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Case Report

Annular elastolytic giant cell granuloma presented with annular erythematous patches over the face and cheek in a Chinese lady

環狀彈性纖維溶解性巨細胞肉芽腫在一位華裔女性的面上雙頰表現為環狀紅色班塊

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A 76-year-old lady presented with asymptomatic annular erythematous patches with hypopigmented atrophic centres distributed over the sun-exposed area including the forehead and cheek. Histopathologic examination showed epithelioid granuloma with multinucleated giant cells. Elastophagocytosis was evident by the granuloma engulfing masses of elastic tissue. Verhoeff stain showed absence of elastic fibers in the upper and mid dermis. The clinicopathological features were suggestive of annular elastolytic giant cell granuloma.

一位七十六歲女仕在身體曝光位置包括前額和面頰呈現無自覺症狀的環狀紅色班塊，其皮膚內有中央萎縮及色素減退。組織病理檢查顯示含有多核巨細胞的上皮樣肉芽腫。彈性纖維吞噬作用可見於肉芽腫內一團團被吞沒的彈性纖維組織，而維爾赫夫染色亦顯示出真皮中上層的彈性纖維缺失。以上的臨床病理特徵均與環狀彈性纖維溶解性巨細胞肉芽腫的診斷吻合。

Keywords: Actinic granuloma, annular elastolytic giant cell granuloma, Chinese, elastophagocytosis

關鍵詞：光化性肉芽腫，環狀彈性纖維溶解性巨細胞肉芽腫，華裔，彈性纖維吞噬作用

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Introduction

Annular elastolytic giant cell granuloma (AEGCG) is a rare granulomatous disease of unknown aetiology. AEGCG is clinically characterized by annular patches and plaques with erythematous borders and hypopigmented atrophic centres, which are mainly located in the sun-exposed areas. It is most commonly found in middle-aged white women. AEGCG has a chronic course and various treatment modalities have been reported

with inconsistent results. Before defining AEGCG as a unique disease entity, similar clinical cases were diagnosed as "atypical annular necrobiosis lipoidica", "Miescher's granuloma of face" and "actinic granuloma" in the literature. It points towards the difficulties in proper classification of AEGCG within the spectrum of granulomatous disease. Radial elliptical skin biopsy including both the centre and periphery of the lesion is essential to establish the clinicohistopathological diagnosis.

Case report

A 76-year-old lady presented with asymptomatic facial rash for 1 year. She was a housewife all along and there were no precipitating factor identified. She had hypertension and was on atenolol for over 10 years, otherwise her past health was unremarkable.

On physical examination, there was multiple annular erythematous patches measuring 0.5 cm-1 cm in size with atrophic and hypopigmented centres (Figures 1 and 2). The lesions were distributed over the forehead and cheek. The scalp, back of ear, limbs, trunk and mucosa were all spared. There was no obvious scaling and no accompanying loss of sensation over the hypopigmented centre. She was clinically well and had no palpable lymphadenopathy. The complete blood count, liver and renal function test, antinuclear antibody and erythrocyte sedimentation rate (ESR) were all unremarkable. Chest X-ray showed clear lung fields with no hilar lymphadenopathy.

Histopathologic examination of the radial elliptical skin biopsy over the forehead patches showed epithelioid granuloma with multinucleated giant cells in the centre of the lesion. The periphery of the lesion showed solar elastosis while the central portion was devoid of elastic tissue in the upper and mid dermis as illustrated by Verhoeff stain. Elastophagocytosis was evident by the granuloma engulfing masses of elastic tissue (Figures 3-6). The clinical features and histopathological findings were suggestive of annular elastolytic giant cell granuloma.

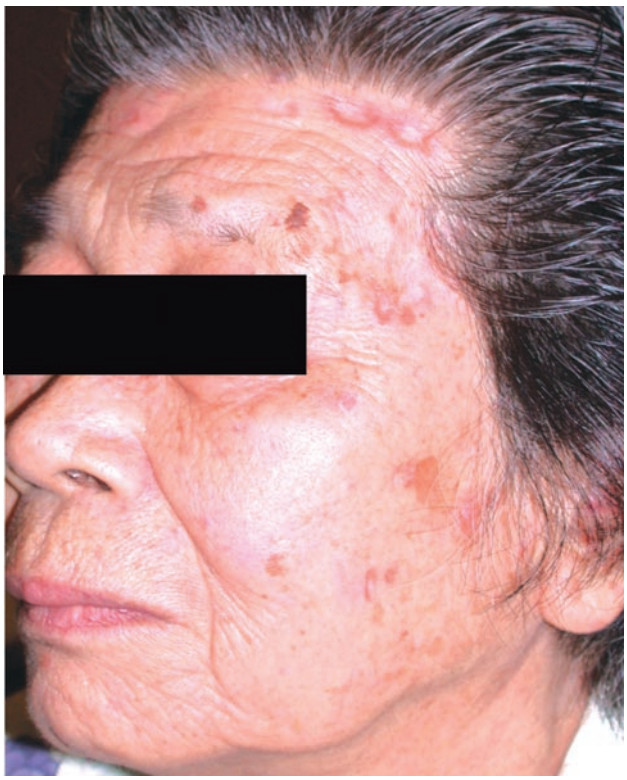

Figure 1. Annular erythematous patches with atrophic hypopigmented centre distributed over the forehead and cheek.

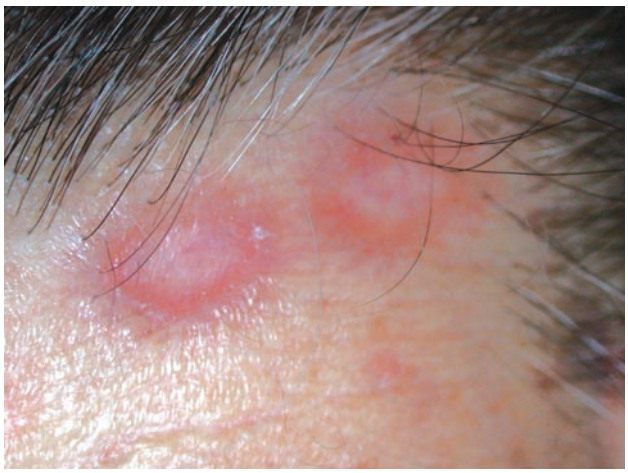

Figure 2. Close up view of the forehead lesion. (site of radial elliptical skin biopsy)

Topical fluticasone propionate cream was prescribed. The lesions remained static with no clinical deterioration.

Discussion

This disease entity was recognized as actinic granuloma by O'Brien in 1975.[1] O'Brien used 'actinic' in the name of this disorder because he believed that its etiology was linked to ultraviolet and infrared radiation. Another theory was proposed by Hanke et al whose patients did not all demonstrate significant solar elastosis histologically. These authors used the descriptive term "annular elastolytic giant cell granuloma (AEGCG)" for this condition in 1979.[2] Annular elastolytic giant cell granuloma is an uncommon disease that occurs predominantly in middle-aged women, usually over 40. The disease had been reported from several countries including Australia, the United Kingdom, the United States of America, the Caribbean, and Africa.

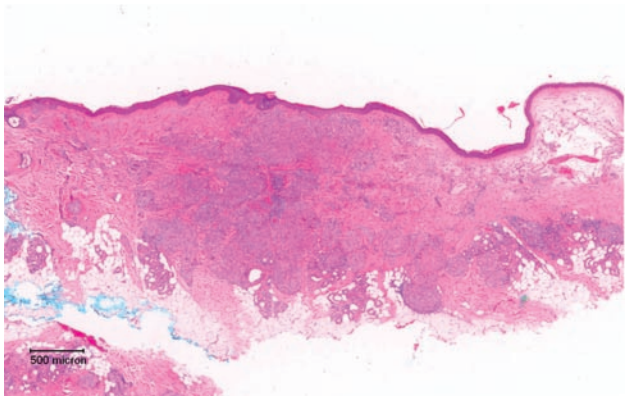

Figure 3. Low power view showing granulomatous inflammation in the dermis. (H&E stain)

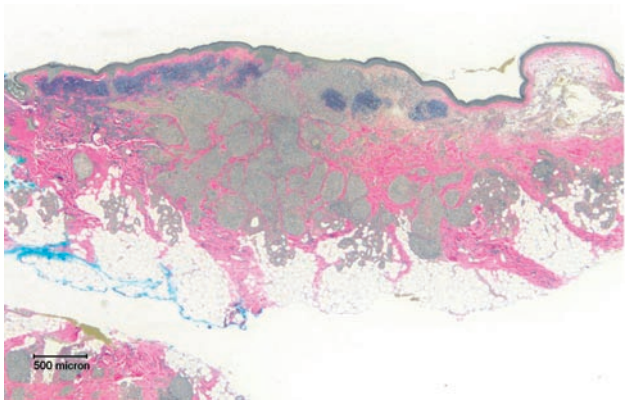

Figure 4. Complete absence of elastic tissue on the right side of the specimen in contrast with that on the left. Granulomatous inflammation is noted in the mid portion. (Verhoeff stain)

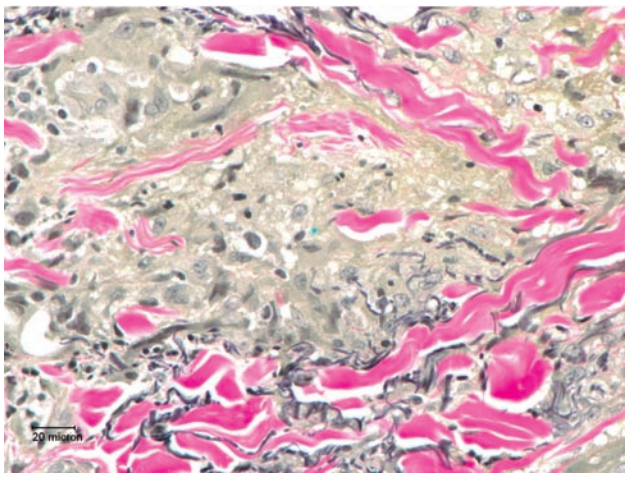

Figure 5. High power view showing elastophagocytosis by histiocytes in edge of a granuloma i.e. centre of the clinical lesion. (Verhoeff stain)

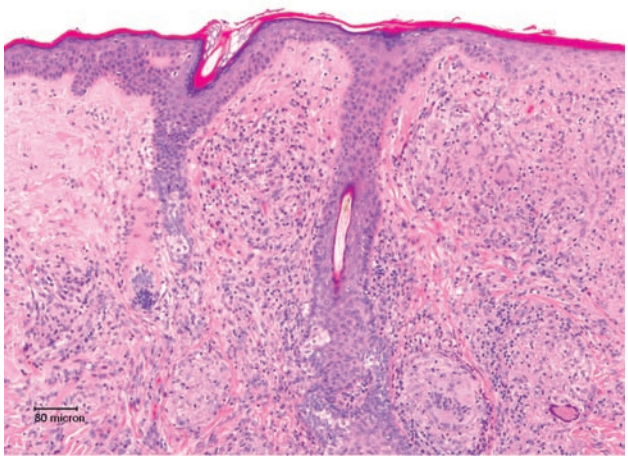

Figure 6. Medium power view showing granuloma with giant cells in the dermis. (H&E stain)

There is conflicting opinion that whether AEGCG is a distinct clinical entity or a variant of granuloma annulare.[3] Its pathogenesis is not fully understood, but it may be related to inflammation precipitated by actinic damage of the elastic fibers.[4] T-helper cells are predominately involved in AEGCG, suggesting that the pathogenesis may involve a cell-mediated immunologic response to a weakly antigenic determinant on altered elastic fibres.[5]

Annular elastolytic giant cell granuloma usually presents with annular patches and plaques with raised erythematous borders and slightly atrophic, hypopigmented central regions that were distributed in sun-exposed areas. The sites of predilection include the neck, face, chest and arms. Scale is rarely observed. Individual lesions usually measure 0.5-10 cm in diameter, and the total number of lesions are generally less than ten. Patients are usually asymptomatic, although pruritus has been reported. A single plaque may last for months to years, after which spontaneous remission may occur, leaving mottled dyspigmentation or normal-appearing skin.[1]

Histopathologic section over the raised border of the skin lesion showed the non-palisading granulomatous infiltrate of histiocytes, foreign body type multinucleated giant cells with haphazardly arranged nuclei, and lymphocytes in the mid to upper dermis, with an absence of altered collagen, mucin or lipid deposition.[5] Elastic fibres are identified adjacent to and within the giant cells, giving rise to the term elastophagocytosis. Asteroid bodies which stained like elastic fibres with acid orcein may be observed within the multinucleated giant cells. These findings are not specific for AEGCG. They are seen less frequently in other granulomatous conditions such as granuloma annulare and necrobiosis lipoidica. Elastin tissue stains, such as the Verhoeff-van Gieson stain, showed a characteristic complete absence of elastic fibres in the areas affected by granulomatous inflammation. There are no vascular changes other than a sparse perivascular lymphocytic infiltrate. Direct immunofluorescence studies for fibrin, C3, IgG, IgM and IgA are all negative.

The diagnosis of AEGCG is based on the clinical morphology and distribution along with the characteristic histopathological features. There is no helpful diagnostic laboratory test. Several clinical differential diagnoses for the annular erythematous plaques over the face should be considered including granuloma annulare, sarcoidosis, necrobiosis lipoidica, discoid lupus erythematosus, infective causes such as tinea faciale and granulomatous infections like lupus vulgaris and leprosy.

Annular elastolytic giant cell granuloma is a persistent condition that responds poorly or inconsistently to various kind of treatments including topical or intralesional corticosteroids, phototherapy such as PUVA and systemic therapy such as antimalarial and cyclosporine. One patient who responded to 16 weeks of chloroquine therapy (200-400 mg/day) and one patient who responded well to 8 weeks of cyclosporine (5 mg/kg/day) were reported.[6,7] Excision of the lesions with partial-thickness skin grafting has been reported with no recurrence at 15 months of follow-up.[8] Cryotherapy, cauterization and oral methotrexate were reported to be ineffective in single case reports.

In summary, our patient has asymptomatic AEGCG with fair response to topical steroid. Radial elliptical skin biopsy is crucial to establish the clinicohistopathological diagnosis.

References

1. O'Brien JP. Actinic granuloma: an annular connective tissue disorder affecting sun- and heat-damaged (elastotic) skin. Arch Dermatol 1975;111:460-6.
2. Hanke CW, Bailin PL, Roenigk HH Jr. Annular elastolytic giant cell granuloma. A clinicopathologic study of five cases and a review of similar entities. J Am Acad

Dermatol 1979;1:413-21.
3. McGrae JD Jr. Actinic granuloma. A clinical, histopathologic, and immunocytochemical study. Arch Dermatol 1986;122:43-7.
4. Regaz A, Ackerman AB. Is actinic granuloma a specific condition. Am J Dermatopathol 1979;1: 43-53.
5. Lim KB, Phay KL. Annular elastolytic giant-cell granuloma. Int J Dermatol 1987;26:463-4.
6. Tsutsui K, Hirone T, Kubo K, Matsui Y. Annular elastolytic giant cell granuloma: response to cyclosporin A. J Dermatol 1994;21:426-9.
7. Ozkaya-Bayazit E, Büyükbabani N, Baykal C, Oztürk A, Okcu M, Soyer HP. Annular elastolytic giant cell granuloma: sparing of a burn scar and successful treatment with chloroquine. Br J Dermatol 1999;140: 525-30.
8. Schwarz T, Lindlbauer R, Gschnait F. Annular elastolytic giant cell granuloma. J Cutan Pathol 1983; 10:321-6.