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1. Atrophoderma of Pasini and Pierini

Atrophoderma of Pasini and Pierini (APP) is a rare idiopathic condition of dermal atrophy, which primarily affects women in their adolescence and young adulthood.<sup>1</sup> First described by Pasini in 1923<sup>2</sup> and Pierini and Vivoli in 1936,<sup>3</sup> the disease went by many names, including atrophic scleroderma d’emblem and dyschromic and atrophic variation of scleroderma, until the official diagnosis of APP was suggested in 1958.<sup>4</sup>

The incidence of APP is unknown. The condition is rare, but some professionals suggest that it is merely underdiagnosed because it is largely asymptomatic. Diagnosis is based on clinical findings; however, histology can play a role in excluding other diagnoses.

Asymptomatic round or oval depressed lesions develop insidiously over many years. They are most commonly seen on the trunk—primarily the lower back and abdomen—and can spread unilaterally or in a bilaterally symmetric distribution. Zosteriform distribution has also been observed.<sup>1</sup> Affected skin is described as having an inverted plaque appearance, which Stoner and Dixon described as resembling “footprints in the snow.”<sup>5</sup> The lesions can be bluish-violet, slate-gray, or brown in colour and have irregular, sharply demarcated borders. Initially, single or multiple patches, 2 to 3 cm in diameter, appear on the skin. Although the lesions start out small, they can grow and coalesce to encompass large areas of the trunk and proximal extremities.

Over time, pigmentation of affected areas fades and atrophic characteristics develop; lesions typically sink 1 to 2 mm from adjacent unaffected areas. The resulting sharply demarcated edges, referred to as “cliff drop” borders, tend to be less abrupt on abdominal lesions and more obvious on the back and proximal extremities. Generally, lesions become quiescent upon reaching this atrophic phase, and tend not to progress or enlarge further.

Differential diagnosis

Although debate continues, most of the recent literature indicates that APP and morphea are related. Buechner and Ruffi describe APP as an “abortive, primarily atrophic variant of morphea”<sup>6</sup> and Amano et al believe that morphea and APP are a “similar nosologic entity.”<sup>7</sup> Although similarly atrophic in nature, the conditions can be distinguished by several features. Morphea tends to have a later age of onset, occurring mainly in the third through fifth decades of life. Morphea runs a shorter course than APP, burning out after 3 to 5 years compared with APP’s 10- to 20-year progression. Morphea plaques are ivory with a lilac border and tend to appear more inflamed, exhibiting induration, edema, and erythema.<sup>5</sup> The atrophic lesions of APP are violaceous or brown with sharply demarcated borders and no evidence of ongoing inflammation.

Further debate also exists over the relationship of the disease to *Borrelia* infections. Past studies have found that


28.5% to 53% of patients with APP also have anti-*Borrelia burgdorferi* antibodies compared with 14% of control subjects.<sup>6,8</sup> The role of *B burgdorferi* in APP is unclear, but some physicians will use positive test results for *B burgdorferi* as support for treating APP with antibiotics.

Histologically, the main indicator of APP is attenuated dermal thickness. This can be assessed clinically by skin punch biopsy or, in research settings, by ultrasound scanning with linear transducer frequency of 13 MHz and penetration depth of 40 mm.<sup>9</sup> Other obvious histologic features include flattened rete ridges, normal or slightly atrophied epidermis, and normal eccrine sweat glands, sebaceous glands, and hair follicles. Collagen bundles in the mid and deep dermis might show evidence of edema.<sup>1</sup>

Management

No definitive treatment for APP exists. In 2006, Carter et al described a 38-year-old patient who responded dramatically to a 400-mg daily dose of hydroxychloroquine. The patient had positive antinuclear antibody serology, low levels of rheumatoid factor, and no Lyme disease antibodies detectable on serology, and experienced “notable improvement” in all lesions after 5 months and complete clearing of facial and trunk lesions after 1 year.<sup>10</sup>

Other treatment options include topical steroids and topical retinoids. For those with new-onset APP, oral antibiotics such as doxycycline or amoxicillin in doses appropriate for Lyme disease might be attempted. Sun precautions are important, as lesions can darken with sun exposure. Unfortunately, data in the current literature do not enable endorsement of any one particular treatment.

Although cosmetically distressing, APP is a benign condition. The challenge to family physicians lies mostly in the diagnosis. Once this has been established through careful history, physical examination, histologic evidence, or dermatology referral, reassuring patients that the condition is benign and will eventually cease to progress is the family physician’s best tool in the management of APP. 

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Competing interests

None declared

References

1. Pullara TJ, Lober CW, Fenske NA. Idiopathic atrophoderma of Pasini and Pierini. *Int J Dermatol* 1984;23(10):643-5.
2. Pasini A. Atrofodermia idioatica progressiva. *G Ital Dermatol* 1923;58:785.
3. Pierini L, Vivoli D. Atrofodermia progressiva (Pasini). *G Ital Dermatol* 1936;77:403-9.
4. Canizares O, Sachs P, Jaimovich L, Torres VM. Idiopathic atrophoderma of Pasini and Pierini. *AMA Arch Derm* 1958;77(1):42-60.
5. Stoner MF, Dixon SL. Purple depressions on the trunk and extremities. Atrophoderma of Pasini and Pierini. *Arch Dermatol* 1990;126(12):1641,1644.
6. Buechner SA, Ruffi T. Atrophoderma of Pasini and Pierini. Clinical and histopathologic findings and antibodies to *Borrelia burgdorferi* in thirty-four patients. *J Am Acad Dermatol* 1994;30(3):441-6.
7. Amano H, Nagai Y, Ishikawa O. Multiple morphea coexistent with atrophoderma of Pasini-Pierini (APP): APP could be abortive morphea. *J Eur Acad Dermatol Venereol* 2007;21(9):1254-6.
8. Wojas-Pelc A, Wielowieyska-Szybinska D, Kieltyka A. Presence of the antinuclear antibodies and antibodies to *Borrelia burgdorferi* among patients with morphea en plaque, deep linear scleroderma and atrophoderma Pasini-Pierini [in Polish]. *Przegląd Lekarski* 2002;59(11):898-902.
9. Kim SK, Rhee SH, Kim YC, Lee ES, Kang HY. Congenital atrophoderma of Pasini and Pierini. *J Korean Med Sci* 2006;21(1):169-71.
10. Carter JD, Valeriano J, Vasey FB. Hydroxychloroquine as a treatment for atrophoderma of Pasini and Pierini. *Int J Dermatol* 2006;45(10):1255-6.