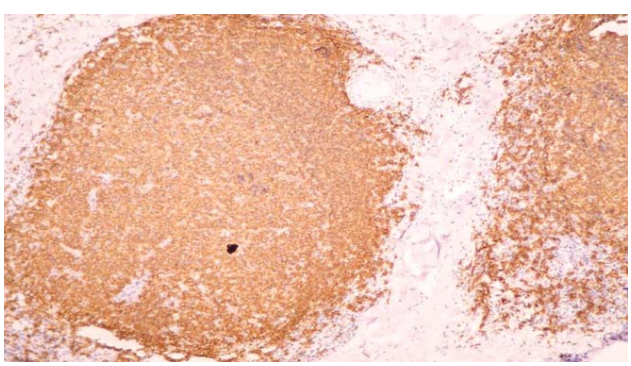

Figure 4: Immunohistochemistry: CD 20: B lymphocytes

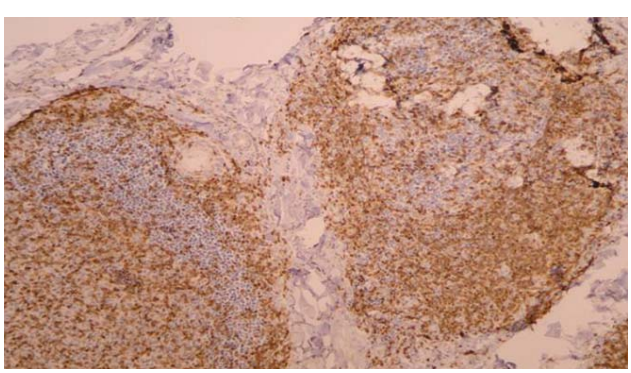

Figure 5: Immunohistochemistry: CD3: T lymphocytes

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)

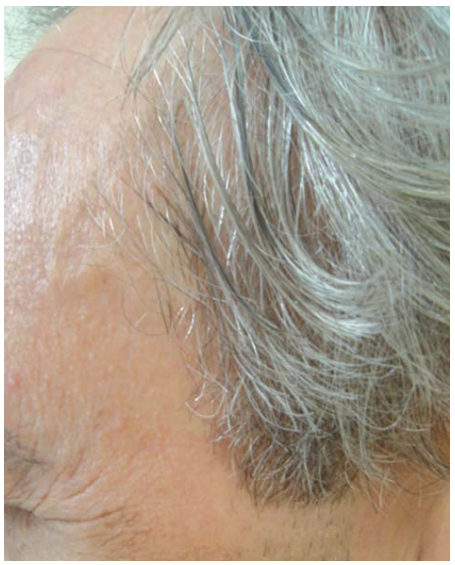

Figure 6: Regression of lesions after infiltrations of triamcinolone

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).[2,4-7] However, cases involving different age groups have been reported.[3] It affects predominantly caucasians.[5]

Clinically, lesions present as papules and/or nodules that can be shiny.[1-5,8] 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.[5-6,8] The clinical condition in which there is dissemination of lesions is termed miliary lymphocytoma cutis.[2,5]

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 from the epidermis 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] ❑

ACKNOWLEDGEMENTS

The authors are grateful for the valuable contributions of: Dr. Daniela Chaves Tarquínio, who conducted the phototherapy sessions; and Dr. Mariângela Esther Alencar Marques, who helped in the histopathological study.

REFERENCES

1. Sampaio SAP, Rivitti EA. Dermatologia 3 ed. São Paulo: Artes Médicas; 2007. p.1270-1.
2. Lemus SMV, Alarcón H, Ruiz SM. Linfocitoma Cutis. Reporte de un caso. Rev Cent Dermatol Pascua. 2003;12:75-8.
3. Peçanha PLVB, Pereira Jr AC, Castro O. Diagnóstico diferencial histopatológico e imuno-histoquímico entre linfoma maligno e pseudolinfomas (linfocitoma): Estudo de três casos. An Bras Dermatol. 1995;70:319-21.
4. du Vivier A. Micose fungoide e quadros correlatos. Atlas de Dermatologia Clínica. 3. ed. São Paulo: Manole; 2000. p.10.17-18.
5. Nasser N, Nasser Filho N, Rosa TSC. Linfocitoma cutis no nariz. PE-338. In: 67. Congresso Brasileiro de Dermatologia; 2012 set 1–4; Rio de Janeiro, Brasil. Rio de Janeiro: Sociedade Brasileira de Dermatologia; 2012.
6. Arai E, Shimizu M, Hirose T. A review of 55 cases of cutaneous lymphoid hyperplasia: reassessment of the histopathologic findings leading to reclassification of 4 lesions as cutaneous marginal zone lymphoma and 19 as pseudolymphomatous folliculitis. Hum Pathol. 2005;36:505-11.
7. Leinweber B, Colli C, Chott A, Kerl H, Cerroni L. Differential diagnosis of cutaneous infiltrates of B Lymphocytes with follicular growth pattern. Am J Dermatopathol. 2004;26:4-13.
8. van Vloten WA, Willemze R. The many faces of lymphocytoma cutis. J Eur Acad Dermatol Venereol. 2003;17:3-6.
9. Stoll DM. Treatment of cutaneous pseudolymphoma with hydroxychloroquine. J Am Acad Dermatol. 1983;8:696-9.
10. O'Neill J, Fien S, Zeitouni NC. ALA-PDT for treatment of cutaneous psudolymphoma: a case repor. J Drugs Dermatol. 2010;9:688-9.

Mailing address:
Ederson Valei Lopes de Oliveira
Avenida Santa Luzia, nº 550, 2° andar.Jardim Sumaré
14025-090 - Ribeirão Preto - SP
Brazil
E-mail: edersonvalei@yahoo.com.br

How to cite this article: Oliveira EVL, Badiale GB, Moraes MMFS. Lymphocytoma cútis - Case report. An Bras Dermatol. 2013;88(6 Suppl 1):S128-31.