



Milia en plaque

Milia em placa

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Abstract: Milia en plaque is a rare type of primary milia. Issues related to esthetics and differential diagnoses represent a concern to patients and a challenge for physicians. In this paper, a case of milia en plaque is reported and a review of the literature is described.

Keywords: Epidermal cyst; Pathology; Therapeutics

Resumo: Milia em placa é uma forma rara e primária de milia. Questões referentes à cosmética e aos seus diagnósticos diferenciais perturbam os pacientes e desafiam os médicos. Relatamos um caso de milia em placa e discutimos a literatura pertinente.

Palavras-chave: Cisto epidérmico; Patologia; Terapêutica

INTRODUCTION

Milia are small keratin-filled cysts that result from the obstruction of a hair follicle or eccrine sweat duct. Milia are classified as primary when their onset is spontaneous and secondary when they appear following trauma (such as a burn, dermabrasion or ablative laser resurfacing), inflammatory skin diseases (particularly those involving the basement membrane zone such as subepidermal bullous diseases) or the use of topical medication (topical corticosteroids, 5-fluorouracil) or systemic drugs (cyclosporin and benoxaprofen).^{1,2} Milia en plaque (MEP) is a rare form of primary milia characterized by the presence of milia on an erythematous-edematous or infiltrated erythematous plaque. The condition was first described by Balzer and Fouquet in 1903 and was denominated MEP by Hubler et al. in 1978.¹ Although it is a benign and generally asymptomatic condition, treatment management and clinical and histological differential diagnoses are challenging. The present report describes a case of retroauricular milia en plaque. A review of the literature was also performed.

CASE REPORT

A 32-year old male Caucasian patient from Piracicaba, São Paulo, who had been living in Sorriso in the midwestern state of Mato Grosso, Brazil, presented with an asymptomatic lesion of approximately one year's duration in the left retroauricular region. The patient had been previously healthy and reported no history of having suffered trauma or burns, having been submitted to any medical procedure or having used any topical or systemic medication. He also reported no similar cases in the family.

Physical examination revealed the presence of numerous, whitish-yellow papules of 1-3 mm in diameter on a well-defined, oval, erythematous-edematous plaque measuring approximately 2.5 x 1.3 cm, with the greater axis following the left retroauricular fold (Figure 1).

Saucerization of the lesion was performed for histopathological evaluation. The histological sections of the skin fragment removed revealed various epidermal cysts in the dermis that contained lamellar keratin material. No vellus hairs were seen inside the

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FIGURE 1: : Milia over an oval, erythematous-edematous plaque in the left retroauricular region

cavities. A dense lymphocyte infiltration was also found in the dermis adjacent to the cysts, with vacuolar alteration of the basal layer of the epithelium of the cyst and the presence of lymphocytes among the epithelial cells (Figure 2).

The patient was given a diagnosis of MEP and opted not to undergo any treatment. One year later, the lesion remained unaltered.

DISCUSSION

Milia en plaque (MEP) is a rare form of primary milia characterized by clusters of milia on one or more erythematous-edematous or erythematous, infiltrated plaques with or without comedones.¹⁻³ Around 30 such cases have been reported in the medical literature.³ MEP predominantly affects the periauricular and periorbital regions of adults, with a female/male ratio of 2:1. However, the condition has also been described both in infancy and in senescence and at other sites on the face such as the nose, forehead and in the perimandibular, malar and supraclavicular regions.^{1,4,6}

Evidence shows that the primary forms of milia originate in the hair follicle infundibulum, whereas the secondary forms may originate in various annexal structures such as the sweat ducts, sebaceous glands and hair follicles.^{1,7}

Clinical differential diagnosis is made between MEP and secondary milia with plaques and other conditions that may simulate MEP. Cases of secondary milia en plaque have been described in patients with lupus erythematosus, pseudoxanthoma elasticum,

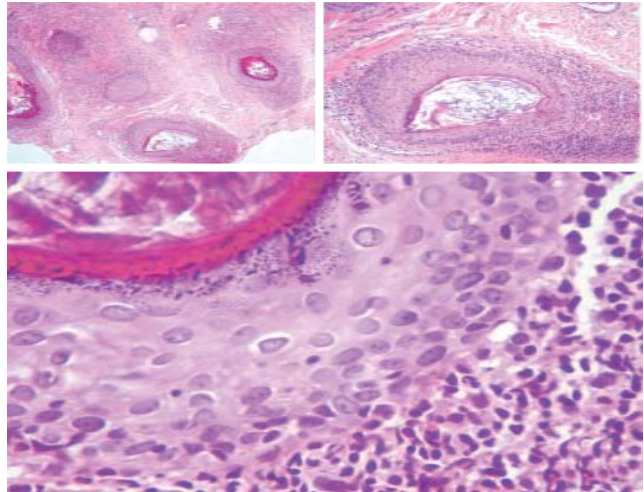


FIGURA 2: A. Photomicrography of the skin showing various epidermal cysts surrounded by a dense inflammatory infiltrate. Hematoxylin and eosin (HE); magnification x4. B. Detail of one of the epidermal cysts containing keratin inside the cavity and dense monomorphonuclear infiltrate in the adjacent dermis. HE; magnification x10. C. Detail of the wall of one of the epidermal cysts with dense lymphocyte infiltration in the adjacent dermis, smudging of the basal layer and the presence of some lymphocytes among the epithelial cells. HE; magnification x40.

lichen planus follicularis tumidus and lichenoid eruption, contact dermatitis, folliculotropic mycosis fungoides with cysts and comedones, follicular mucinosis and in patients in use of cyclosporine following organ transplantation, although doubt remains with respect to this last group.^{1,8-10} Comedone nevus and Favre-Racouchot disease must also be differentiated from MEP. Distinction between primary and secondary milia is based on the presence of a causal factor for milia, as shown by the patient's history, physical examination or histopathology. Clinical examination is sufficient to distinguish MEP from comedone nevus and Favre-Racouchot disease. In the former, onset generally occurs prior to 10 years of age and is characterized by comedones with a nevoid, linear or zosteriform distribution and, in some cases, inflammatory acneiform lesions and scars. On the other hand, Favre-Racouchot disease has a late onset and presents as gigantic comedones associated with cysts, principally at the lower and lateral rims of the periorbital region, with signs of chronic actinic damage. Nevertheless, exclusion of the disorders associated with secondary milia en plaque generally requires histopathological evaluation. Lichen planus follicularis tumidus occurs principally in the retroauricular region, as does MEP, and presents with tumid, violaceous plaques covered by comedones and milia and associated or not with other lesions typical of lichen planus.⁸ The condition is so similar to MEP that it has been suggested that it may represent the final stage of this form of lichen planus.⁹ It has also been speculated that MEP may represent a rare variant of mycosis fungoides or follic-

ular mucinosis; however, histopathological and immunohistochemical evaluation of reported cases has rendered these hypotheses unlikely.⁵ Three cases of milia secondary to lupus erythematosus have been reported in the literature. Lesions generally have a cicatricial appearance that, associated with the milia, is suggestive of this condition. However, in one case of de novo lupus erythematosus, there was a noncicatricial lesion on the chin, reinforcing the need for biopsy.¹¹

In the present case, the epidermal cysts characteristic of MEP were surrounded by lymphocyte infiltration immediately beneath the epithelium of the cyst, showing a lichenoid pattern. This type of inflammation appears to be characteristic of the disease, since it has been reported in various cases of MEP published in the literature.^{1,3,7,11} In some of the reported cases, there was infiltration of lymphocytes in the dermis adjacent to the cyst or confined to the perivascular space.^{4,5} The presence of eosinophils and vellus hair inside the cysts has been described, but was not found in the present case.^{1,3,5} From a histological point of view, differential diagnosis is made with diseases that present with cysts and comedones surrounded by dense lymphocyte infiltration, principally lichen planus follicularis tumidus (LPFT) and folliculotropic mycosis fungoides with cysts and comedones. Microscopic findings are identical in cases of LPFT and MEP, reinforcing the hypothesis that they represent the same disease.^{9,12} Folliculotropic mycosis fungoides with cysts and comedones, on the other hand, is a cutaneous lymphoma, a variant of mycosis fungoides, in which the neoplastic T-lymphocytes are usually found in the epithelium of the hair follicles.^{13,14} Mucin deposits may or may not be present among the epithelial cells and, as is characteristic in cases of mycosis fungoides, the lymphocytes are small and monomorphic in the majority of cases.^{13,14} In the absence of follicular mucinosis and lymphocytes with prominent nuclear atypia, a clinical correlation must be performed to differentiate it from MEP.¹⁵ In cases of ordinary epidermal cysts unrelated to MEP there is no lichenoid infiltration. Chronic granulomatous inflammation of the foreign body type is found when the cysts are ruptured. There are various situations in which epidermal cysts are secondary, as previously

mentioned, and characteristics of the primary disease will be found at skin biopsy. When epidermal cysts are present in lupus erythematosus, for example, alterations typical of this disease such as atrophy and vacuolar alteration of the basal layer with necrotic keratinocytes are generally found in the epidermis in addition to mucin deposit in the reticular dermis.^{2,11} A case of MEP in lichen planus has been reported; however, the epidermis showed typical alterations of the disease such as acanthosis, hypergranulosis, vacuolar alterations and underlying bands of lymphocyte infiltration.⁷

MEP may regress spontaneously; however, it generally remains unchanged if untreated.⁴ Although benign, the lesion's appearance may be distressful to the patient, who may request treatment for this reason. Treatment options include manual extraction, topical tretinoin, etretinate, minocycline, electrocauterization, cryotherapy, dermabrasion, surgical excision and photodynamic therapy.^{1,4} In cases in which histopathology shows the lesions to be superficial, manual extraction and the use of topical tretinoin have been found to be effective. When the milia extend as far as the reticular dermis, procedures capable of accessing these lesions are required in order to obtain results; however, the difficulty in accessing a specific depth has generated varied results.¹ Minocycline has been used successfully in cases with dense inflammatory infiltrate.¹ The advantages of treatment with cryotherapy, electrocauterization and dermabrasion include the low cost of these modalities and the fact that they are commonly used by dermatologists and known to provide good results; however, it should be emphasized that the number of cases that have been treated using these methods is small.^{1,4} Surgical excision may be considered for small lesions.⁴ Photodynamic therapy resulted in a partial focal improvement that was unsatisfactory considering the cost and the results that may be obtained with simpler methods.¹ In view of the few cases described in the literature, the presence of lesions at different depths in the skin, inflammatory infiltrate of varying degrees of intensity and the short follow-up period of the patients treated, no consensus has been reached with respect to the optimal treatment for MEP, and the choice of therapy should be individualized. □

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