Annular erythematous plaques in the Marianas

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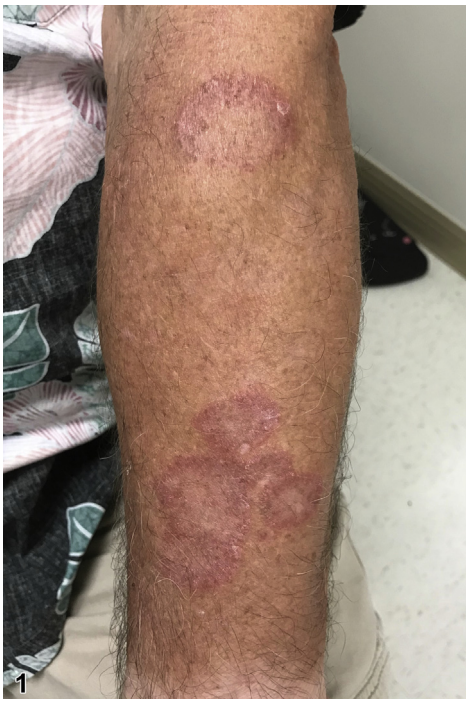
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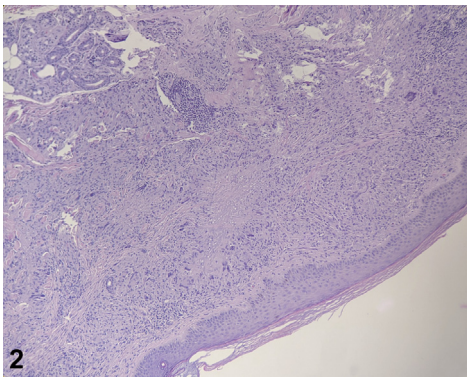
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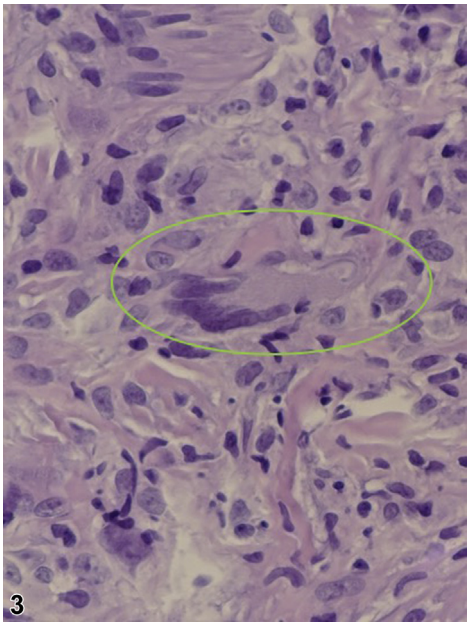
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A 66-year-old man with type 2 diabetes mellitus working as a tree farmer in the Northern Mariana Islands, western Pacific Ocean presented with a 12-month history of several 1- to 5-cm asymptomatic annular erythematous scaly plaques on the left forearm (Fig 1). He denied associated bleeding, discharge, pain, or pruritus and had trialed triamcinolone 0.1% ointment and ketoconazole 2% cream without improvement. A left forearm punch biopsy was obtained. Although special staining was unavailable locally, the diagnosis was made based on clinical and histologic data (Figs 2 and 3). The patient's annular plaques were treated with a trial of intralesional kenalog, 40 mg/mL injections, with notable response.

Question 1. Based on clinical and histologic findings, which leading diagnosis should be considered in this patient?

A. Granuloma annulare (GA)

B. Subacute cutaneous lupus erythematous (SCLE)

C. Majocchi granuloma

D. Tuberculoid leprosy

E. Annular elastolytic giant cell granuloma (AEGCG)

Answers:

A. GA — Incorrect. Although localized GA may share a similar clinical presentation, histology findings show foci of altered collagen, increased mucin deposition, and necrobiosis with surrounding histiocytes, lymphocytes, and fibroblasts, often in palisading array.

B. SCLE — Incorrect. Predominately afflicting white women around age 40, annular SCLE often begins with small, erythematous papules with slight scale that progress to form larger polycyclic patterns involving the upper torso and extensor surfaces of the upper extremities. Histopathology shows hyperkeratosis, focal degeneration of the basal layer, and perivascular lymphocytic infiltrate.

C. Majocchi granuloma — Incorrect. Majocchi granuloma would be clinically marked by focal erythematous perifollicular papules or nodules, often located at sites of frequent trauma. Histology findings show perifollicular granulomatous inflammation; the granulomatous inflammation in this case was not perifollicular.

D. Tuberculoid leprosy — Incorrect. Although presenting as a solitary erythematous plaque, histology is characterized by epithelioid cell granuloma involvement along neurovascular bundles.

E. AEGCG — Correct. The hallmarks of AEGCG are annular erythematous plaques with central clearing (Fig 1) in the setting of distinct histologic findings of multinucleated giant cells with elastolysis and elastophagocytosis. Although elastin staining is required for definitive diagnosis, other features on the Hematoxylin-eosin slide (Fig 3), including rare intracytoplasmic elastin fibrils, aid in the diagnosis.[1] Other features include the absence of necrobiosis and mucin deposition.

Question 2. Given resource limitations, only select special stains were available. Based on the initial histologic findings in Fig 3, which stain would be most helpful in consolidating this diagnosis?

A. Bodian stain

B. Verhoeff-van Gieson stain

C. Ziehl-Neelsen stain

D. Grocott methenamine silver stain

E. Alcian blue stain

Answers:

A. Bodian — Incorrect. Bodian staining is useful for the evaluation of nerve fibers.

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B. Verhoeff-van Gieson — Correct. AEGCG was confirmed via follow-on Van Gieson staining in the United States, which showed elastophagocytosis at the periphery of the granuloma and a paucity of elastic fibers in the center.

C. Ziehl-Neelsen — Incorrect. Ziehl-Neelsen staining is used to identify acid-fast organisms. Although a mycobacteria infection should be excluded in the presence of dermal granulomas, and were in this case, rare cytoplasmic fibrils on histology suggest an elastolytic process.

D. Grocott methenamine silver — Incorrect. Filamentous fungi were not evident on histology. Fungal organisms would have stained black and are larger (2 μm for dermatophytes, 75 μm for Aspergillus niger) when compared with elastin fibrils (typically 0.1-0.2 μm). In this case, Grocott methenamine silver was negative.

E. Alcian blue stain — Incorrect. Despite the amphophilic background, one of the characteristics of AEGCG is the lack of mucin deposition, which would appear amphophilic but as a fine, stringy substance.

Question 3. All of the following are proposed to be effective in the treatment of this condition, with the exception of:

A. Topical or intralesional corticosteroids

B. Cyclosporine

C. Hydroxychloroquine

D. Dapsone

E. Rifampin

Answers:

A. Corticosteroids — Incorrect. First-line treatment options for AEGCG includes topical and intralesional steroids.

B. Cyclosporine — Incorrect. AEGCG lesions have been responsive to an 8-week trial of cyclosporine A at 5 mg/kg/d.[2]

C. Hydroxychloroquine — Incorrect. A trial of hydroxychloroquine, 200 mg/d for 22 weeks has successfully resolved AEGCG lesions.[3]

D. Dapsone — Incorrect. A trial of dapsone, 50 mg/d successfully treated AEGCG in a 72-year-old fisherman with a history of hepatitis C infection without evidence of recurrence 1 year later.[4]

E. Rifampin — Correct. Although rifampin shows efficacy as a therapeutic agent in granuloma annulare and leprosy, this agent has not been successful in the treatment of AEGCG.[5]

Abbreviations used:

AEGCG: annular elastolytic giant cell granuloma
GA: granuloma annulare
SCLE: subacute cutaneous lupus erythematous

REFERENCES

1. Klemke CD, Siebold D, Dippel E, Hildenbrand R, Bleyl U, Goerdt S. Generalised annular elastolytic giant cell granuloma. *Dermatology*. 2003;207(4):420-422.
2. Tsutsui K, Hirone T, Kubo K, Matsui Y. Annular elastolytic giant cell granuloma: response to cyclosporin A. *J Dermatol*. 1994;21(6):426-429.
3. Babuna G, Buyukbabani N, Yazganoglu KD, Baykal C. Effective treatment with hydroxychloroquine in a case of annular elastolytic giant cell granuloma. *Indian J Dermatol Venereol Leprol*. 2011;77(1):110-111.
4. Igawa K, Maruyama R, Katayama I, Nishioka K. Anti-oxidative therapy with oral dapsone improved HCV antibody positive annular elastolytic giant cell granuloma. *J Dermatol*. 1997;24(5):328-331.
5. Marcus DV, Mahmoud BH, Hamzavi IH. Granuloma annulare treated with rifampin, ofloxacin, and minocycline combination therapy. *Arc Dermatol*. 2009;145(7):787-789.