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Case and Review

A Case Report of Idiopathic Follicular Hyperkeratotic Spicules and Literature Review

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Keywords
Follicular hyperkeratosis · Myeloma · Spiky · Spinulosis · Spiny

Abstract
Follicular hyperkeratotic spicules is a rare skin disorder that is usually associated with multiple myeloma. The condition typically presents with tiny hyperkeratotic spicules in follicular distribution and predominantly on the face. To our knowledge, there has been one reported case of this condition without underlying disease. We herein report the second case of idiopathic follicular hyperkeratotic spicules in a 54-year-old Thai woman presenting with multiple follicular horn-like spicules on her face and neck.

Introduction

Follicular hyperkeratotic spicules is a rare cutaneous disorder characterized by spiky, skin-colored to yellowish, follicular digitate hyperkeratosis. This condition was firstly introduced by Heidenstorm and Tottie in 1944 [1]. Since then, several subsequent case reports had

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been published, sharing similar locations on the face, particularly the nose, chin, and forehead. Although its etiology is still unknown, it is often reported in association with multiple myeloma. It is also associated with other conditions such as Crohn's disease and drug-induced reactions [2–4]. We herein report a case of follicular hyperkeratotic spicules without associated condition.

Case Report

A 54-year-old Thai female presented with a group of multiple spiky spicules along her face, jawline, and neck for 1 month. No pain or pruritus accompanied the lesions. The spicules were easily removed by scratching or rubbing without bleeding, then reappeared within a few weeks. She denied previous illness before the development of the skin lesion. She had no underlying disease and was not taking any supplement or medication.

Physical examination revealed multiple discrete tiny filiform hyperkeratotic papules on the face and neck (Fig. 1). The histopathological study demonstrated dilated follicles with digitate follicular hyperkeratosis and parakeratosis. Neither trichostasis nor koilocyte was observed (Fig. 2). Laboratory investigations, including complete blood count, blood chemistry, serum protein electrophoresis, and urinalysis were within normal limit. Based on the history, physical examination, histopathological study, and laboratory findings, the diagnosis of idiopathic follicular hyperkeratotic spicules was performed. The patient received topical 0.1% adapalene gel to be applied once daily. Two months later, the condition showed mild improvement.

Discussion

Follicular hyperkeratotic spicules is a rare skin disorder presenting with tiny, skin-colored to yellowish, hyperkeratotic spikes that are commonly located on the face, specifically on the nose, and occasionally on the trunk and extremities. However, the lesion can appear anywhere on the body. Most of the affected patients were middle-aged and elderly [4]. Histopathological examination reveals focal spike-like orthokeratotic or parakeratotic column filling the infundibulum with eosinophilic inclusions and protruding above the epidermis. Lymphocytic infiltrations in the papillary dermis are usually sparse [5–7].

The disease is often reported as being a paraneoplastic cutaneous sign of multiple myeloma or cryoglobulinemia [3]. Additionally, it has also been reported in associations with other conditions such as Crohn's disease, HIV infection, and drug-induced reactions from cyclosporine, sorafenib, and acitretin [8–11]. There has been one reported case without the associated disease [5]. To our knowledge, our patient is the second reported case of idiopathic follicular hyperkeratotic spicules. Previously reported cases of this disorder are demonstrated in Table 1 and Table 2 [1–3, 5–10, 12–23].

Differential diagnosis of follicular hyperkeratotic spicules includes lichen spinulosus, multiple minute digitate hyperkeratosis, trichodysplasia spinulosa, and spiky follicular mycosis fungoides [4, 24–27]. Details of these diseases regarding clinical manifestations, histopathological findings, and their associated conditions are summarized in Table 3.

The etiology of follicular hyperkeratotic spicules is still inconclusive. There have been various hypotheses regarding the pathogenesis of the disease. Bork et al. [14] demonstrated that myeloma dysprotein and cryoglobulins precipitated in the follicular infundibulum, resulting in follicular plugs and spiky presentation. A subsequent report of cryoglobulins accumulation in follicular content obtained from a patient with multiple myeloma supports this finding [3]. Besides, there has been speculation that Merkel cell polyomavirus, *Propionibacterium acne*, and *Demodex folliculorum* are the causes of the disease [15, 19–23]. However, there is a lack of evidence to support that these microorganisms play a role in the disease pathogenesis.

Several topical agents, including 12% lactic acid cream, adapalene gel, tretinoin cream, fluocinolone acetonide oil, and antibiotics have been tried to treat follicular hyperkeratotic spicules [5, 16]. None of them showed effectiveness. However, improvement of the lesions in patients with multiple myeloma after receiving systemic chemotherapy has been reported [6, 12, 13, 16].

In conclusion, we present a rare case of idiopathic follicular hyperkeratotic spicules. Due to multiple reports of associated conditions, we recommend that complete physical examination and laboratory evaluation are important to find the associated disease. Moreover, long-term monitoring and re-assessment are suggestive in all cases of idiopathic follicular hyperkeratotic spicules.

Statement of Ethics

The patient provided written informed consent to perform all necessary investigations, to take clinical photographs, and use them for research purposes and publication. This case report was conducted ethically in accordance with the World Medical Association Declaration of Helsinki.

Disclosure Statement

The authors have no conflicts of interest to declare.

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Author Contributions

All named authors meet the International Committee of Medical Journal Editors (ICMJE) criteria for authorship for the manuscript, take responsibility for the integrity of the work as a whole, and have given final approval to the version to be published.

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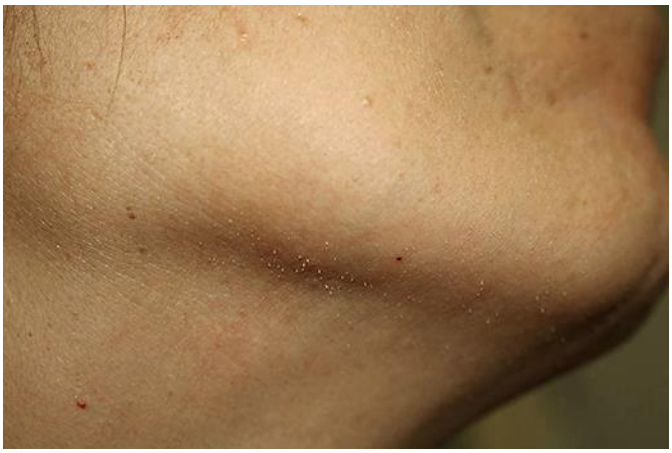

Fig. 1. Multiple discrete tiny filiform hyperkeratotic papules on the face and neck.

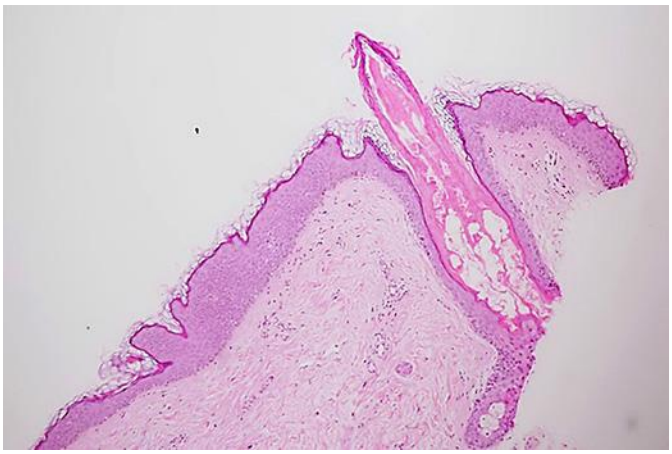

Fig. 2. Microscopic examination showing dilated follicle with digitate follicular hyperkeratosis and parakeratosis together with sparse superficial perivascular cells infiltration of lymphocytes (hematoxylin-eosin, original magnification ×20).

Table 1. Reported cases of follicular hyperkeratotic spicules

Author	Year	Sex	Age	Distribution	Associations	Treatment response
Heidenstorm and Tottie [1]	1944	M	39	Face, chest	Multiple myeloma	NA
Braverman[a]	1970	M	NA	Face	Multiple myeloma	NA
Lukitsch et al. [12]	1985	M	42	Face, arms	Multiple myeloma, cryoglobulinemia	Disappearance
Kuokkanen et al. [13]	1987	M	54	Face, legs	Multiple myeloma	Disappearance
Castanet et al.[a]	1987	M	59	Face, scalp, chest, extremities	Multiple myeloma	Disappearance
Brunner et al.[a]	1987	M	66	Face, scalp	Multiple myeloma	Disappearance
Bork et al. [14]	1990	M	62	Face	Multiple myeloma, cryoglobulinemia	No response
Requena et al. [3]	1995	M	79	Face, scalp, back	Multiple myeloma, cryoglobulinemia	Disappearance
Paul et al. [15]	1995	M	61	Face, scalp, lumbar area	Multiple myeloma	Partial response
Pestarino et al.[a]	2000	M	79	Face	Multiple myeloma	Partial response
Braun et al. [7]	2002	M	58	Face	Multiple myeloma	Increase with worsening of multiple myeloma
Satta et al. [6]	2003	F	68	Face	Multiple myeloma, cryoglobulinemia	Disappearance, recurrence with worsening multiple myeloma
Satta et al. [6]	2003	M	79	Face, trunk, arms	Multiple myeloma	NA
Miller et al. [16]	2006	F	70	Face	Multiple myeloma	Partial response
Tay et al. [17]	2010	F	55	Face	Multiple myeloma	Disappearance
Dalal et al. [18]	2010	F	67	Face	Multiple myeloma	Increase with worsening of multiple myeloma
van Boheemen et al. [19]	2015	M	70s	Face, chest, arms	Multiple myeloma	Disappearance (cidofovir gel 1% added)
Aloi et al. [2]	1989	F	38	Trunk, neck, upper extremities	Crohn's disease	No response

NA, not available. [a] Reference cited in Satta et al. [6].

Table 2. Reported cases of follicular hyperkeratotic spicules (continued)

Author	Year	Sex	Age	Distribution	Associations	Treatment response
Farina et al. [20]	1998	F	78	Face	Polycythemia vera (suspected of demodicidosis)	No response
Franck et al. [8]	2010	M	61	Face, scalp, trunk, arms	Sorafenib-induced reaction	Disappearance after stop medication
Franck et al. [8]	2010	M	72	Face, scalp, trunk, arms	Sorafenib-induced reaction	Disappearance after stop medication
Franck et al. [8]	2010	M	78	Face, scalp, trunk, arms	Sorafenib-induced reaction	Disappearance after stop medication
Franck et al. [8]	2010	M	59	Face, scalp, trunk, arms	Sorafenib-induced reaction	Disappearance after stop medication
Franck et al. [8]	2010	M	49	Face, scalp, trunk, arms	Sorafenib-induced reaction	Disappearance after stop medication
Franck et al. [8]	2010	F	79	Face, scalp, trunk, arms	Sorafenib-induced reaction	Disappearance after stop medication
Franck et al. [8]	2010	M	65	Face, scalp, trunk, arms	Sorafenib-induced reaction	Disappearance after stop medication
Franck et al. [8]	2010	M	60	Face, scalp, trunk, arms	Sorafenib-induced reaction	Disappearance after stop medication
Franck et al. [8]	2010	M	60	Face, scalp, trunk, arms	Sorafenib-induced reaction	Disappearance after stop medication
Chia et al. [21]	2010	M	22	Neck	Suspected *Propionibacterium acnes*	Response to oral erythromycin
Nemeth et al. [22]	2016	M	NA	Scalp	Lung transplantation	Response to systemic valganciclovir
Yanik et al. [9]	2016	F	51	Face, neck	Acitretin-induced reaction	Disappearance after stop medication
Ruiz-Rivero et al. [23]	2017	F	16	Face	Plaque morphea (suspected demodicosis)	Response to ivermectin
Maddy et al. [10]	2018	M	52	Face	HIV infection	Increased with worsening HIV infection
Kim et al. [5]	1997	F	52	Face	Idiopathic	No response
Current case	2019	F	54	Face	Idiopathic	Partial response

NA, not available.

Table 3. Differential diagnosis of follicular hyperkeratotic spicules

Diseases	Clinical findings	Histopathology	Associations
Lichen spinulosus	Patches of follicular papules topped by keratotic spines , predilection for trunk and extremities	Dilated infundibulum filled with columnar orthokeratotic keratin plug, occasionally dense lymphocytic perifollicular infiltrates	Ichthyosis, Atopic dermatitis, HIV infection
Multiple minute digitate hyperkeratosis	Multiple minute digitate hyperkeratosis, predominantly affect trunk and extremities	Nonfollicular, focal columns of orthokeratotic hyperkeratosis arising from a tented epidermis, with prominent stratum granulosum	Familial, sporadic, post-inflammation, paraneoplastic
Trichodysplasia spinulosa	Multiple keratotic spicules on follicular erythematous papules, mostly on face	Dilated and dystrophic hair follicles with proliferation of the inner root sheath cells containing large trichohyaline granules	Immunocompromised host, associated with papovavirus
Spiky follicular mycosis fungoides	Slightly erythematous, hyperkeratotic spiky or cone-shaped follicular papules	Hyperkeratotic columns protruding from follicular plugs with infiltration of atypical lymphocytes around follicular epithelium	Mycosis fungoides, Sezary syndrome