Zosteriform Eruptive Vellus Hair Cyst: A Rare Entity with an Uncommon Presentation

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Abstract

Eruptive vellus hair cyst (EVHC) represents a rare developmental anomaly of vellus hair follicles. The infrequency with which it is encountered makes it a formidable diagnostic challenge. Herein, we report a case of zosteriform EVHC in a 23-year-old male who presented to our dermatology clinic with asymptomatic, brown-black colored, follicular papules for 15 years. This case highlights a unique presentation of an uncommon entity.

Keywords: Eruptive vellus hair cyst, vellus hair, zosteriform distribution

Introduction

Eruptive vellus hair cyst (EVHC) represents an under-reported anomaly of vellus hair follicle, characterized by multiple, small, asymptomatic, dome-shaped follicular papules of size 1–5 mm with colors ranging from skin colored, yellow, blue, red to red brown in young adults. The lesions are usually located in anterior chest wall and extremities. Histopathologic examination (HPE) of these lesions is necessary to confirm the diagnosis. Herein, we report a case of EVHC in a 23-year-old-man who presented with asymptomatic papules and plaques over the abdomen in a zosteriform distribution. Till date, zosteriform variant has not been reported.

Case Report

A 23-year-old man presented with multiple, asymptomatic, purple to dark brown colored raised papules on the right side of the abdomen extending to the back and flexor aspect of the right upper limb. The lesions started during childhood and they gradually increased in number. The patient's past medical and family history was unremarkable. Examination of the skin revealed discrete, numerous 1–2 mm, hyperpigmented, dome-shaped, follicular, non-tender, papules in a zosteriform distribution over the right side of abdomen, back and right upper limb [Figure 1]. Few papules on the back coalesced to form a hyperpigmented plaque [Figure 2]. Areas of post-inflammatory hyperpigmentation were also present. Hair, nail, oral, and genital mucosa was normal. The differential diagnoses for these lesions were keratosis pilaris and zosteriform lichen planus and a punch biopsy from an abdominal lesion was performed. HPE revealed a well-defined cystic structure in the upper dermis filled with laminated keratinous material and multiple vellus hair shafts [Figure 3]. The cyst cavity was lined by stratified squamous epithelium with a prominent granular layer.

Based on the above findings, the diagnosis of Zosteriform EVHC was rendered. The patient was advised to apply retinoic acid (0.05%) and 30% improvement was noted after 3 months.

Discussion

The term EVHC was introduced by Esterly and Cols in 1977 in two pediatric patients.[1] Since then, few cases of EVHC have been reported in the literature.

Submission: 05-02-2022 **Revision:** 17-03-2022
Acceptance: 11-05-2022 **Web Publication:** 15-09-2022

Access this article online

Quick Response Code:

Website: www.tjdonline.org

DOI: 10.4103/tjd.tjd_30_22

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How to cite this article: Poddar S, Gayen, T, Chatterjee G. Zosteriform eruptive vellus hair cyst: A rare entity with an uncommon presentation. Turk J Dermatol 2022;16:98-100

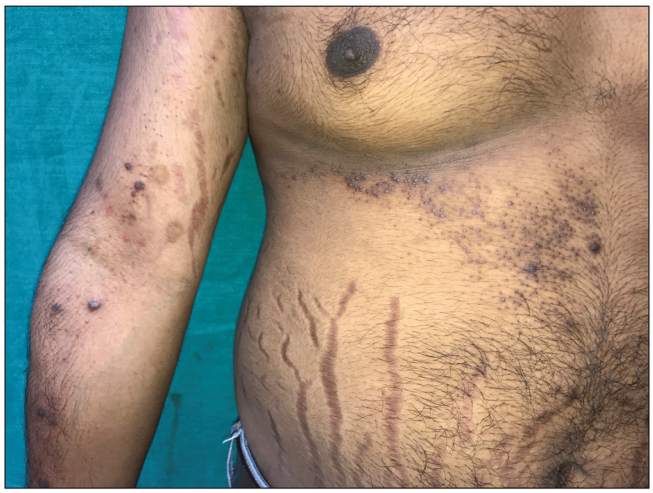

Figure 1: Multiple, hyperpigmented papules over the abdomen and right upper limb

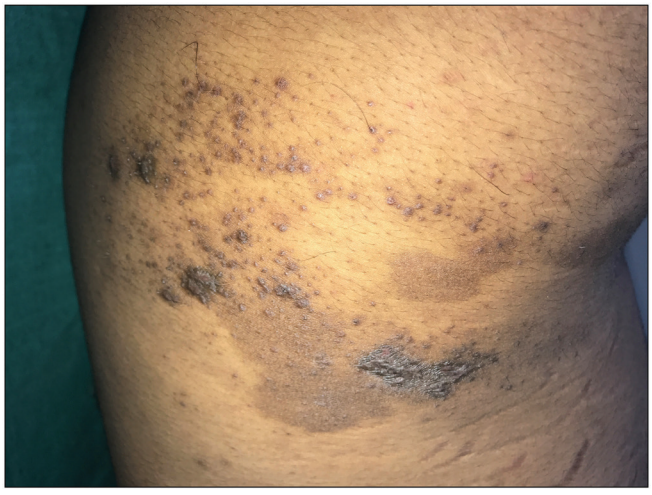

Figure 2: Papules and hyperpigmented plaque on abdomen in a zosteriform distribution

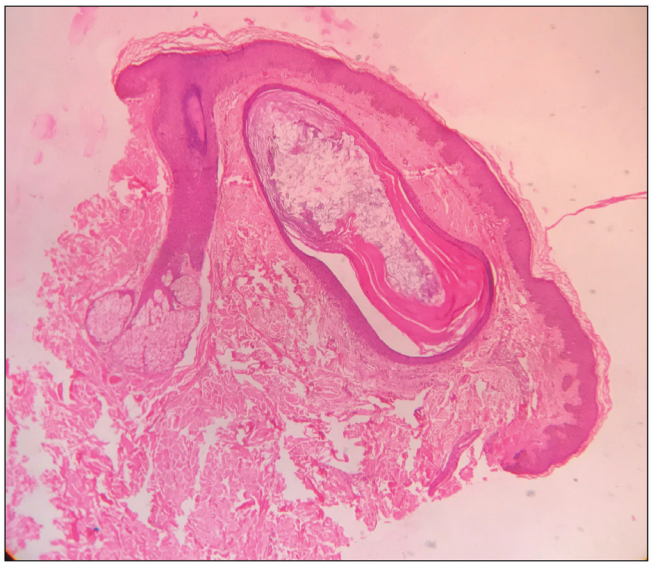

Figure 3: A well-defined cyst in the upper dermis lined by stratified squamous epithelium (H&E, X40)

EVHC can be inherited in an autosomal dominant pattern or sporadic in occurrence affecting patients of all ages, seen usually in the first three decades of life. The pathogenesis of EVHC is not yet fully understood. Esterly *et al.*[1] suggested that a developmental anomaly of vellus hair follicle predisposes to infundibular occlusion leading to hair retention, cystic dilatation, and secondary atrophy of the hair bulb. Steatocystoma multiplex, Lowe syndrome, pachyonychia congenital, syringoma, hidrotic ectodermal dysplasia, and anhidrotic ectodermal dysplasia have been associated with EVHC. In addition, EVHCs were reported in two Japanese patients with end-stage kidney disease on hemodialysis.[2]

EVHCs are generally asymptomatic; however, occasional tenderness and pruritus have been reported. The chest and flexor surfaces are the most involved sites. Other rare sites are abdomen, back, neck, axilla, groin, and labium major.[3] Generalized and facial variants resembling Nevus of Ota have been reported.[4] Bhushan and Singh[5] also reported facial localization of EVHC. Zosteriform presentation of EVHC has not been reported yet in the literature.

The clinical differential diagnoses include steatocystoma multiplex, milia, keratosis pilaris, folliculitis, other cysts (infundibular, trichilemmal, epidermal), acne, perforating folliculitis, trichostasis spinulosa, and molluscum contagiosum.

EVHC despite being a benign condition can be of cosmetic concern. Lesions can resolve spontaneously in some cases via transdermal elimination of cyst contents or their granulomatous dissolution. There are no standardized therapeutic options available. Pharmacotherapy with topical keratolytic agents can be utilized. Oral isotretinoin and Vit A therapy resulted in minimal clinical improvement in a few studies.[6] Improvement with CO2 laser vaporization, pulsed erbium:yttrium-aluminum-garnet (Er:YAG) laser, dermabrasion, and needle incision have been reported in the literature.

EVHC is an infrequently diagnosed condition. The uncommon clinical manifestations can occasionally cause diagnostic challenges. It should be kept in mind that EVHC can present in a zosteriform distribution and it should be considered as a differential diagnosis in the presence of zosteriform dermatoses.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Not applicable.

Conflicts of interest

There are no conflicts of interest.

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