

A rare case of Eruptive Vellus Hair Cysts: facial variant

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Abstract Eruptive vellus hair cysts are rare benign tumors of pilosebaceous unit with vellus hairs, which appear in childhood or young adults. Their etio-pathogenesis is poorly known. A review of the literature shows that eruptive vellus hair cysts (EVHC) might be inherited as autosomal dominant or acquired, and might be associated with other genodermatoses like pachyonychia congenita and anhidrotic ectodermal dysplasia. Although it is a benign tumor of pilosebaceous unit, this condition is rather difficult to treat. Here we report a case of 23 years old female with multiple, asymptomatic, slate-grey to bluish papules on face and ears for last 5 years, with a positive family history of similar lesions in other siblings. To the best of our knowledge, this is the first case of autosomal dominant facial variant of eruptive vellus hair cysts to be published from Pakistan.

Key words

Eruptive vellus hair cyst (EVHC), pilosebaceous unit, vellus hair.

Introduction

Eruptive vellus hair cyst is an extremely rare and underdiagnosed follicular developmental abnormality of the vellus hair follicles. They are commonly seen in children and young adults, and appear clinically as reddish-brown smooth dome-shaped papules mostly found over the chest, extremities and abdomen.¹ Histological examination of these papules shows dermal cysts which contain small vellus hairs.¹ Eruptive vellus hair cysts were first described by *Esterly and colleagues* in 1977, who reported two cases with symmetrically distributed asymptomatic follicular dome-shaped papules on the chest and extremities.²

Case report

A 23 years old female, lady health visitor by

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profession, presented to the Dermatology Out-patient Department of Sir Ganga Ram Hospital, Lahore with asymptomatic multiple raised lesions on face and ears for last 5 years. There was no history of use of any cosmetic products or excessive sun exposure or exposure to chemicals or toxins before the eruptions. On cutaneous examination, there were numerous grouped, dome-shaped, slate-grey to bluish colored, soft to firm, non-tender 5 to 10mm papules present on the forehead, periorbital area, temples, cheeks and ears bilaterally that gradually increased in number over a period of 5 years and involved most of her face (**Figure 1-3**). The patient was misdiagnosed for comedonal acne and remained on oral retinoids for six months, but no cure. According to the patient, she was operated for a liver cyst when she was seven years old but no record available at present. Her family history was positive of similar lesions in two siblings which also started to appear at teenage. Rest of the cutaneous examination revealed dry xerotic skin with follicular hyperkeratosis over the extensor aspects of upper extremities.



Figure 1



Figure 2



Figure 3



Figure 4



Figure 5 Vellus hair cyst. Low-power view of a mid-dermal cyst

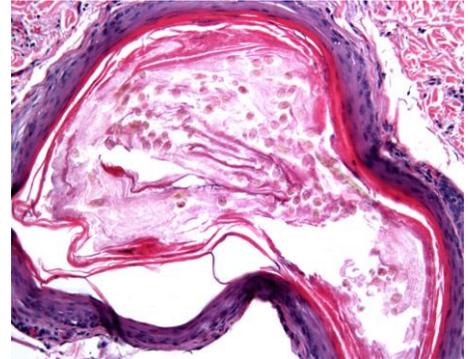


Figure 6 Vellus hair cyst. High-power view showing stratified squamous epithelium and contents consisting of laminated keratin and numerous vellus hairs

The general physical and systemic examination was unremarkable. The patient was thoroughly investigated to look for any other association. Her eye examination revealed degenerative changes in right conjunctiva suggestive of early pterygium formation. Bilateral cup disc ratio was also increased, and this association was not observed in the review of literature. Blood investigations like complete blood count, liver function tests, fasting and random blood sugar, renal function tests and thyroid function tests were all within normal range. CT scan chest, abdomen and pelvis was also unremarkable except for a small follicular cyst noted in right

ovary. Her brother was also examined and similar multiple soft to firm, bluish, dome-shaped papules were present over the forehead and temples (**Figure 4**). His baseline investigations were also within normal range, and general physical and systemic examination was unremarkable. Based on our differentials like giant comedones, fibrofolliculomas, trichodiscomas, steatocystoma multiplex and cutaneous mucinosis, we planned to carry out a lesional skin biopsy from the face of both the patients. Histopathological examination findings were similar in both the siblings and revealed a well circumscribed-cyst in the dermis lined by

stratified squamous epithelium and the lumen was filled with vellus hair and some keratinous material (**Figure 5 & 6**). The wall of the cyst showed no associated adnexal structure. Therefore, based on the above findings, a final diagnosis of eruptive vellus hair cysts; a rare facial variant was made. The patient was advised split face trial of cauterization of lesions with plasma pen on one side of the face and monthly Fraxel CO₂ laser session on the other side, and is currently under follow-up period.

Discussion

Eruptive vellus hair cysts are proposed to arise as a congenital abnormality of the vellus hair follicles. There is a predisposition for occlusion of hair follicles at the level of the infundibulum, which results in cystic dilatation of the proximal hair follicle and thus leading to secondary atrophy of the hair bulb.³ Mutations in the gene that encodes keratin 17 have also been described in immune-histochemical analysis.⁴ Morgan et al.⁵ reported a patient with mild intellectual impairment, developmental delay and congenital eruptive vellus hair cysts. There is no racial predilection, and both males and females are equally affected.⁶

This benign condition is characterized by multiple, small skin-colored to brownish, dome-shaped, soft to firm papules ranging from 1 to 5mm in diameter.⁶ They may have a central punctum or umbilication. Sites commonly involved are chest and extremities and rarely seen on face, neck, axillae, abdomen and groin.⁷ Our case is a 'facial variant' of Eruptive Vellus Hair Cysts which is extremely rare.

The clinical differentials include giant comedones, milia, syringomas, colloid milium, infundibular cysts and steatocystoma multiplex.⁸ The definite diagnosis can only be confirmed by cutaneous biopsy which reveals mid-dermal cyst

containing laminated keratin and many vellus hairs, and lined by several layers of stratified squamous epithelium, often with a granular cell layer, and no adnexal structures are seen within the cyst wall.⁹

Although benign, this condition is very difficult to treat and shows no response to oral retinoids, as seen in our case as well.¹⁰ Spontaneous resolution is seen in only about 25% of the cases due to transepidermal elimination.⁸ Currently, there is no promising treatment but some improvement with dermabrasion, erbium:YAG or carbon dioxide laser and needle incision and drainage has been cited in the literature, but due to presence of numerous lesions, complete resolution is not achieved.¹⁰

It is believed that the entity 'Eruptive vellus hair cysts' is more common than it actually appears but it has been underdiagnosed and underreported because of its marked similarity to many other conditions and thus, we could not find any case reported from Pakistan till now. So, we report this case to generate awareness about this rarely diagnosed condition.

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