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Eruptive Vellus Hair Cysts: Report of a Pediatric Case With Partial Response to Calcipotriene Therapy

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Eruptive vellus hair cysts (EVHCs) are characterized by asymptomatic, follicular, comedonelike papules usually located on the anterior chest and abdomen. We present a pediatric case of EVHC associated with attention deficit hyperactivity disorder that partially responded to calcipotriene cream within 2 months. Our aim is to refamiliarize clinicians with a common albeit frequently unrecognized disorder of vellus hair follicles.

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Eruptive vellus hair cysts (EVHCs) were initially described by Esterly et al[1] in 1977. Although the disorder is relatively common, it rarely has been reported in the literature.

Case Report

A 6-year-old boy presented with a 5-year history of numerous asymptomatic papules on the chest and abdomen. He had received several topical remedies for a presumptive diagnosis of acne vulgaris. The medical history was notable for attention deficit hyperactivity disorder. There was no history of EVHC, steatocystoma multiplex (SCM), or pachyonychia congenita in the family.

On dermatologic examination, there were multiple follicular, comedonelike, yellow-brown, 1- to 3-mm papules with smooth surface scattered on the chest and abdomen (Figure 1). Histopathologic examination of a punch biopsy specimen revealed an oval cystic structure in the superficial dermis lined with a thin, stratified, squamous epithelium of 2 to 3 layers (Figure 2). The cyst wall was noted to include a granular layer at focal areas. The cyst lumen contained lamellate keratin flakes and nonpigmented vellus-type hairs (Figure 3). In serial sections, there was no evidence of sebaceous glands within the cyst wall.

The patient was diagnosed with EVHC, and twice daily calcipotriene cream 0.005% was prescribed for the lesions. At the end of the second month of follow-up, a partial response was attained, with complete resolution of some cysts and flattening of the remaining lesions (Figure 4).

Comment

Eruptive vellus hair cysts represent a sporadic disorder of vellus hair follicles.[2,3] Familial cases are rare and suggest an autosomal dominant inheritance.[2,4-6] There is no racial predilection and both sexes are equally affected.[2,5] The age of onset ranges from 2 to 64 years, and both congenital and pediatric cases have been documented.[1-3,7,8] In our patient, the disease was sporadic and the age of onset was 1 year.

Currently, the etiopathogenesis of EVHC is based on various theories. One theory attests that faulty development of the infundibulum of hair follicles results in keratotic occlusion at this level. Thereafter, keratin and vellus hair shafts accumulate, with subsequent cystic dilatation of the infundibulum and loss of connection with the overlying epidermis.[1,2,7,9,10] An alternative theory postulates that EVHC is a hamartoma of the pilosebaceous unit with the potential to differentiate toward the vellus hair matrix.[4,9,10] This theory may explain

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The authors report no conflict of interest.

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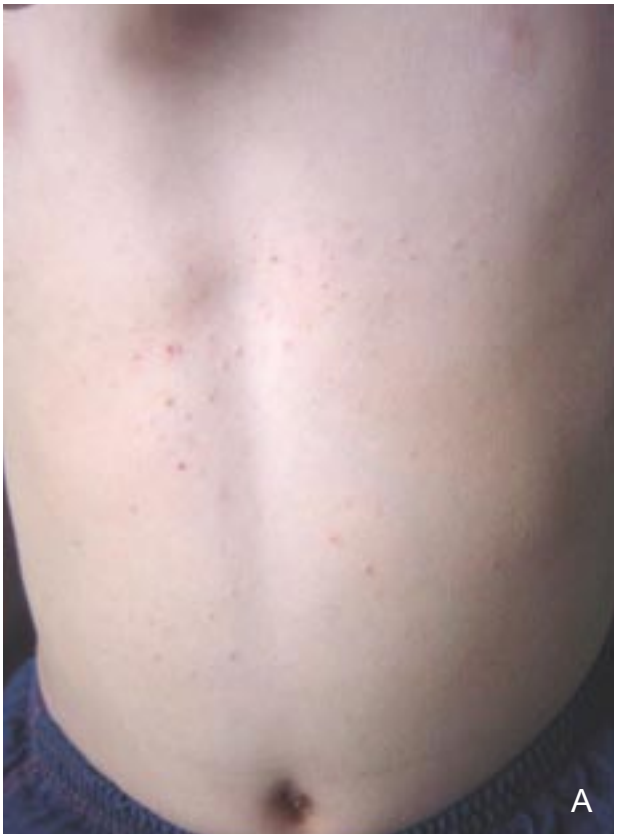

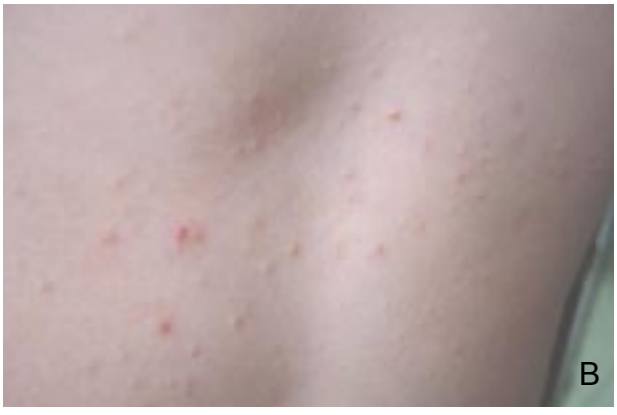

Figure 1. Follicular, comedonelike, yellow-brown papules on the chest and abdomen (A and B).

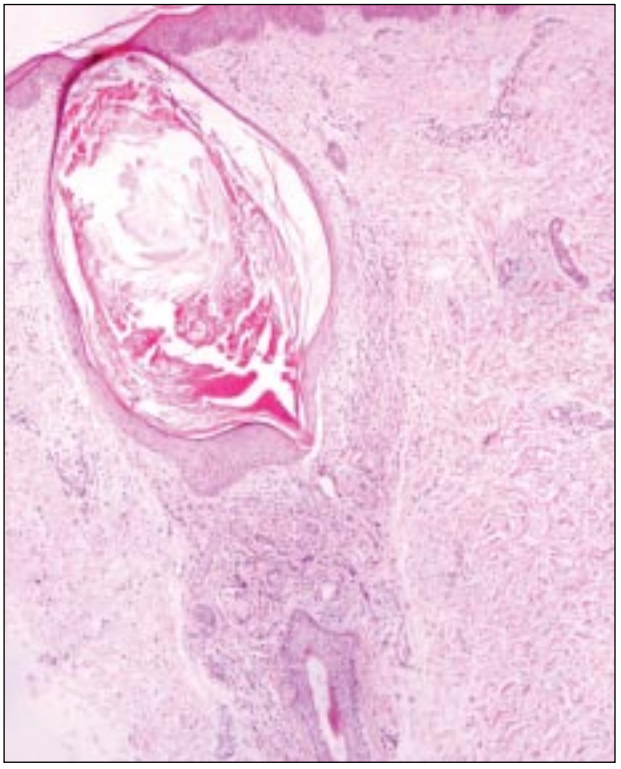

Figure 2. An oval cystic structure in the superficial dermis lined with a thin, stratified, squamous epithelium of 2 to 3 layers (H&E, original magnification ×40).

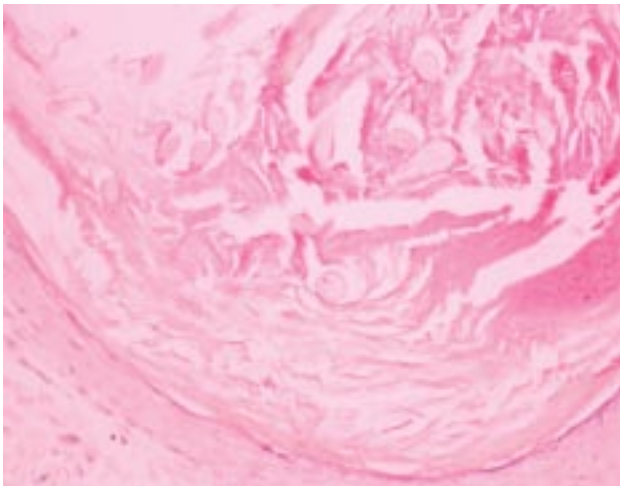

Figure 3. Lamellate keratin flakes and nonpigmented vellus-type hairs within the cyst lumen (H&E, original magnification ×200).

cases with congenital or early onset. A final theory aims to clarify acquired and late-onset EVHC and advocates proliferation of ductal follicular keratinocytes (ductal occlusion) or loss of perifollicular elastic fibers (follicular collapse) prompted by exogenous factors.[7,10] UV rays, mechanical trauma, and scratching are assumed to trigger these pathogenic mechanisms.[9,10] The child presented herein had a history of frequent trauma due to attention deficit hyperactivity disorder. We believe that trauma might represent an incipient trigger in EVHC and leads to the formation and/or progression of these lesions, at least in patients inherently susceptible to the disorder.

Clinically, EVHC presents with multiple, 1- to 5-mm, dome-shaped, comedonelike, discrete or clustered follicular papules.[1,2,5,7-9,11] The lesions have a tendency for localization on the chest and

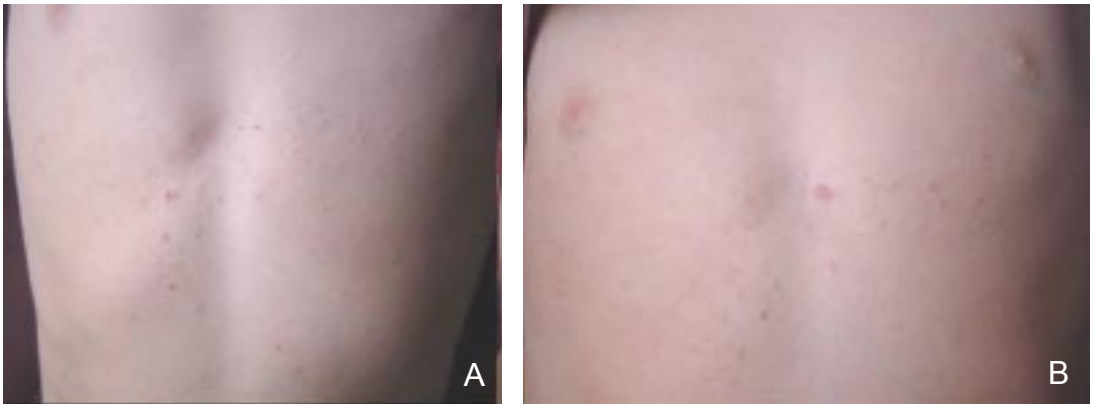

Figure 4. Appearance of the lesions before (A) and after 2 months of treatment with calcipotriene cream 0.005% (B). There was complete resolution of some cysts and flattening of the remaining lesions. The red macule is a former biopsy site.

abdomen, though the extremities, buttocks, back, axillae, groin, neck, face, and eyelids also may be involved.[1,2,4,5,9-12] Lesions may vary in number from 20 to 200.[4,8] The color of the lesions may be diverse, including flesh colored, white, yellow, blue, brown, red, and black.[2,4-6,8,11,12] The papules usually have a smooth surface; however, crusting, umbilication, or central puncta may be noted.[2,5,8,9] A yellow-white, cheesy, sticky material with offensive odor may be expressed upon incision of the lesions.[9] The disorder usually is asymptomatic; pruritus and tenderness are exceptional concerns.[5] Spontaneous regression, experienced by one-fourth of patients, has been attributed to transepidermal elimination and formation of foreign body granulomas by cyst rupture.[4-6]

Methodologies that may aid the diagnosis include incision and expression of cyst contents under local anesthesia, aspiration of cyst contents with an 18-gauge injector, and biopsy.[8,10,13] Histology of EVHC is characterized by round to oval cystic structures in the superficial or mid dermis, encircling keratin flakes, and nonpigmented nonmedullated vellus-type hairs.[2-5,7,9,12] Vellus hairs are transverse or obliquely cut and doubly refract under polarized light.[4,7] The cyst wall is composed of a stratified squamous epithelium. The granular layer may be observed within the cyst wall.[5]

Steatocystoma multiplex constitutes the main differential diagnosis.[2,5,12,14] Clinically, EVHC and SCM are similar in terms of inheritance, age of onset, appearance of lesions, and sites of predilection.[5,12,14] Histologically, in contrast to SCM, EVHC lacks sebaceous glands within the vicinity of the cyst wall.[5,9] Another histologic clue for differentiation between these disorders is the staining of the cyst wall with the protein keratin 17 only in EVHC and with both keratin 10 and keratin 17 in SCM.[5,9,11,13] However, hybrid cysts with histologic features of both EVHC and SCM have been reported.[3,5,12,14,15] In addition, biopsy specimens from different lesions of the same patient may display morphologies consistent with either EVHC or SCM[3,12,14,15]; thus, the term *multiple pilosebaceous cysts* has been coined.[12,14] Some reports suggest that EVHC and SCM are interrelated and represent variant entities within the spectrum of pilosebaceous unit disorders.[3,5,9,11,12,15]

Although it is generally observed as an isolated phenomenon, EVHC has been reported in association with several genodermatoses. The most notable association is pachyonychia congenita.[7,9,11,16] Hidrotic and anhidrotic ectodermal dysplasia, trichostasis spinulosa, and Lowe syndrome also have been described in conjunction with EVHC.[5,7,9,11]

The treatment of EVHC is a challenge. Several modalities have been documented in the literature, including abrasion and application of urea 10% in a moisturizer or lotion formulation, salicylic acid, lactic acid lotion 12%, tretinoin, and adapalene gel 0.1%; oral vitamin A; oral retinoids; incision and expression of cyst contents; curettage; cryotherapy; dermabrasion; surgical excision; and CO_2 or erbium:YAG lasers.[4,5,17,18] Surgical approaches may be time consuming and often disappointing because of early recurrences or complications such as atrophy, pigmentation, and scarring.[5,6,13] Our patient partially responded to calcipotriene within 2 months. Some of the lesions completely resolved with this therapy and the remaining lesions revealed flattening. The efficacy of calcipotriene cream in EVHC could be ascribed to its antiproliferative and prodifferentiating effects on ductal follicular keratinocytes. However, the response was partial and spontaneous resolution could not be discounted in our case.

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