

Benign Atrophic Papulosis Revealing Undiagnosed Systemic Lupus Erythematosus and Antiphospholipid Syndrome: A Rare Case of Secondary Degos Disease

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Introduction

- **Benign atrophic papulosis (BAP)**, or Degos disease, is a rare occlusive vasculopathy with porcelain-white, atrophic papules and telangiectatic rims.
- Systemic lupus erythematosus (**SLE**) and antiphospholipid syndrome (**APS**) cause immune complex-mediated vasculopathy and thrombosis.
- **Coexistence** of Degos-like lesions with SLE and APS is **rare yet clinically significant**.

Case Presentation

- **62-year-old woman** with obesity, diabetes, and tobacco use.
- **Tender erythematous papules** and plaques on trunk and extremities, many with **central necrosis** and crusting.
- Lesions evolved into **porcelain-white atrophic papules** with **telangiectatic rims** within two weeks.
- **Labs**
 - ANA: positive, 1:640
 - Anti-dsDNA: elevated
 - Anti-phospholipid antibodies: positive
- **Biopsy:** wedge-shaped dermal necrosis and thrombosed vessels, later showing dermal fibrosis and epidermal atrophy → **benign atrophic papulosis**
- **Treatment:** antiplatelet & anticoagulant therapy, hydroxychloroquine, and methotrexate.

Clinical Presentation



Figure 1 (A) Numerous erythematous papules and plaques with central necrosis and crusting on the bilateral thighs, consistent with an occlusive vasculopathy. **(B)** Evolution to porcelain-white, atrophic papules with telangiectatic rims consistent with benign atrophic papulosis.

Dermoscopy and Histopathology



Figure 2. Dermoscopic appearance of benign atrophic papulosis lesion. Shows a central porcelain-white atrophic area with a surrounding rim of light brown to erythematous pigmentation, consistent with the characteristic targetoid pattern of Degos-like lesions.

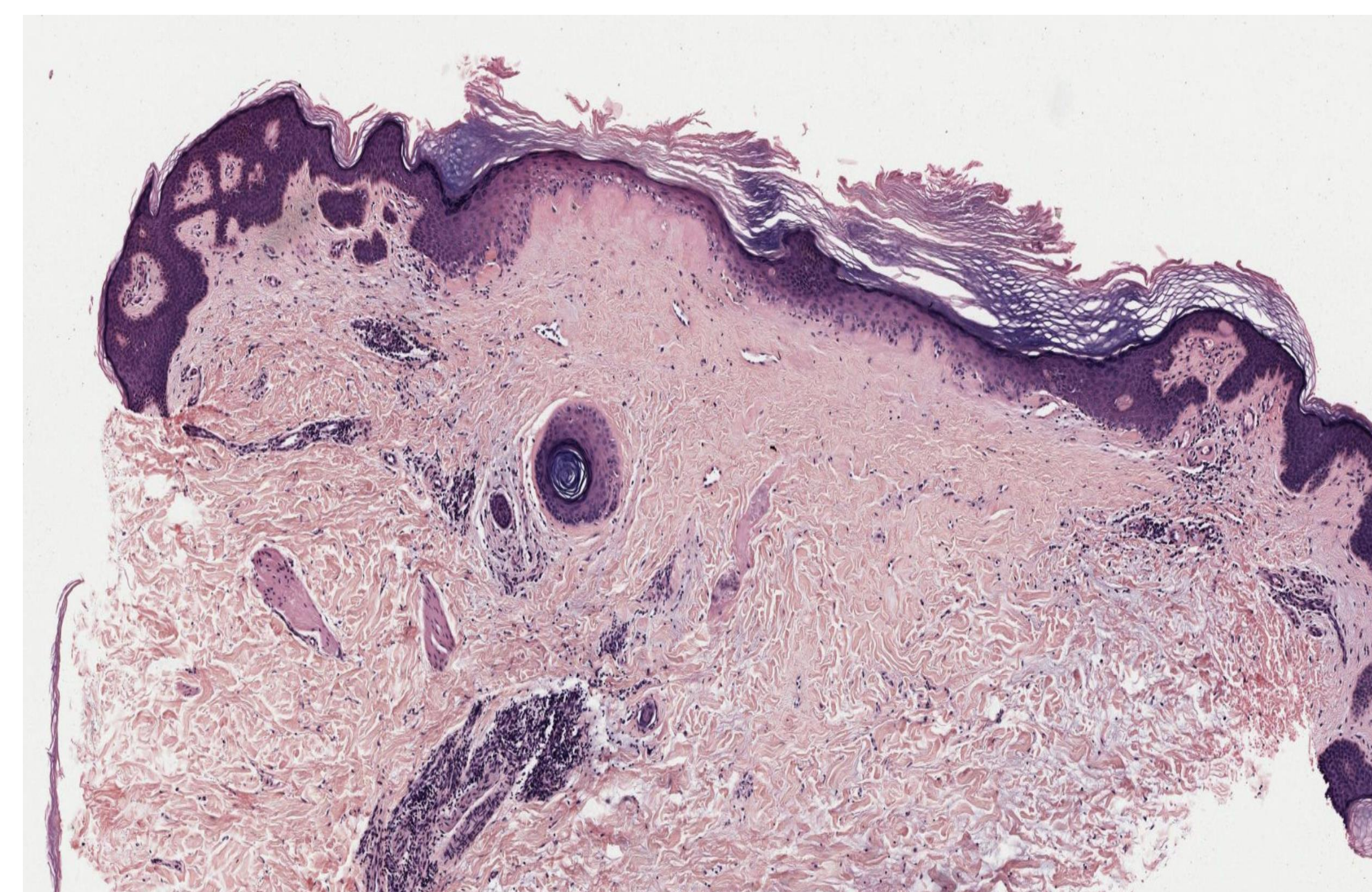


Figure 3. Histopathology of benign atrophic papulosis lesion Epidermal atrophy with underlying dermal fibrosis and mild perivascular lymphocytic infiltrate, consistent with Degos-like changes in benign atrophic papulosis.

Discussion

- Recognition of Degos-like lesions should prompt **evaluation for secondary systemic disorder, such as autoimmune and thrombotic disorders**.
- Early treatment can **prevent systemic progression and detect catastrophic vascular neurologic and GI events**.
- Emphasizes the **dermatologist's role** in recognizing cutaneous signs of systemic disease and ensuring **multidisciplinary management**.

Conclusion

- Benign atrophic papulosis (BAP) can serve as an **early cutaneous marker of systemic autoimmune and thrombotic disease**.

References

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