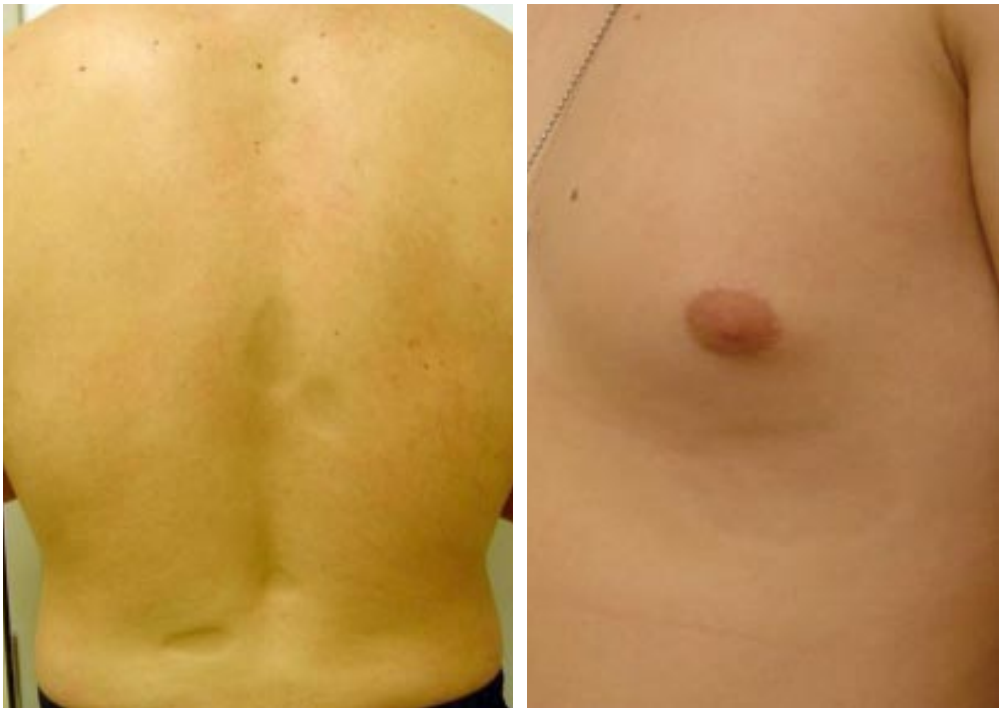
What Is Your Diagnosis?



A 19-year-old otherwise healthy man presented with a 2.5-month history of progressively enlarging, asymptomatic, depressed patches on his back and chest. An elliptical incision biopsy specimen of affected and healthy skin showed the affected dermis to be only half as thick as the unaffected dermis.

PLEASE TURN TO PAGE 475 FOR DISCUSSION

Sarah M. Howell, MD; Stephen J. Krivda, MD; Dermatology Service, Walter Reed Army Medical Center, Washington, DC.

The authors report no conflict of interest.

The opinions expressed are those of the authors and should not be construed as official or as representing those of the US Department of the Army or the US Department of Defense.

The Diagnosis: Idiopathic Atrophoderma of Pasini and Pierini

Idiopathic atrophoderma of Pasini and Pierini (IAPP) is a form of dermal atrophy presenting either as a solitary patch or multiple sharply demarcated, depressed patches occurring most frequently on the back in adolescents or young adults in the second and third decades of life. It still is debated if IAPP is a variant of morphea or its own separate disease entity.[1] The etiology of IAPP is unknown. However, *Borrelia burgdorferi* infection may play some role, as serologic tests were positive for the bacterial species in up to 53% (18/34) of patients with IAPP in one study.[2] The lesions occur most frequently on the back (Figure 1) but also may present on the chest (Figure 2), arms, and abdomen. The face, hands, and feet usually are spared.[3] The lesions range from a few centimeters in diameter to plaques covering the entire trunk. The plaques usually have a brown coloration. The surface of the skin is otherwise healthy in appearance and has average consistency on palpation. There is no induration or sclerosis. The skin surrounding the plaques is healthy in appearance, unlike morphea with a pinkish or purplish halo. However, typical lesions of morphea, lichen sclerosus et atrophicus, and IAPP have simultaneously occurred at different body sites in the same patient, indicating these conditions may be related.[3] Idiopathic atrophoderma of Pasini and Pierini may be progressive and lesions can continue to appear and spread throughout many decades of life. Although cosmetically displeasing, this disease is benign and there has been no association with any internal malignancies or systemic diseases.

In our patient, an elliptical incision biopsy specimen from affected skin showed a reduced dermal thickness compared to adjacent healthy skin (Figure 3). The epidermis and subcutaneous tissue were unaffected. Basal cell pigmentation may be increased. A perivascular infiltrate of T cells also may be seen.[4] A punch biopsy of only the affected skin may appear unaffected without comparison to healthy skin. Therefore, an elliptical incision biopsy specimen including unaffected skin is preferred.

Differential diagnosis of IAPP includes morphea linearis, generalized morphea, superficial morphea, morphea profunda, extragenital lichen sclerosus et atrophicus, and anetoderma. Morphea linearis often presents in the first decade of life, and the linear lesions typically follow the Blaschko lines. The lesions may extend down an entire extremity; therefore, physical therapy is important to prevent contractures and frozen joints. Generalized

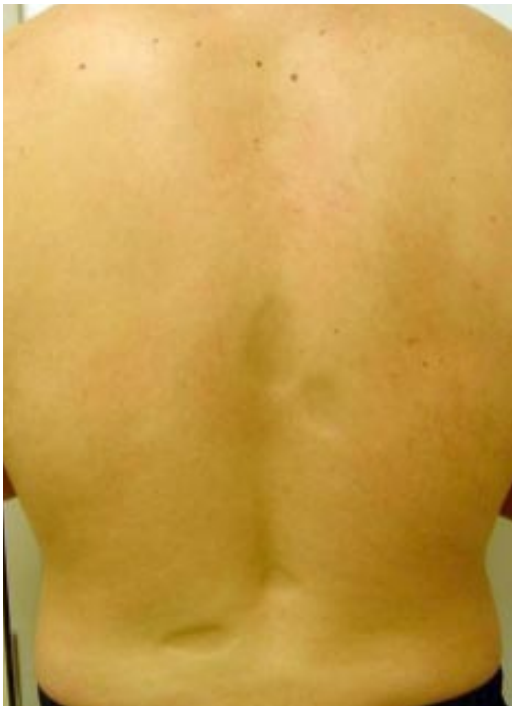


Figure 1. Depressed patches on the back.

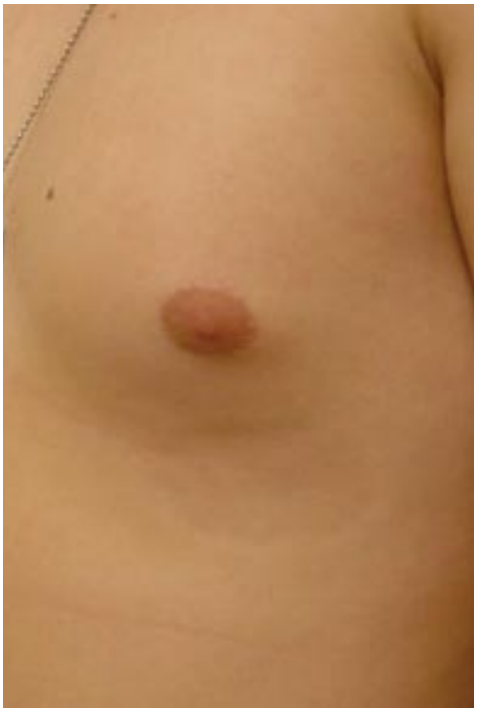


Figure 2. Depressed patch on the chest.

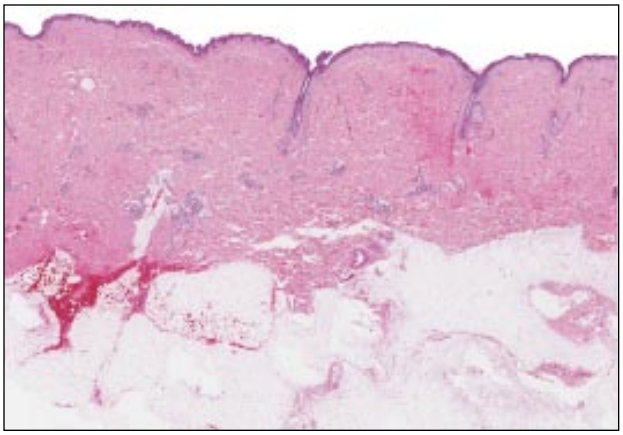


Figure 3. An elliptical incision biopsy specimen with healthy skin (left) and skin affected by idiopathic atrophoderma of Pasini and Pierini (right)(H&E, original magnification ×2).

morphea may appear similar to IAPP clinically, but the pathology is different. Generalized morphea usually has a squared off punch biopsy due to dermal sclerosis. The dermis replaces part of the subcutaneous tissue and the subcutaneous tissue that normally surrounds adnexal structures is lost. Morphea profunda is a deeper process involving the subcutaneous tissue and fascia. Clinically, extragenital lichen sclerosus et atrophicus causes similar lesions to IAPP that also are asymptomatic and appear most commonly on the back and chest. However, the pathology of lesions associated with extragenital lichen sclerosus et atrophicus is completely different from IAPP lesions. With lichen sclerosus et atrophicus, the epidermis is atrophic and there is a lichenoid band of lymphocytes in a washed-out–appearing dermis. Anetoderma is caused by a localized loss of elastic fibers in the dermis and results in flaccid protruding skin due to herniation of the subcutaneous tissue, as opposed to depressed patches of IAPP.[5]

No treatment has been proven to be consistently effective. In one study, patients were treated with oral penicillin or tetracycline for 2 to 3 weeks and 20 of 25 patients showed improvement.[2] In theory, oral antibiotics would be effective if *B burgdorferi* is the cause of IAPP and the antibiotics are treating the underlying infection. In one case, the Q-switched alexandrite laser was effective in decreasing hyperpigmentation, but it did not help with atrophy.[6]

Our patient's plaques presented immediately after beginning a rigorous basic training program in the military. The patient had no remarkable medical or surgical history. He denied taking any medications and had no known drug allergies. The patient also denied any family history of skin disease. The histologic sections showed a healthy epidermis and a thinned dermis. The affected portion of the biopsy specimen showed a dermis that measured 0.3 cm in thickness, whereas the dermis of the unaffected portion measured 0.7 cm. There was no evidence of sclerotic dermal collagen bundles, perivascular inflammatory cellular infiltration, vascular thickening, or basilar hyperpigmentation. Staining for $CD34^+$ showed no difference between the affected and unaffected portions of the biopsy specimen. The patient's Lyme (IgM/IgG) antibody screen was 0.32 (reference range, 0.00–1.09), and results of the *B burgdorferi* DNA polymerase chain reaction were negative. The patient was administered an empiric 21-day course of oral doxycycline hyclate 100 mg twice daily to treat possible *B burgdorferi* infection. The patient was lost to follow-up prior to completion of his treatment.

REFERENCES

1. Maari C, Powell J. Atrophies of connective tissue. In: Bolognia JL, Jorizzo JL, Rapini RP, eds. *Dermatology*. New York, NY: Mosby; 2003:1539-1548.
2. Buechner SA, Rufli T. Atrophoderma of Pasini and Pierini. clinical and histopathologic findings and antibodies to *Borrelia burgdorferi* in thirty-four patients. *J Am Acad Dermatol*. 1994;30:441-446.
3. Wakelin SH, James MP. Zosteriform atrophoderma of Pasini and Pierini. *Clin Exp Dermatol*. 1995;20:244-246.
4. Yokoyama Y, Akimoto S, Ishikawa O. Disaccharide analysis of skin glycosaminoglycans in atrophoderma of Pasini and Pierini. *Clin Exp Dermatol*. 2000;25:436-440.
5. James W, Berger T, Elston D, eds. *Andrews' Diseases of the Skin: Clinical Dermatology*. 10th ed. Philadelphia, PA: Saunders Elsevier; 2006.
6. Arpey CJ, Patel DS, Stone MS, et al. Treatment of atrophoderma of Pasini and Pierini–associated hyperpigmentation with the Q-switched alexandrite laser: a clinical, histologic, and ultrastructural appraisal. *Lasers Surg Med*. 2000;27:206-212.