Follicular mucinosis - Case report*

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Abstract: Follicular mucinosis, also known as alopecia mucinosa, is a cutaneous mucinosis histologically characterized by accumulation of dermal type mucin in the pilosebaceous follicle and sebaceous glands. It presents in two forms, a primary or idiopathic form and a secondary form associated with various benign or malignant processes. Among the malignant processes, the main association is with mycosis fungoides. The frequent overlap of clinical, histopathological, immunohistochemical and molecular biology characteristics makes the correct classification of these conditions difficult, therefore a long follow-up of all cases is recommended. We report the case of an adolescent with disseminated lesions and discuss the difficulty of early identification of secondary follicular mucinosis associated with cutaneous lymphoma.
Keywords: Lymphoma, T-Cell, cutaneous; Mucinosis, follicular; Mycosis fungoides

INTRODUCTION:

Follicular mucinosis (FM) is a cutaneous mucinosis in which there is a build-up of mucin at the external root sheath and with much less frequency in the sebaceous gland.[1] It was initially described in 1957 by Pinkus who named it *alopecia mucinosa* and renamed it *mucinose folicular* in 1959, due to the fact that alopecia is not always present.[2]

Two variants are found, one idiopathic or primary form, usually present in children and young adults, of spontaneous remission, and a secondary form that affects a higher age range and is associated with other benign and malignant processes, the mycosis fungoides (MF) being the main association.[2]

Clinical and histopathological criteria are fundamental for the distinction between primary and secondary forms; however, they are not specific enough for such definition, thus making the follow-up of patients necessary.[2]

CASE REPORT

Male patient, white, 13 years old, complained about spots on his body associated with pruritus for six months. At the physical examination he presented erythematous and hypopigmented macules, well defined, alopecic, diameter of 5 to 10 cm, topped by follicular papules with thorny spicules which detached easily, on the cervical region, back, abdomen and flexural creases (axillae, antecubital and popliteal fossae) (Figures 1-3). Organomegaly was not found and the patient was previously healthy.

Histopathology showed mucin inside the follicular epithelium, detected by HE coloration and Coloidal Iron, with disconnection of keratinocytes, perivascular and periannexal inflammatory infiltrate and without atypical lymphocytes. (Figures 4 e 5).

The exams for investigation of associated systemic diseases (blood count, renal function, liver function, thyroid function, anti-nuclear factor) were normal. Lesions improved spontaneously as the patient used only a cream with 5% urea prescribed at his first visit.

DISCUSSION

FM is a rare condition, of unknown cause, which affects all races, ages and both sexes.[3]

FM is considered a reaction pattern to several conditions, being either idiopathic or secondary to benign diseases (lupus erythematosus, insect bites, eczema, alopecia areata, hypertrophic lichen planus) or malignant (mycosis fungoides and Sezary syndrome, leukemia cutis, cutaneous B-cell lymphoma and Hodgkin's disease).[2]

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Figure 1: Erythematous, hypochromic, well-delimited, alopecic macules on the back and cervical region

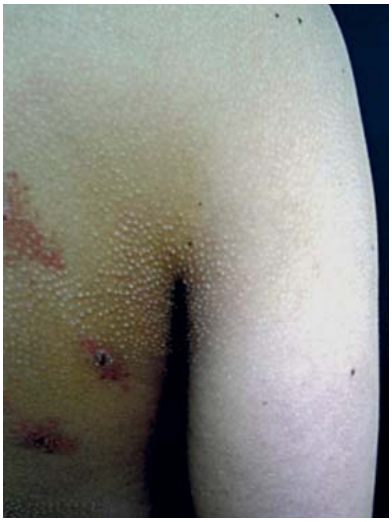

Figure 2: Pinkish, well-delimited plaques, topped by follicular papules, with thorny spicules

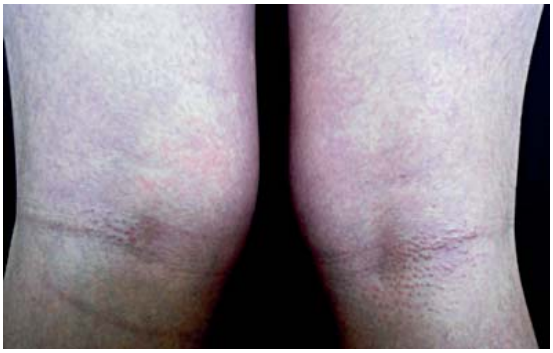

Figure 3: Pinkish, well-delimited plaques, topped by follicular papules, with thorny spicules

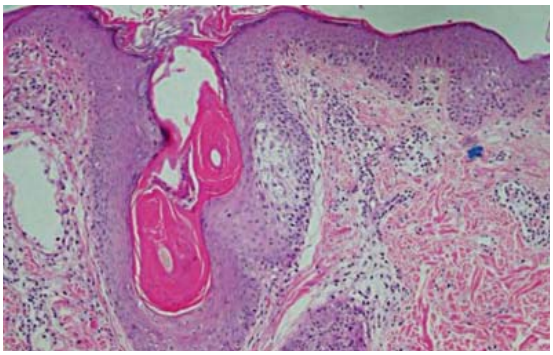

Figure 4: 100x -HE. Follicular wall area, clear, with mucin deposition

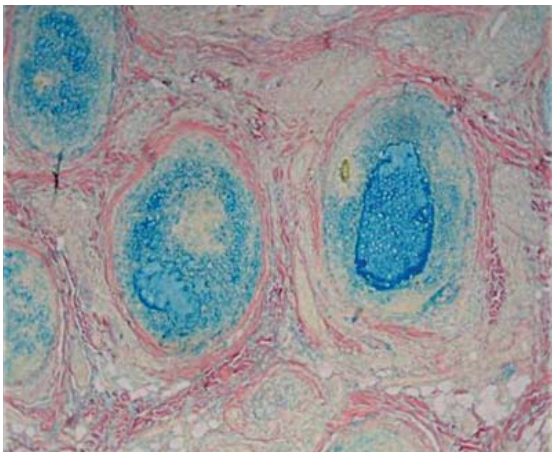

Figure 5: 40x -Colloidal iron. Three follicles with mucin deposition on wall

The association of FM with the development of lymphoproliferative diseases is well documented and association with mycosis fungoides and its variants is more frequent.[1,3] Around 15 to 30% of FM patients will develop MF.[4] Some authors believe that FM is an indolent form of MF with an usually favorable prognosis, although its exact course cannot be predicted.[5]

The morphology of lesions on idiopathic and secondary forms is identical. Lesions present as single or multiple plaques, usually erythematous and alopecic, well delimited, discretely elevated, sometimes scaly and on top of them there may be follicular papules. The follicular ostia may be dilated or filled with keratin and the pressure of lesion cause the exit of mucin. However, clinical manifestation may not be typical and there are reports of nodulary forms and presentations that simulate folliculitis, alopecia areata, scarring alopecia, chronic eczema, acne, urticaria and erythrodermic forms.[3]

Histopathology is essential for the diagnosis, showing mucin accumulation at the hair follicle external root sheath and sebaceous gland, besides inflam-

matory infiltrate of lymphocytes, macrophages and eosinophils with follicle tropism of lymphocytes.[1]

The onset of MF might precede, occur concomitantly or years after the diagnosis of FM. In almost all cases in which there is development of MF this process happens within 5 years, one year on average.[6] There are however a few reports where lymphoma onset was late, occurring 15 years after the FM diagnosis.[4,5]

Patients are classified into two phenotypes with different evolution. It is accepted that when FM occurs in adults older than 40 years with infiltrative, persistent and diffuse lesions there is more serious risk of progression to MF; nevertheless, cumulative experience shows that the age of the patient, distribution of lesions, duration and extension of the disease do not distinguish primary benign FM from FM secondary to mycosis fungoides in a reliable way.[4] In addition to this distinction not being absolute, several patients, as reported, do not fit into these two groups.

Despite primary forms occurring at a lower age than those associated with lymphomas, in a study 62% of patients older than 40 did not develop lymphomas after an average 10-year follow-up and 23% of patients with FM between 30 and 40 years of age ended up developing MF.[2,5,6] Furthermore, MF and Hodgkin's disease were associated with FM in patients under 20 years of age.[4]

Location has not been a precise criterion either, 44% of patients with FM lesions limited to head and trunk presented MF whereas 42% of patients with FM lesions on trunk or limbs did not present progression to lymphoma.[4]

In contrast, the presence of a single lesion is a strong indication of primary FM; the development of lymphomas in single lesions, even though possible, is not expected.[2,5]

Something similar occurs in histological evaluation of these cases.[2] In the absence of clear criteria to confirm the diagnosis of lymphoma there is no way to know if there will be evolution to lymphoma or not.[2,4,6]

There is no standard conduct for the treatment of FM and it is possible to adopt an expectant management (watchful waiting) in cases of primary FM, since many of them resolve spontaneously between 2 and 24 months. Several therapeutic modalities have been reported with variable results: topical corticoids, intralesional or oral, topical retinoids, oral isotretinoin, dapsone, indomethacin, minocycline, PUVA, cyclophosphamide and methotrexate, but no drug has a consistent result and the evaluation is difficult since there may be spontaneous involution.[7,8,9] Treatment of secondary form is the treatment of the associated disorder.

The patient with idiopathic FM must be oriented regarding the necessity of a follow-up for the early detection of alterations signs secondary to malignancies. Such orientation is mandatory, and also suggested is patient follow-up for a minimal period of 5 years.[4] Our patient remains under clinical follow-up with improvement of the aspect of the lesions, although without complete remission up to this moment. The joint family / doctors option was for not doing other forms of treatment. ❑

REFERENCES

1. Zvulunov A, Shkalim V, Ben-Amitai D, Feinmesser M. Clinical and histopathologic spectrum of alopecia mucinosa/follicular mucinosis and its natural history in children. J Am Acad Dermatol. 2012;67:1174-81.
2. Rongioletti F, De Lucchi S, Meyes D, Mora M, Rebora A, Zupo S, et al. Follicular mucinosis: a clinico pathologic, histochemical, immunohistochemical and molecular study comparing the primary benign form and the mycosis fungoides - associated follicular mucinosis. J Cutan Pathol. 2010;37:15-9.
3. Fonseca APM, Campelo FS, Bona AH, Rêgo PMM, Fonseca WSM. Follicular mucinosis: literature review and case report. An Bras Dermatol 2002;77:701-6
4. Brown HA, Gibson LE, Pujol RM, Lust JA, Pittelkow MR. Primary follicular mucinosis: Long-term follow up of patients younger than 40 years with and without clonal T-cell receptor gene rearrangement. J Am Acad Dermatol. 2002;47:856-62.
5. Cerroni L, Fink-Puches R, Bäck B, Kerl H. Follicular Mucinosis. A Critical Reappraisal of Clinicopathologic Features and Association With Mycosis Fungoides and Sézary Syndrome. Arch Dermatol. 2002;138:182-9.
6. Gibson LE, Muller SA, Leiferman KM, Peters MS. Follicular mucinosis: clinical and histopathologic study. J Am Acad Dermatol. 1989;20:441-6.
7. Parker SR, Murad E. Follicular mucinosis: clinical, histologic, and molecular remission with minocycline. J Am Acad Dermatol. 2010;62:139-41.
8. Kim KR, Lee JY, Kim MK, Yoon TY. Successful treatment of recalcitrant primary follicular mucinosis with indomethacin and low-dose intralesional interferon alpha. Ann Dermatol. 2009;21:285-7.
9. Schneider SW, Metze D, Bonsmann G. Treatment of so-called idiopathic follicular mucinosis with hydroxychloroquine. Br J Dermatol. 2010;163:420-3.

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