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Periorbital eruptive vellus hair cyst: a challenging treatment in an atypical location

Cisto veloso eruptivo periorbital: tratamento desafiador em localização atípica

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ABSTRACT

Eruptive vellus hair cyst is a rare condition resulting from the abnormal development of vellus hair follicles, leading to the formation of multiple follicular papules. While it can appear on the face, only five cases specifically located in the periorbital region have been described in the literature. Its treatment is challenging, particularly in the case we report here due to the peculiar location of the lesions, which increases the risk of postoperative scarring and dyschromia. We present a report that addresses the challenging diagnosis and successful treatment of a rare case of eruptive vellus hair cyst located in the periorbital region.

Keywords: Follicular Cyst; Eyelids; Surgical Procedures, Operative.

RESUMO

O cisto veloso eruptivo é uma entidade rara decorrente do desenvolvimento anormal dos folículos vellus, que se manifesta na forma de múltiplas pápulas foliculares. Eventualmente, pode localizar-se na face, sendo descritos na literatura apenas cinco casos na região periorbital. Seu tratamento é desafiador, especialmente neste relato de caso, devido à peculiaridade da localização das lesões, com risco de cicatriz e discromia pós-procedimento. Relata-se o diagnóstico desafiador e o tratamento satisfatório de um caso raro de cisto veloso eruptivo localizado na região periorbital.

Palavras-chave: *Cisto folicular; Pálpebras; Procedimentos cirúrgicos menores*

Case report

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INTRODUCTION

Eruptive vellus hair cyst (EVHC), a rare condition first described in 1977,[1] arises from the abnormal development of vellus hair follicles at the infundibular level, leading to retention of hairs, cystic dilation of the proximal part of the follicle, and secondarily, atrophy of hair bulbs.[2,3] EVHC is most commonly observed in children and young adults, with no sex or race predilection. Its pathogenesis remains unknown, and it may be associated with either the sporadic form of the disease or autosomal dominant inheritance with incomplete penetrance,[2,3] based on numerous reports of families in which two or more members are affected.[4] Typically, these lesions are asymptomatic but may sometimes be associated with mild pruritus or altered sensitivity. Occasionally, they can appear on the face.[2] Treatment can be challenging, and currently, there is no consensus on the optimal treatment. We report the case of a patient with EVHC located on the eyelids, which, due to its unusual location, was initially misdiagnosed as milia during childhood. This case report aims to illustrate and demonstrate a treatment approach that resulted in a satisfactory aesthetic outcome.

METHODS

A 19-year-old male patient with no known comorbidities presented with asymptomatic normochromic and hypochromic papules of firm consistency on the upper and lower eyelids, present since the age of 6 years. There was no family history of similar lesions. Previous medical consultations resulted in a diagnosis of milia, and topical exfoliants were prescribed without improvement. Due to the progressive increase in the number of lesions, eventually forming clustered papules (Figure 1), an excisional biopsy was performed. Histopathological analysis revealed cysts lined by keratinized squamous epithelium filled with laminated keratin and occasional vellus hair shafts (Figure 2), confirming the

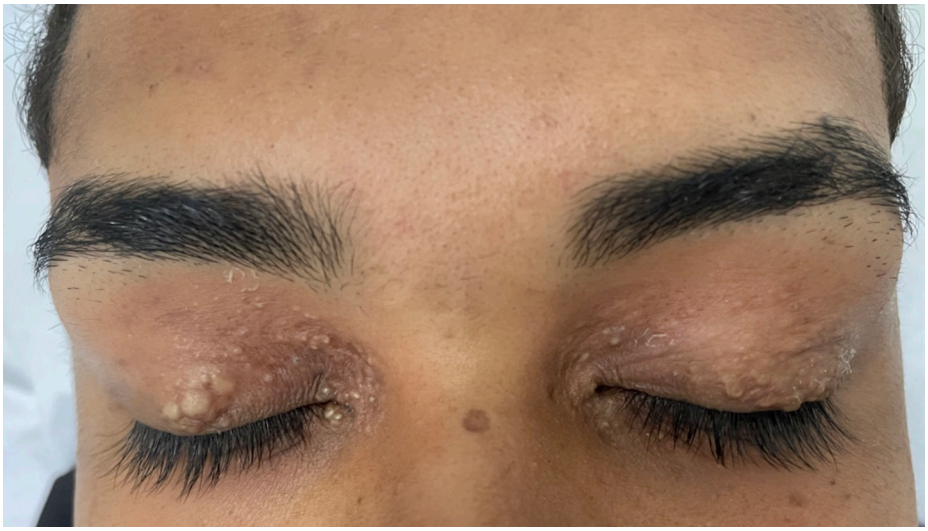

Figure 1: Normochromic and hypochromic papules of firm consistency, some clustered, located on the upper and lower eyelids

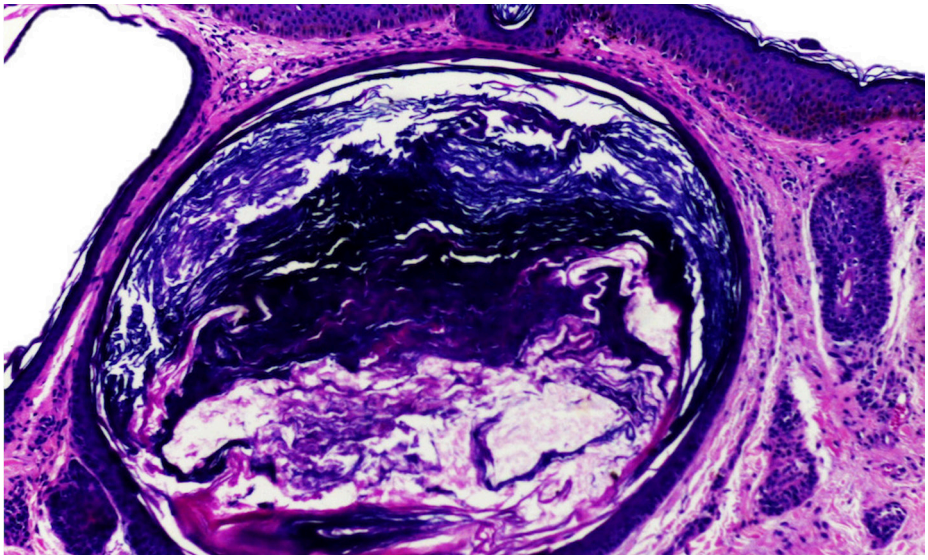

Figure 2: Anatomopathologic examination: cyst located in the dermis lined by keratinized squamous epithelium filled with laminated keratin and vellus hair shafts (HE, 100x)

diagnosis of EVHC. Given the patient's significant aesthetic discomfort, we opted for needle evacuation in some lesions using a 30G needle. To minimize the risk of post-inflammatory hyperchromia, the patient's skin was prepared topically for 14 days with Kligman's formula (hydroquinone 40 mg/g + tretinoin 0.5 mg/g + fluocinolone acetonide 0.1 mg/g cream) prior to the procedure.

Description of the technique:

Infiltrative anesthesia using 2% lidocaine with vasoconstrictor.

Incision of the lesions with a 30G needle (Figure 3a).

Needle evacuation using a 30G needle or manual drainage (Figure 3b).

Hemostasis achieved with local compression using gauze.

Local cleaning with a 0.9% saline solution.

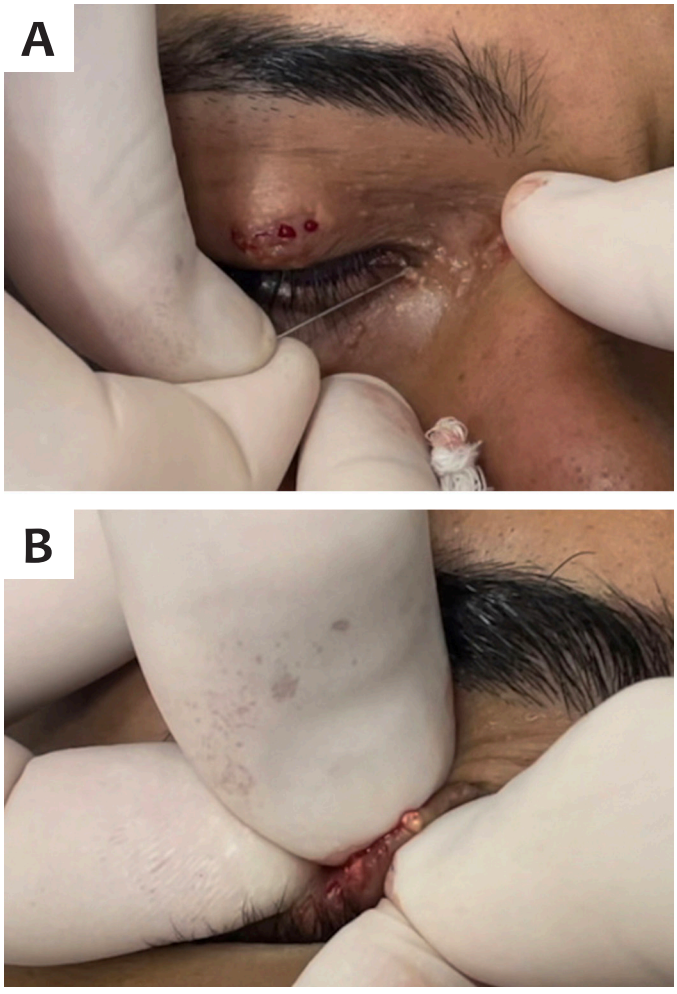

Figure 3: A - Incision of lesions with a 30G needle
B - Manual drainage of lesion contents

RESULTS

The patient experienced mild edema in the first postoperative days. Two months post-procedure, the patient has shown good healing and achieved a satisfactory aesthetic outcome, with no recurrence of the lesions to date (Figure 4).

DISCUSSION

EVHC manifests clinically as multiple follicular papules, flat or dome-shaped, ranging from 1 to 5 mm in diameter, characterized by a smooth surface and firm consistency, with variations of color (erythematous, normochromic, hyperchromic, or even whitish and yellowish). The papules may also exhibit a central hyperkeratotic crust and umbilication.[2,3] While these lesions are mostly located on the thorax and upper and lower extremities, they can occasionally affect the abdomen, neck, axillae, groins, and face.[2] This distribution is unlikely to be random and appears to overlap with that of pilosebaceous and apocrine units.[4] Given its clinical resemblance to other conditions, the definitive diagnosis of EVHC relies on anatomopathologic examination. In the case reported here, this clinical similarity led to misdiagnosis and a prolonged treatment for milia.[2,3,5] In addition to milia, the differential diagnosis includes molluscum contagiosum, keratosis pilaris, steatocystoma multiplex, folliculitis, multiple adnexal tumors, and syringomas.[2,3,5] Histopathological analysis of EVHC typically reveals its presence in the middle or upper dermis, lined by multiple layers of keratinized stratified squamous epithelium (up to 12 layers). The cyst's contents consist of varying amounts of laminated keratin, along with numerous vellus hairs that are cut both obliquely and transversely, causing occlusion and dilation of the follicular unit. Usually, no sebaceous gland is found in the cyst wall.[2,3,5] EVHC in the periorbital region, as reported in this case, is rare, with only five cases described in the literature.[4-7] While treatment approaches for EVHC are not well-established, it is estimated that spontaneous remission occurs in approxima-

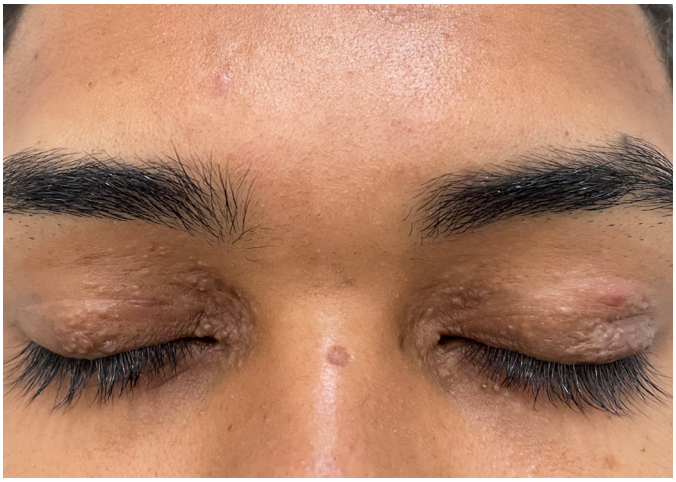

Figure 4: Reduction in size and number of lesions after needle extraction

tely 25% of cases by transepidermal elimination or destruction secondary to the local inflammatory process.[2,3,5] Currently, there is no universally accepted treatment. Reports mention the use of lactic acid and retinoids (tretinoin and isotretinoin), but their results are still insufficient.[2,3] Dermabrasion, erbium:YAG laser, carbon dioxide laser vaporization, and needle evacuation have also been noted as potential treatments. However, these methods have limitations, including the risk of scarring and the high rate of early recurrence.[2,3,8]

CONCLUSION

We report this case of periorbital EVHC due to its rarity, particularly in this atypical location. Furthermore, this report aims to increase awareness of this often underdiagnosed condition, as its primary differential diagnoses correspond to more common entities. Although benign, EVHC can cause considerable aesthetic concerns and even psychological distress for patients, making appropriate and effective treatment essential. Therefore, despite the challenges in treating this condition, the satisfactory response to the approach used in this case is noteworthy. ●

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