International Journal of Research in Medical Sciences
Vickneswaran K et al. Int J Res Med Sci. 2025 Feb;13(2):823-825
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20250259

Case Report

Clearing the circles: an unusual way

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Received: 25 November 2024
Accepted: 16 December 2024

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ABSTRACT

A 66-year-old lady with no significant past medical history presented with gradual onset of annular erythematous raised border with central clearing and atrophy plaque on her left arm. Subsequently, she noticed further plaques on her left forehead, and extensor surfaces of her arms. A diagnostic skin punch biopsy was organised from the affected site on right arm. The histology showed multi-nucleated giant cells with surrounding fragmented elastic fibres and elastophagocytosis with no dermal mucinosis and no interface inflammation. A working diagnosis of annular elastolytic giant cell granuloma (AEGCG) was formulated. Initially this was treated with a potent topical steroid, followed with tacrolimus ointment but the dermatoses did not settle. She was then started on hydroxychloroquine 200 mg twice daily. A repeat follow-up in three months showed the plaques has started to flatten out and the treatment was continued for a further three months. The patient was happy with the improvement of the rash. AEGCG is an uncommon granulomatous skin reaction typically involving sun exposed areas of the body. Clinically, this presents as annular erythematous plaques with central clearing or atrophy. The gold standard for diagnosis is based on histopathology showing granulomatous inflammation, multinucleated giant cells with more than one area of elastophagocytosis and absence of necrobiosis. There is no standard treatment, but treatment can range from topical therapy with corticosteroids, to phototherapy including narrow band ultraviolet B to systemic medications including oral antimalarial such as hydroxychloroquine. This case illustrates that hydroxychloroquine can be used as a treatment option.

Keywords: Annular elastolytic giant cell granuloma, Elastophagocytosis, Hydroxychloroquine

INTRODUCTION

Annular elastolytic giant cell granuloma (AEGCG) is a rare skin condition that can often be mistaken for other common skin disorders.[1,2] This enigmatic disorder is characterized by circular or annular lesions on sun-exposed sites that may be accompanied by itching, redness, and scaling.[2] While AEGCG is a benign condition, diagnosis can be difficult with various mimics, hampering timely treatment in the process.[2]

This case is highlighted for a relatively classical appearance and histology.

CASE REPORT

A 66-year-old lady with no significant past medical history presented with a six-month history of gradual onset of annular erythematous, raised border with central clearing atrophic plaque on her left arm (Figure 4). Subsequently, she developed further similar plaques on her left forehead (Figures 1 and 2) and extensor surfaces of her right arm (Figure 3).

Physical examination revealed annular erythematous plaques with raised borders and central clearing and some atrophy on the extensors of her left and right arms and left forehead (Figures 1-4).

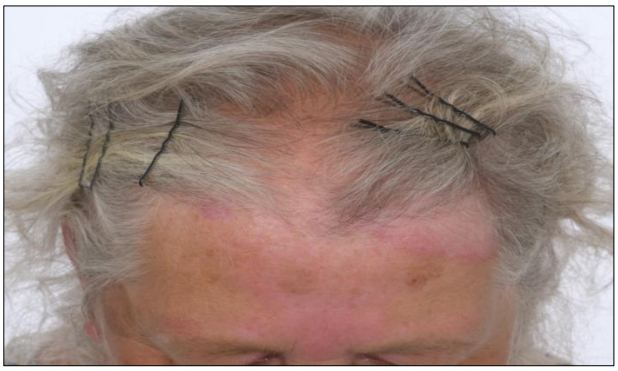

Figure 1: Forehead (front view).

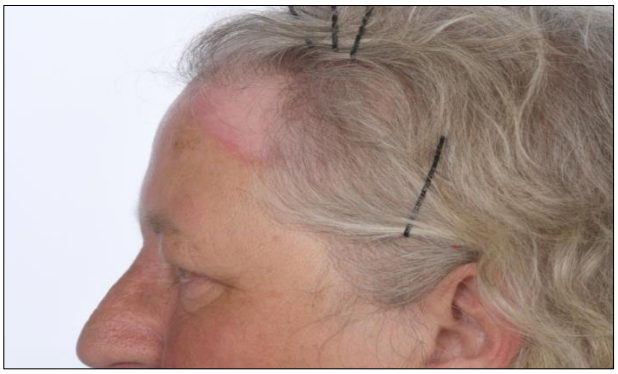

Figure 2: Forehead (left side view).

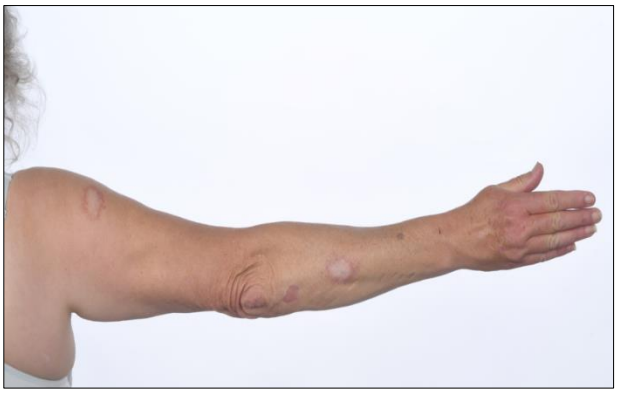

Figure 3: Right arm.

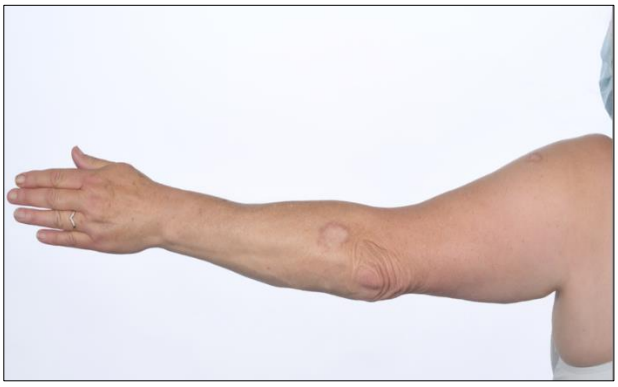

Figure 4: Left arm.

Initial preliminary investigations including full blood count (FBC), erythrocyte sedimentation rare (ESR), liver function tests, serum thyroid stimulating hormone (TSH) and serum anti-nuclear antibody (ANA) titres were within normal limits. A diagnostic skin punch biopsy was organised from the affected site on the right arm. The histology showed multi-nucleated giant cells with surrounding fragmented elastic fibres and elastophagocytosis with no dermal mucinosis and no interface inflammation.

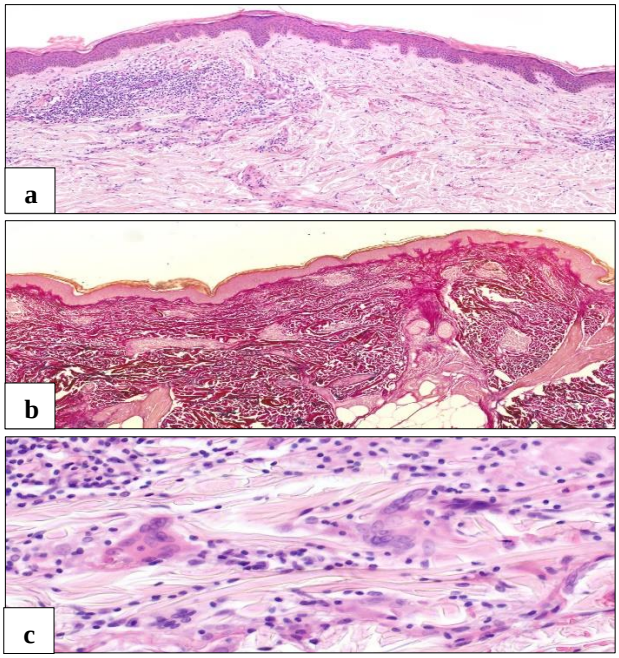

Figure 5: (a) H&E illustrating a normal epidermis with perivascular lymphohistiocytic inflammation, (b) elastic van Gieson illustrating disruption of the elastic architecture with loss of elastin, and (c) high power H&E stain where plasma cells and multinucelated giant cells can be appreciated surrounding fragmented elastic fibres.

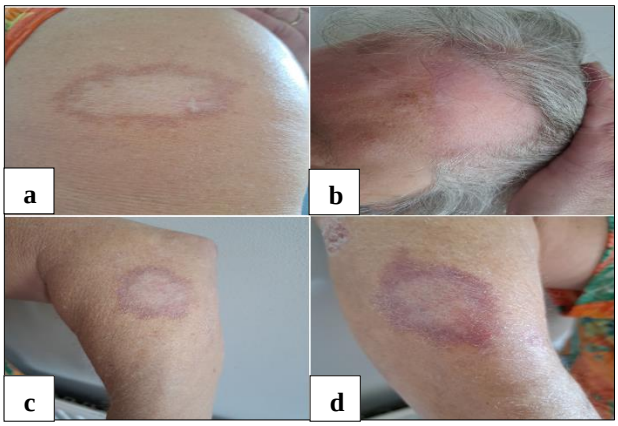

Figure 6: (a) Upper arm annular plaque now flat and paler, (b) patch on forehead, (c) and (d) patches on left and right arm, and (d) image showing the appearance whilst on treatment, respectively.

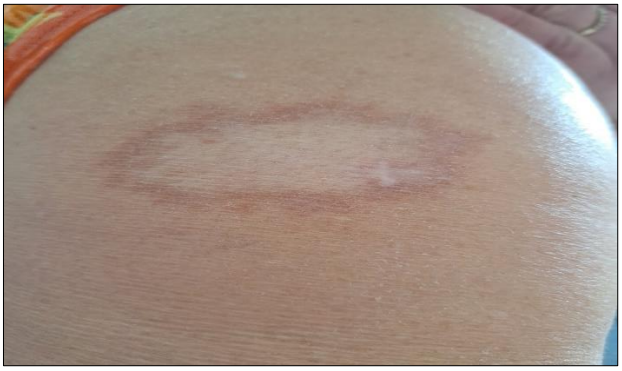

Figure 7: Zoom in image of right upper arm showing annular plaque to be flatter and paler.

Initially, this was treated with a potent topical steroid; mometasone furoate 0.1% ointment once daily for a week followed by topical tacrolimus 0.1% ointment once daily for 3 weeks. She continued this topical treatment regime for 3 months with very little improvement. She was then started on oral hydroxychloroquine 200 mg twice daily. A follow-up in three months revealed that the plaques had started to flatten out and the treatment was continued for a further three months. The patient was satisfied and happy with the response to treatment and improvement in her rash.

DISCUSSION

AEGCG is an uncommon granulomatous skin reaction typically involving sun-exposed skin areas. Pathogenesis is thought to be linked to UV radiation inducing cellular damage to elastin fibers and the formation of granulomas.[1-4]

Clinically, this condition presents as annular erythematous plaques with central clearing or atrophy.[1-3] Differentials include necrobiosis lipoidica and granuloma annulare, which can be distinguished via clinical and histopathological assessment.[2]

The gold standard for diagnosis is based on histopathology showing granulomatous inflammation, multinucleated giant cells with more than one area of elastophagocytosis and absence of mucin deposition or necrobiosis.[2,5]

There is no standard treatment, and clinical outcomes can be variable.[2] Treatment options can range from topical therapy with corticosteroids, to phototherapy including narrow band ultraviolet B to systemic medications including oral antimalarial such as hydroxychloroquine and quinine.[2]

CONCLUSION

Diagnosis of AEGCG is based on histopathological findings. This case illustrates the value of hydroxychloroquine as a treatment option, especially in cases not responding to topical therapy.

ACKNOWLEDGEMENTS

The authors would like to thank Dr. Rand Hawari (Consultant Histopathologist), University Hospitals of Derby and Burton NHS Foundation Trust.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Vickneswaran K, Nambi R. Clearing the circles: an unusual way. Int J Res Med Sci 2025;13:823-5.