CASE REPORT

Eruptive vellus hair cysts: A localized unilateral variant

Ahmed Almutairi, MD, Mohammad Elkhashlan, MD, Samar Zeitony, MD

Department of Dermatology, Al Ahmadi Hospital, Kuwait

ABSTRACT

A 22-year-old female presented with multiple, asymptomatic, follicular papules distributed over the right side of the abdomen. The condition started 3 years ago with sudden onset, and a rapid progressive course in last 6 months. Histological examination showed small dermal cyst in which the cavity was filled with numerous vellus hair confirming the diagnosis of eruptive vellus hair cysts (EVHC). There was no family history of similar condition. Although surgical excision of the lesions was the recommended treatment, the patient refused such management to avoid postoperative scarring or keloid formation. In the report we presented a rare entity of EVHC as localized unilateral variant.

INTRODUCTION

Eruptive vellus hair cysts (EVHC) is a well-known entity since 1977. When it was reported in two children with multiple, asymptomatic, monomorphous, hyperpigmented papules that were distributed on the trunk and flexor aspects of the extremities.[1] Classically, EVHC are characterized by multiple small asymptomatic yellowish or brownish papules with a smooth or centrally umbilicated surface. Their diameters range from 1 to 5 mm.[2]

They are usually localized on the chest and proximal extremities, but they may be present at unusual sites, including the face, groin and buttocks. There are reports of generalized, and of isolated facial variants in the literature.[3] Lesions may appear hyperpigmented, flesh-colored, yellow, erythematous, or blue. Rarely, lesions are umbilicated or hyperkeratotic. The vast majority of cases are asymptomatic. However, there are reports of associated tenderness or pruritus.[4]

EVHC may occur sporadically or be inherited in an autosomal dominant fashion.[5] The majority of cases are isolated and benign findings. However, there are reports of EVHC occurring in association with renal failure and with such disorders as anhidrotic ectodermal dysplasia, hidrotic ectodermal dysplasia, pachyonychia congenita, and Lowe syndrome.[6]

Diagnosis of EVHC is confirmed by histologic examination of a cutaneous biopsy specimen, which shows a mid-dermal cyst that contains laminated keratin and multiple transversely- and obliquely-cut vellus hairs. A flattened squamous epithelium lines the cyst wall, and it usually shows a prominent granular layer. Ruptured cysts demonstrate a granulomatous reaction in the adjacent dermis, with presence of mixed inflammatory infiltrate associated with a variable number of multinucleated giant cells.[7]

An alternative, less invasive diagnostic approach has been described in which a superficial incision at the top of the lesion enables expression of the cystic contents. Microscopic examination of the

Correspondence: *Dr. Ahmed Almutairi, Department of Dermatology, Al Ahmadi Hospital, Kuwait. Almutairi@kockw.com*

expressed contents in a potassium hydroxide preparation shows numerous vellus hairs.[8]

We report here a case of an EVHC that developed in a rare unilateral distribution as a localized form in the abdomen. Although the lesions were completely asymptomatic, cosmetic disability was the chief concern of our patient. The treatment of persistent lesions is often challenging with disappointing results. We have reviewed the literature for atypical variants and different treatment modalities.

CASE REPORT

A 22-year-old female presented with multiple, asymptomatic, follicular papules that were distributed over the right side of abdomen. The condition started 3 years ago with sudden onset and rapid progressive course over 6 months duration. The clinical examination revealed several well-defined pinhead-to-match head-sized skin coloured and slightly pigmented papules that were localized on the right side of the abdomen (Fig. 1 A & B).

The patient had felt mild itching on some occasions. However, most of the time the papules were asymptomatic. There was no family history of any such occurrences or any chronic skin disease. There was no history of any systemic disease or chronic infection.

The case was diagnosed initially as superficial folliculitis and she was treated with topical antibiotic, adapalene cream and benzoyl peroxide gel. After 2 months, the lesions showed no response to any of these medications and moreover, they were increased in number. A punch skin biopsy (3mm in diameter) was performed from the papule under local anaesthesia. Histologic examination showed a small mid-dermal cyst that

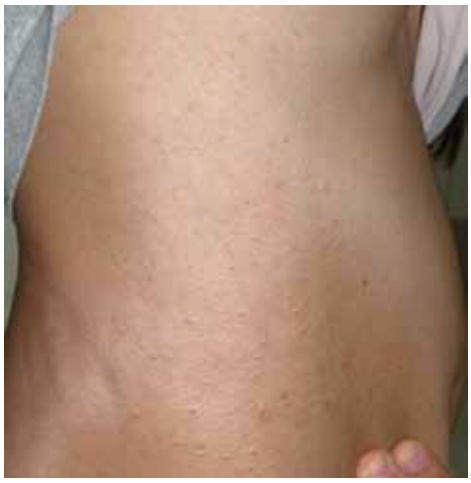

Fig. 1A Several well-defined pinhead-to-match head-sized papules are localized on the right side of the abdomen.

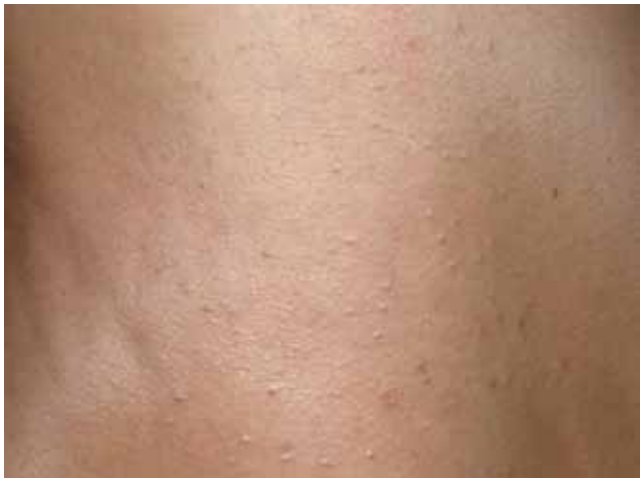

Fig. 1B Closeup view of the lesions.

was lined by a stratified squamous epithelium cell (Fig. 2). Most of the lining epithelium showed a granular layer. The cavity of the cyst was filled with laminated keratin materials and cutting vellus hair shafts (Fig. 3). These findings were consistent with the diagnosis of vellus hair cyst.

Although surgical excision of the lesions was the recommended treatment, the patient refused such management to avoid postoperative scarring or keloid formation. Follow up of the patient showed no progression of the disease over 2 years.

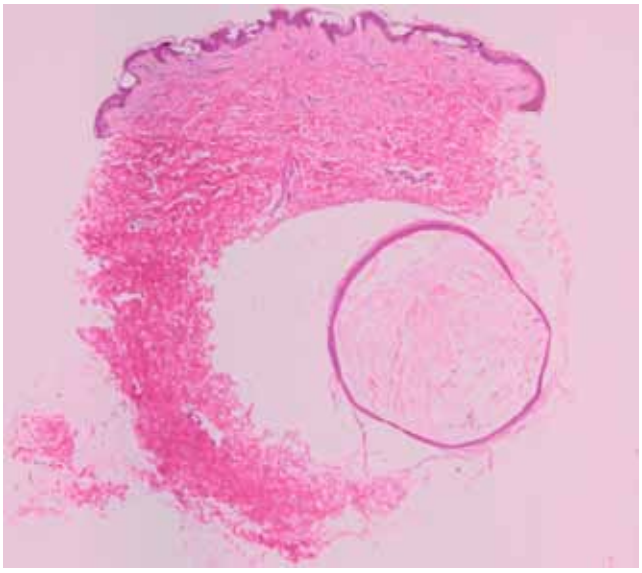

Fig. 2 A cyst lined by squamous epithelium in the mid-dermis (H&E, ×40).

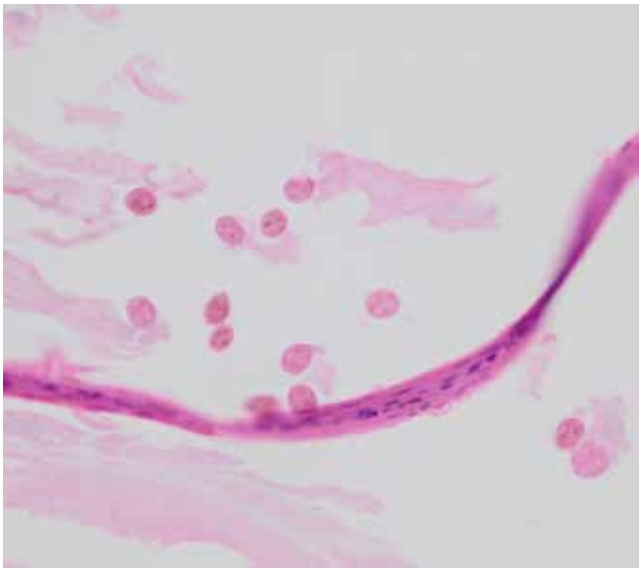

Fig. 3 The lining epithelium of the cyst showed granular layer. The Cavity is filled with vellus hair (H&E, ×400).

DISCUSSION

Eruptive vellus hair cyst (EVHC), described initially in 1977, is a benign dermatologic condition that is characterized by the sudden appearance of monomorphic, follicular, asymptomatic, small papules in children and young adults. EVHC demonstrates no sex or racial predilection.[9] The literature contains reports of the onset of EVHC ranging from birth to adulthood. Generally, in familial cases, lesions are first noted at birth or shortly thereafter. Whereas, the usual age of onset is between 4 and 18 years in the acquired form.[10]

Although the precise pathogenesis of EVHC is unknown; it may represent an unusual developmental abnormality of vellus hair follicles. However, some postulate that developmental abnormalities predispose vellus hair follicles to infundibular occlusion. Follicular occlusion leads to retention of hairs, proximal cystic dilatation, and consequent atrophy of the hair bulb.[11]

The diagnosis is based on the histopathologic findings of stratified-squamous epithelium with a granular layer that surrounds a cystic space filled with laminated keratin and a variable number of vellus hair cysts. EVHC can be associated with steatocystoma multiplex. A current hypothesis suggests that EVHC originates from a cystic change at the insertion of the pilosebaceous duct.[12] Overlapping histopathologic features and reports of EVHC and steatocystoma multiplex appearing together in the same patient have led some to put an etiologic link between these two entities.[13]

Familial EVHC is characterized by autosomal dominant transmission with complete penetrance, early age of onset (nine years), and a tendency toward persistence. Late onset EVHC (35 years or older) primarily is described in Asian women and is characterized by the appearance of a solitary lesion on the face or anterior chest, with an average age of onset of 57 years.[14] The clinical differential diagnosis includes steatocystoma multiplex, milia, keratosis pilaris, folliculitis (i.e., microbial, sterile, or perforating), infundibular cysts, trichilemmal cysts, dermoid cysts, acne cysts, and molluscum contagiosum.[15]

Although generally an asymptomatic and

innocuous condition, EVHC can be cosmetically bothersome. Spontaneous resolution occurs in an estimated 25 percent of lesions and is thought attributable either to transepidermal elimination or to granulomatous dissolution.[16] Trichostasis spinulosa is another important disorder in the differential diagnosis of EVHC. Protruding vellus cysts in Trichostasis spinulosa may be unapparent to naked eyes and the disease may clinically simulate EVHC. Histological examination is essential to differentiate these similar skin lesions.[17]

Although the diagnosis of EVHC should be confirmed with punch biopsy and histological examination. Nevertheless, punch biopsy can be both distressing and painful, and may leave an unpleasant scar. An alternative method to support the clinical diagnosis was implicated by utilizing a sterile, large (18 Gx2) blood-collecting needle to aspirate cystic contents from anesthetized skin. While in a different technique, the skin was punctured using a sharp-tipped cautery point. After that the base of the cyst was grasped using a standard dissecting forceps and extracted out. These techniques are less invasive methods than punch biopsy.[18]

In most cases, EVHC is found on the anterior chest wall, abdomen and/or extremities, yet other unusual sites can be involved. Kumakiri et al.[19] experienced a localized form of EVHC on the forehead and they called it the facial variant, and there has since been several case reports of such facial variants. Less than 15 cases of the so-called facial variant form of EVHC, which is localized exclusively on the face, have been reported. There have also been a few cases on an unusual site such as the buttocks.[19]

The uniqueness of this case is its atypical distribution. Until now, there have been reports of EVHC on the chest, extremities, face, neck and buttocks, but not as a unilateral localized form in the abdomen. Unilateral form was previously reported by Lew et al. on the face.[20] The reason why the lesions were localized in a unilateral distribution in this case is obscure. Some studies have postulated that cystic lesions may be formed by such triggers as minor trauma or scratching, but there is no clear evidence about it. In this case, the itching was mild and not associated with any scratch marks.

EVHC are usually asymptomatic lesions, but treatment is necessary for cosmetic reasons. Spontaneous resolution can be seen in one-fourth of the patients because of transepidermal elimination. Topical (retinoic acid, 12% lactic acid, 10% urea), mechanical (cauterization, curettage, dermabrasion, CO_2 laser) and systemic (oral isotretinoin) treatment modalities are available.[21] Surgical techniques, which include excision, curettage, needle evacuation, and incision and drainage, have been utilized but are time consuming and may result in scars. Other destructive modalities, which include dermabrasion and laser therapy using a CO_2 or erbium:yttrium-aluminum-garnet (Er:YAG) laser, have been employed. These approaches enable rapid treatment of multiple lesions. However, there utility is limited by inconsistent results and an increased risk of scars.[22]

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