

Unusual variant of keratoacanthoma with spontaneous regression

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ABSTRACT :

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Keratoacanthoma centrifugum marginatum (KCM) is an extremely rare variant of keratoacanthoma(KA). In contrast with classic KA, KCM is a special type of KA that shows little tendency to clear spontaneously. We report an exceptional case of keratoacanthoma centrifugum marginatum with spontaneous regression within 5 months. The key point in the history is a lesion diminishing in size or at least one that has stopped enlarging. Histology of involuting tumor gave us confident to merely give close observation.

Key words : Keratoacanthoma, Tumor, Regression

บทคัดย่อ :

ชัชฎาภรณ์ กิริติขจรชัย, ฤมุตนาถ จันทร์ประภาพร รายงานผู้ป่วย KERATOACANTHOMA ชนิดที่พบได้น้อย และหายได้เอง วารสารโรคผิวหนัง 2554; 27: 142-146.

หน่วยโรคผิวหนัง ภาควิชาอายุรศาสตร์ คณะแพทยศาสตร์โรงพยาบาลรามาธิบดี มหาวิทยาลัยมหิดล

Keratoacanthoma centrifugum marginatum เป็นโรคที่พบบได้น้อย จัดเป็นชนิดหนึ่งของ keratoacanthoma ซึ่งมีความแตกต่างคือพบว่ามีกรหายเองได้น้อย ผู้เขียนได้รายงานผู้ป่วยหนึ่งราย ที่วินิจฉัยเป็น Keratoacanthoma centrifugum marginatum ที่มีรอยโรคขนาดเล็กลงเรื่อยๆ ในระยะเวลา 5 เดือน ลักษณะสำคัญคือรอยโรคไม่ขยายขนาดขึ้น การตรวจทางพยาธิวิทยาพบว่ารอยโรคอยู่ในช่วงกำลังเสื่อมถอย

คำสำคัญ : เคอราโตอะแคนโทมา (keratoacanthoma), เนื้องอกผิวหนัง, หายเอง

Introduction

The keratoacanthoma(KA) and its variants are clinically and histologically heterogeneous. Some consider the keratoacanthoma (KA) to be benign, whereas other classify it as a subtype of squamous cell carcinoma. Keratoacanthoma centrifugum marginatum (KCM) presents a diagnostic challenge, as it may resemble a subcutaneous mycosis, an atypical mycobacterial infection or especially squamous cell carcinoma (SCC).

Case Report

A 82-year-old woman presented with an expanding lesion on the left leg for 2 months. The patient noticed asymptomatic faint brownish to erythematous plaque on her left leg. The surface contained multiple keratotic punctums arranged closely set together. The rash gradually expanded over time. Her past medical history included diabetes mellitus, hypertension and

dyslipidemia. Physical examination revealed a solitary well-defined erythematous annular plaque with indurated border and multiple keratotic punctums on her left leg (Figure1). Skin biopsy was obtained and stained with hematoxylin-eosin (Figure2). Histopathologic examination of the skin biopsy specimen showed deep invasion of flattened epidermis with a hint of keratotic crater. Scar was seen in the surrounding dermis. Histologic study was consistent with keratoacanthoma (KA). The clinical finding of multiple tiny closely set dome shaped cratered papules with central keratin plug together with histologic finding typical of KA suggested the diagnosis of keratoacanthoma centrifugum marginatum in this patient. Histology was compatible with KA in the involution stage. Hence, there was tendency towards a self-remitted disease. Conservative approach was adopted as a treatment option, the patient was on close followed up. The tumor was photograph

every 2-3 weeks. The patient was fully advised about the natural history of the disease and compliance with the follow-up schedule. Finally the tumor spontaneously regressed within 5 months (Figure 3).



Figure 1 Keratoacanthoma centrifugum marginatum on left leg

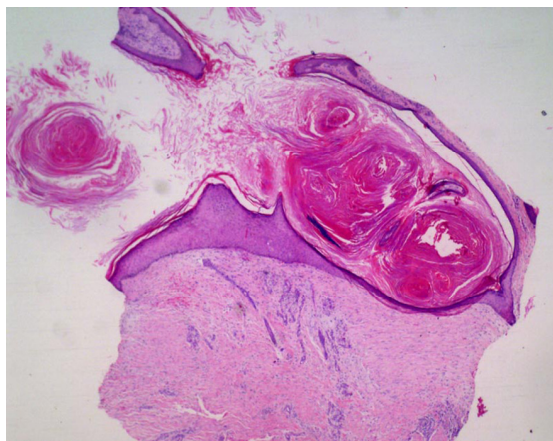


Figure 2 Deep invasion of flattened epidermis with a hint of keratotic crater. Scar was seen in the surrounding dermis (H&E 20X)



Figure 3 Spontaneous regression of tumor within 5 month

Discussion

Keratoacanthoma may present clinically as a solitary lesion or multiple lesions, in a sporadic fashion or in an inherited syndrome, or in association with inflammatory disease. Some KA may metastasize, and there is debate over the relationship of KA to squamous cell carcinoma (SCC) and whether to classify KA as benign or malignant. Solitary KA usually develops in sun-exposed areas, primarily on the face (lower lip, cheek, nose, eyelid), neck, and hands, and undergo three distinct stages¹. The first stage is a rapid proliferative phase, in which the tumor increases in size to reach 10–25 mm in diameter in 6–8 weeks. Histopathological finding demonstrates horned-filled invagination of the epidermis protruding into the dermis. The strands are poorly demarcated from the surrounding stroma in some

areas and may contain cell with nuclear atypia. The second stage is the mature KA, where the lesion stops growing, maintains its crater form, and appears as a bud, dome, or berry-shaped structure containing hair, a central keratin plug, and signs of fragmentation. Histology of fully developed lesion shows in its center a large, haphazardly shaped crater filled with keratin. Irregular epidermal proliferation extends both upward into the crater and downward from the base of the crater. Less nuclear atypia is seen than that of the initial stage. Finally, in the involution stage, 50% of KA undergoes spontaneous resolution in 4–6 weeks, with expulsion of the keratin plug and resorption of the tumoral mass, leaving an atrophic and hypopigmented scar. After proliferation has ceased, cells at crater base undergo keratinization. There may be shrunken, eosinophilic cells among the tumor cells located in the stroma, suggesting cell degeneration. Gradually, the crater flattens and disappears.

Keratoacanthoma centrifugum marginatum (KCM) is an extremely rare variant of KA since it was first described in 1962.¹ It is usually seen as a solitary lesion, most frequently appearing on the legs. In contrast with classic keratoacanthoma(KA), KCM extends horizontally across the surface, shows less tendency towards spontaneous resolution.^{1,2,3,4,5,6} There is persistent peripheral extension with a raised border, and atrophic center. Histologic study is the same as in KA. The clinical finding of multiple tiny closely set dome shaped cratered papules with central keratin plug

(Figure 1) together with histologic finding typical of KA suggested the diagnosis of KCM in this patient (Figure 2).

Numerous treatment modalities have been described for KCM, including surgical excision, Moh's micrographic surgery⁵, topical and intralesional bleomycin⁷, topical 5-fluorouracil⁸, topical imiquimod⁹, oral isotretinoin¹⁰ and etretinate.⁶ A recent literature reported a case of refractory, locally aggressive KCM of scalp having strong epidermal growth factor receptors (EGFRs) expression and demonstrating excellent response to erlotinib.¹¹

The total number of cases of KCM that have been reported throughout the world is very rare. This case illustrates an exceptional case of keratoacanthoma centrifugum marginatum with spontaneous regression. The key point in the history is a lesion diminishing in size or at least one that has stopped enlarging. Histology of involuting tumor gave us confidence to merely just wait and see with close observation. However in general, therapeutic intervention is preferable because there are no reliable criteria to differentiate with confidence between KA and squamous cell carcinoma.

References

1. Miedzinski F, Kozakiewicz J. Keratoacanthoma centrifugum--a special variety of keratoacanthoma. *Hautarzt* 1962;13:348-52.
2. Attili S, Attili VR. Keratoacanthoma centrifugum marginatum arising in vitiligo: a case report. *Dermatol Online J* 2006;12:18. 633-7.
3. Weedon D, Barnett L. Keratoacanthoma centrifugum marginatum. *Arch Dermatol* 1975;111:1024-6.
4. Divers AK, Correale D, Lee JB. Keratoacanthoma centrifugum marginatum: a diagnostic and therapeutic challenge. *Cutis* 2004;73:257-62.

5. Benest L, Kaplan RP, Salit R, Moy R. Keratoacanthoma centrifugum marginatum of the lower extremity treated with Mohs micrographic surgery. *J Am Acad Dermatol* 1994;31:501-2.
6. Ogasawara Y, Kinoshita E, Ishida T, Hamamoto Y, Fujiyama J, Muto M. A case of multiple keratoacanthoma centrifugum marginatum: response to oral etretinate. *J Am Acad Dermatol* 2003;48:282-6
7. de la Torre C, Losada A, Cruces MJ. Keratoacanthoma centrifugum marginatum: treatment with intralesional bleomycin. *J Am Acad Dermatol* 1997;37:1010-1.
8. Yuge S, Godoy DA, Melo MC, Sousa DS, Soares CT. Keratoacanthoma centrifugum marginatum: response to topical 5-fluorouracil. *J Am Acad Dermatol* 2006;54:S218-9.
9. Aroni K, Mastoraki A, Agrogiannis G, Ioannidis E, Patsouris E. Successful treatment of keratoacanthoma centrifugum marginatum with local application of imiquimod cream. *Int J Dermatol* 2007;46:1321-2.
10. Schaller M, Korting HC, Wolff H, Schirren CG, Burgdorf W. Multiple keratoacanthomas, giant keratoacanthoma and keratoacanthoma centrifugum marginatum: development in a single patient and treatment with oral isotretinoin. *Acta Derm Venereol* 1996;76:40-2.
11. Bulj TK, Kronic AL, Cetner AS, Villano JL. Refractory aggressive keratoacanthoma centrifugum marginatum of the scalp controlled with the epidermal growth factor receptor inhibitor erlotinib. *Br J Dermatol* 2010; 163: 633-7.