Eruptive Vellus Hair Cysts in Identical Triplets With Dermoscopic Findings

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PRACTICE POINTS
- Eruptive vellus hair cysts (EVHCs) are 1- to 3-mm round, dome-shaped, flesh-colored, asymptomatic, benign papules typically occurring on the chest and extremities.
- Pathogenesis and inheritance are unclear. Although the majority of EVHC cases are sporadic, the strong influence of genes is indicated by numerous reports of families in whom 2 or more members were affected.
- Dermoscopy is a noninvasive diagnostic procedure that should be utilized to diagnose EVHCs in the pediatric population; specifically, EVHCs exhibit light yellow, homogenous, circular structures with a maroon or erythematous halo.
- The main indication for treatment of EVHCs is cosmetic concern; however, one-quarter of cases may resolve spontaneously.

Eruptive vellus hair cysts (EVHCs) are asymptomatic, follicular, flesh-colored to hyperpigmented papules that are typically located on the chest and extremities with an unclear inheritance pattern and pathogenesis. We report a case of EVHCs in 4-year-old identical triplet girls. Our aim is to present another rare case of autosomal-dominant inheritance of EVHCs and to emphasize the utility of dermoscopy as a diagnostic aid for EVHCs that may be misdiagnosed clinically.

Case Report
Four-year-old identical triplet girls with numerous asymptomatic scattered papules on the chest of 4 months' duration were referred to a dermatologist by their pediatrician for molluscum contagiosum. The patients' father reported that there was no history of trauma, irritation, or manipulation to the affected area. Their medical history was notable for prematurity at 32 weeks' gestation and congenital dermal melanocytosis. Family history was notable for their father having acne and similar papules on the chest during adolescence that resolved with isotretinoin therapy.

On physical examination there were multiple smooth, hyperpigmented to erythematous, comedonal, 1- to 2-mm papules dispersed on the anterior central chest of all 3 patients (Figure 1). Clinically, these lesions were fairly indistinguishable from other common dermatologic conditions such as acne or milia. Dermoscopic examination revealed homogenous yellow-white areas surrounded by light brown to erythematous halos (Figure 2). Histopathologic examination was not performed given the benign clinical diagnosis and avoidance of biopsy in pediatric populations. Based on dermoscopic features and history, a diagnosis of eruptive vellus hair cysts (EVHCs) in identical triplets was made.

Comment
Pathogenesis—Eruptive vellus hair cysts, first introduced by Esterly et al in 1977, are uncommon benign lesions presumed to be caused by an abnormal development of the infundibular portion of the hair follicle. They are usually 1- to 3-mm, reddish brown, monomorphous papules overlapping with pilosebaceous and apocrine units. Although the lesions typically are located on the chest and extremities, they may occur on the face, abdomen, axillae, buttocks, or genital area. The inheritance of EVHCs is unclear. The majority of reported cases are sporadic; however, the literature mentions 19 families affected by autosomal-dominant EVHCs based on phyllostrophy. In 2015, EVHCs were reported in identical twins, further supporting the case for a genetic mutation. We augment this autosomal-dominant inheritance pattern...
by presenting a case of identical triplets with EVHCs. The patients’ father reported similar lesions in childhood, further underscoring a genetic basis.

The pathogenesis of EVHC is uncertain, with 2 main theories. Some propose retention of vellus hair and keratin in a cavity formed by an abnormal vellus hair follicle causing infundibular occlusion. Others consider the growth of benign follicular hamartomas that differentiate to become vellus hairs.1

Clinical Presentation—The sporadic form of EVHCs is noted to be more common and clinically presents later, with an average age at onset of 16 years and an average age at diagnosis of 24 years.3 The sporadic form occurs without trauma or manipulation as a precursor. Less
commonly, lesions present at birth or in early infancy and may show an autosomal-dominant inheritance pattern with a similar distribution across relatives.3

Other variants of EVHCs have been described. Late-onset EVHC usually occurs at 35 years or older (average age, 57 years), with a female to male predominance of 2.5 to 1.2 This late onset may be attributed to proliferation of ductal follicular keratinocytes or loss of perifollicular elastic fibers exacerbated by exogenous factors such as manipulation, UV rays, or trauma.5

For unilational EVHC, the average age at diagnosis is 27 years.3 Some of these lesions may be pedunculated or greater than 8 mm. There is a female to male predominance of 2 to 1. Eruptive vellus hair cysts with steatocystoma multiplex can be seen with an average age at onset of 19 years and a female to male predominance of 0.2 to 1. There may be a family history of this subset, as reported in 3 patients with this pattern.2

Diagnosis—The recommended workup for EVHCs varies by patient and age. Eruptive vellus hair cysts present an opportunity to utilize noninvasive diagnostic procedures, especially for the pediatric population, to avoid scarring and pain from manipulation or biopsies. Although many practitioners may comfortably diagnose EVHCs clinically, 6 cases were misdiagnosed as steatocystoma multiplex, keratosis pilaris, or milia prior to histopathology revealing vellus hair cysts.6

Dermoscope presents as a useful diagnostic aid. Eruptive vellus hair cysts exhibit light yellow homogenous circular structures with a maroon or erythematous halo.2,7 A central gray-blue color point may be seen due to melanin in the pigmented hair shaft.7 A dermoscopy review of EVHCs reported radiating capillaries. Occasionally, nonfollicular homogenous blue pigmentation may be seen due to a connection to atrophic hair follicles in the mid dermis and no normal hair follicle around the cysts.8 In comparison, dermoscopic characteristics of molluscum contagiosum demonstrated a polylobular, white-yellow, amorphous structure at the center with a hardened central umbilicated core and a crown of hairpin vessels at the periphery. Additionally, comedonal acne, commonly mistaken for EVHCs, reveals a brown-yellow hard central plug with sparse inflammation under dermoscopy.2 Thus, differentiation of these entities with dermoscopy should be highly prioritized to better aid in the diagnosis of pediatric dermatologic conditions using painless noninvasive techniques.

Treatment—The main indication for treatment of EVHCs is cosmetic concern. Twenty-five percent of EVHCs spontaneously resolve with transepidermal hair elimination or a granulomatous reaction.4,5 A case report of 4 siblings with congenital EVHCs also described a mother with similar lesions that resolved spontaneously in early adulthood,3 as our patients’ father also noted. Treatment modalities including topical keratolytic agents such as urea 10%, retinoic acid 0.05%, tazarotene cream 0.1%, and lactic acid 12%; incision and drainage; CO2 laser; or erbium-doped YAG laser ablation have been tried with minimal improvement.5 Of note, tazarotene cream 0.1% has demonstrated better results than both erbium-doped YAG laser and drainage and incision of EVHCs.4 Additionally, another report evidenced partial improvement with calcipotriene within 2 months with some lesions completely resolved and others flattened, which may be attributed to the antiproliferative and prodifferentiating effects on the ductal follicular keratinocytes by calcipotriene.5 Lastly, an additional study indicated that isotretinoin and vitamin A derivatives were ineffective for clearing EVHCs.6

Conclusion
We presented 3 identical triplets with the classic pediatric onset and dermoscopic findings of EVHCs on the trunk. Although the definitive diagnosis of EVHCs relies on histopathology, we argue that their unique dermoscopic findings combined with a thorough clinical examination is sufficient to recognize this benign condition and avoid painful procedures in the pediatric population.

REFERENCES