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Milia en plaque on the shoulder as an early manifestation of mycosis fungoides

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Abstract

Milia en plaque (MEP) is an uncommon skin condition identified as retroauricular confluent milium by Boulzer and Fouqet in 1903 [1]. It can be mistaken for other dermatoses like Favre-Racouchot nodular elastosis, steatocystoma multiplex, and nevus comedonicus. Milia en plaque can either be primary or secondary and is typically benign, often triggered by dermatological procedures like cryotherapy, as reported in this journal. In some cases, MEP can arise as a secondary manifestation of other diseases, including folliculotropic mycosis fungoides (FMF). Despite this association, there are few documented cases in the literature. Herein, we present a patient in whom MEP served as the initial clinical presentation of FMF; the treatment involved oral retinoids and phototherapy. Furthermore, we highlight distinctive features of both conditions. It is important to emphasize that early diagnosis and treatment of FMF are vital for the patient's quality of life. The presence of MEP can serve as a valuable indicator for identifying it.

Keywords: milia, mycosis fungoides, lymphoma

Introduction

The term milia en plaque (MEP) was originally coined by Boulzer and Fouqet in 1903 [1] and later confirmed by Hubler in 1978 [1]. Histologically, MEP is characterized by several cystic formations filled with keratinous material, arranged along the epidermis and often surrounded by a mononuclear infiltration. These multiple milia are clustered on an erythematous plaque. Milia en plaque can be categorized as either a primary or a secondary manifestation associated with other diseases [1-2].

Case Synopsis

A 42-year-old man with no underlying medical conditions presented with an erythematous plaque on his shoulder that appeared two years prior, along with the recent development of milia points in the affected area (**Figure 1A**). The patient also had pruritus and xerosis. Dermatoscopy revealed pale, sclerotic regions resulting from fibrosis due to follicular damage around yellowish papules resembling milia, with some visible blood vessels (**Figure 1B**). Histopathological examination showed a significant lymphocytic infiltration around hair follicles along with areas showing epidermic lymphocytic exocytosis and hair follicle damage

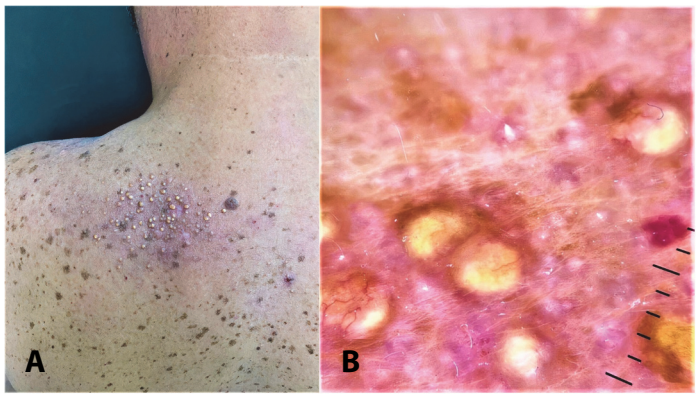

Figure 1. ***A)*** *Milia-en plaque lesion on the shoulder.* ***B)*** *Dermoscopy demonstrating yellow milia dots surrounded by erythematous infiltrate.*

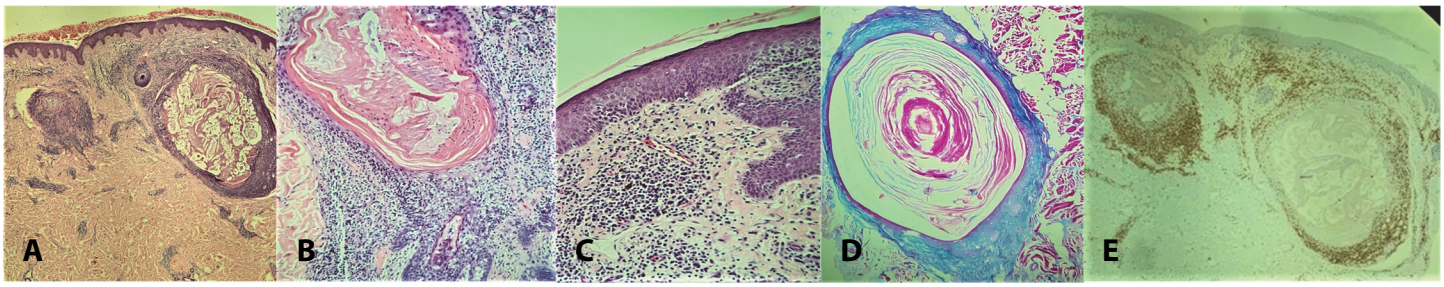

Figure 2. ***A)*** *Intense lymphocytic infiltrate surrounding hair follicles provoking milia.* ***B)*** *The more detailed presence of the perifollicular lymphocytic infiltrate causing milia-like lesions clinically. H&E, 40×.* ***C)*** *Epidermic lymphocytic exocytosis were also observed. H&E, 40×.* ***D)*** *Intense positivity for mucin on colloidal iron coloration, 40×.* ***E)*** *Positive immunofluorescence for CD3, 40×.*

(**Figures 2A-C**). Colloidal iron staining revealed mucin in the dermis (**Figure 2D**). Biopsy and immunohistochemistry confirmed the diagnosis of folliculotropic mycosis fungoides (FMF), showing positive markers for CD4, CD7, and CD3 (**Figure 2E**), but no CD30+ transformation was observed.

Case Discussion

The patient was diagnosed with an early stage of the mycosis fungoides and was prescribed daily doses of 20mg isotretinoin along with UVA phototherapy sessions, leading to significant improvement in the clinical scenario. Diagnosing mycosis fungoides typically proves challenging and time-consuming, often requiring multiple skin biopsies until the characteristic histopathological changes of the disease are observed. Folliculotropic mycosis fungoides represents a more aggressive subtype of classical mycosis fungoides and manifests in various clinical presentations such as plaques, papules, or erythematous tumors. Approximately 81% of patients with FMF experience areas of alopecia and about 30% exhibit acneiform lesions. Milia en plaque lesions can occur in either primary or secondary forms. The primary form lacks a definite cause and usually regresses spontaneously. Moreover, secondary MEP has been linked to other conditions such as discoid lupus, pseudoxanthoma elasticum, amyloidosis, and even follicular hamartomas like trichoadenoma [1-7]. Conversely, secondary MEP has also been associated with radiotherapy, oral cyclosporine use, and certain dermatological procedures like dermabrasion and cryotherapy, as previously described by Beutler et al. in this journal [7]. The characteristics of patients with these two entities are further explained in **Table 1**.

The occurrence of MEP has also been associated with mycosis fungoides regression after treatment. However, there is limited literature highlighting the association between MEP and FMF [1-7]. Notably, the development of MEP-like lesions as a secondary

Table 1. *Some features of milia en plaque and folliculotropic mycosis fungoides.*

Milia en plaque	Folliculotropic mycosis fungoides
Erythematous plaque with milia	Lesional polymorphism, including alopecia areas
Usually an unique lesion	Usually more than one skin lesion
Histologically, multiple cystic structures can be observed at various dermal levels within the dermis. These cysts are lined with a stratified squamous epithelium and are filled with keratinous material. Additionally, they may be surrounded by a mononuclear inflammatory infiltrate	Diverse histopathological aspects including "prototypic" pattern with intact hair follicles, folliculotropism with or without follicular mucinosis, basaloid folliculolymphoid hyperplasia with folliculotropism, granulomatous dermatitis associated with a follicular destruction, eosinophilic folliculitis, or follicular cysts with folliculotropism
Benign in most cases or secondary to underlying disease	Malignant condition
Usually acute	Usually chronic
Spontaneous regression is possible, and treatment options may include laser sessions or the application of retinoids	Typically, there is no definitive cure for the condition, and patients may experience periods of remission achieved through the administration of immunosuppressants and phototherapy
Usually asymptomatic	Usually pruritic

effect of an underlying disease plays a role in the disease progression or remission.

Conclusion

This article reviews various characteristics of FMF and MEP, emphasizing the significance of understanding the link between these two dermatological conditions. It also emphasizes the importance of early diagnosis and prompt treatment of FMF.

Potential conflicts of interest

The authors declare no conflicts of interest.

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