

What Is Your Diagnosis?



A 14-year-old adolescent boy presented with a 2-year history of asymptomatic, soft, outpocketing papules on his abdomen. The lesions disappeared when in a supine position.

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The author reports no conflict of interest.

The Diagnosis: Primary Anetoderma

The patient presented to the clinic with unusual skin bumps (Figures 1 and 2). Prior to their appearance, he denied any other skin abnormalities, such as inflammation, erythema, or urticaria, on his abdomen. Upon examination, the patient had average mental development and no physical abnormalities. Skin examination revealed lesions that were soft saclike papules on the abdomen only, varying from 0.5 to 2 cm (Figure 3). There were no sensory changes associated with the lesions. Otherwise, the patient's general health was good. He did not have a history of medication consumption nor did he have any coexisting medical conditions or allergies to medication or food. His mother denied any known family history of abnormal skin conditions

or immunologic disorders. Results of the patient's laboratory workup were negative or within reference range, specifically for human immunodeficiency virus, rapid plasma reagin, cardiolipin IgG and IgM, C3, C4, antinuclear antibody titer, lupus anticoagulant, protein C and S, thyroid-stimulating hormone, and protein electrophoresis.

A 4-mm punch biopsy of 2 of the lesions was performed. The results for both biopsies showed decreased dermal elastic fibers. More specifically, elastic stains suggested a decrease in elastic fibers in the mid dermis. Periodic acid-Schiff stains were negative for fungal pathogens. There was no appreciable alteration of or thickening of collagen bundles. The differential diagnosis included anetoderma, connective tissue nevus, and nevus anelasticus.

Anetoderma is a benign uncommon condition, with focal loss of dermal elastic tissue resulting in localized areas of flaccid or herniated saclike skin.¹ The lesions usually occur in individuals aged 15 to 25 years and are more predominant in females than males.²

Pathologically, there is complete loss of elastic tissue in the papillary and/or mid reticular dermis. Chaby et al³ found that anetoderma is characterized by focal loss of elastic tissue in the mid dermis, which was revealed in our patient. Benest et al⁴ found that histologic variations in inflammatory infiltrate and elastic tissue loss did not correlate with clinical appearance, disease course, or associated diseases. Perivascular lymphocytes often are present and do not correlate with clinical inflammatory findings. Monoclonal antibody studies have shown that the majority of lymphocytes in these infiltrates are helper T cells.⁵ The pathogenesis of anetoderma is not known. Many theories have been considered to explain the underlying mechanisms of focal elastin destruction, but no single consistent theory has been proven.

Primary anetoderma arises on skin that is not clinically affected by an underlying associated disorder.⁶ Anetoderma is broadly and historically categorized into 2 clinical types: (1) Jadassohn-Pellizzari anetoderma, preceded by macular erythema or papular urticaria, and (2) Schweninger-Buzzi anetoderma, with lesions appearing without preceding erythema or



Figure 1. Soft outpocketing papules on the abdomen of a 14-year-old adolescent boy.



Figure 2. Lesions disappeared when the patient was in a supine position.



Figure 3. Lesions only appeared on the abdomen when the patient was in a prone position.

inflammation. Although several revisions in terminology have occurred, it is now generally accepted that secondary anetoderma refers to lesions that follow an inflammatory dermatosis in the same location, while primary anetoderma refers to the occurrence of atrophic lesions in areas of skin that appear unaffected prior to onset of atrophy.⁶ Both types of anetoderma can coexist in the same patient, as the histopathology is the same, and inflammation has been known to exist in both types of lesions; the presence of or absence of clinical inflammation at the onset of the disease is not related to prognosis.⁴ True secondary anetoderma implies that the characteristic atrophic lesion has appeared in the same site as a previous specific skin lesion. Some researchers also consider lesions associated with an underlying disease as secondary anetoderma; however, with an underlying disease, the atrophic areas do not necessarily develop within known areas of inflammation.²

The clinical features of primary and secondary anetoderma are the same.² Clinical manifestations of anetoderma are characterized by oval plaques with overlying loose skin appearing as depressions or flabby pouchlike protrusions. Areas of predilection include the trunk and proximal extremities. In this patient, lesions were outpocketing saclike papules found only on the abdomen, with no preceding skin lesions (Figure 3).

A study by Peterman et al⁷ described a family of 4 individuals with anetoderma as the only manifestation. Familial forms of anetoderma are rare and have been associated with ocular, bony, or neurologic changes. Only 6 families have been previously reported with a familial form of anetoderma.⁷

Sparsa et al⁸ reported frequent observation of coexisting autoimmune diseases in patients with primary anetoderma, particularly with antiphospholipid antibodies. True secondary anetoderma, with pouched lesions that have developed on the sites of inflammation, can occur after many dermatoses. The most common causes of secondary anetoderma today are probably acne and varicella.⁸

In researching case reports of anetoderma in the literature, numerous systemic conditions, as well as cutaneous tumors, were found to be coexisting disorders in patients with anetoderma, including syphilis, tuberculosis, *Borrelia anserina* infection, urticaria pigmentosa, lupus erythematosus, human immunodeficiency virus, sarcoidosis, antiphospholipid syndrome, leprosy, acrodermatitis chronica atrophicans, mastocytosis, nodular prurigo, granuloma annulare, xanthomas, pilomatrixomas, and cutaneous T- or B-cell lymphomas. Researchers have found that anetoderma is rarely secondary to lupus profundus.⁸ Chaby et al³ also reported associations of anetoderma with cutaneous tumors. Their findings showed that associations of malignant cutaneous tumors with secondary anetoderma are rare. Secondary anetoderma usually is associated with cutaneous infections and benign skin tumors.³

In 1998, Braun et al⁹ reported that numerous therapeutic approaches for anetoderma have been proposed, but the results have frequently been disappointing. They described a patient with primary anetoderma with an inflammatory component responsive to colchicine. Their patient was given a 1-mg daily dosage of oral colchicine. Within 2 weeks, no new inflammatory lesions were present and the atrophic noninflammatory lesions remained unaffected. After 7 months, the treatment was discontinued, and shortly thereafter, new inflammatory lesions began to appear. In their discussion, Braun et al⁹ stated that proposed treatments for anetoderma include intralesional corticosteroids, oral penicillin G, phenytoin, dapsone, vitamin E, and nicotinate. Most treatments have been unsuccessful or of little benefit. Reiss and Linn¹⁰ reported a partial response to epsilon-aminocaproic acid that prevented new inflammatory lesions in a patient with primary anetoderma. In most patients, despite treatment, disease activity persists for at least 15 years. Multiple treatment modalities, such as penicillin, phenytoin, dapsone, vitamin E, and nicotinate, have been unsuccessful.¹¹

REFERENCES

1. Aghaei S, Sodaifi M, Aslani FS, et al. An unusual presentation of anetoderma: a case report. *BMC Dermatol.* 2004;4:9.

2. Maari C, Powell J. Atrophies and disorders of dermal connective tissue. In: Bologna JL, Jorizzo JL, Rapini RP, eds. *Dermatology*. Philadelphia, PA: Elsevier Ltd; 2003: 1539-1548.
3. Chaby G, Viseux V, Chatelain D, et al. Myxofibrosarcoma associated with anetoderma [in French]. *Ann Dermatol Venereol*. 2006;133:35-37.
4. Benest L, Kwark E, Goldwasser J, et al. Primary anetoderma. *Cutis*. 2000;65:188-190.
5. Chapman MS, Perry AE, Baughman RD. Off-center fold: generalized flesh-colored pouchlike papules. *Arch Dermatol*. 2001;137:657-662.
6. Lewis KG, Bercovitch L, Dill SW, et al. Acquired disorders of elastic tissue: part II. decreased elastic tissue. *J Am Acad Dermatol*. 2004;51:165-185.
7. Peterman A, Scheel M, Sams WM Jr, et al. Hereditary anetoderma. *J Am Acad Dermatol*. 1996;35:999-1000.
8. Sparsa A, Piette JC, Wechsler B, et al. Anetoderma and its prothrombotic abnormalities. *J Am Acad Dermatol*. 2003;49:1008-1012.
9. Braun RP, Borradori L, Chavez P, et al. Treatment of primary anetoderma with colchicine. *J Am Acad Dermatol*. 1998;38:1002-1003.
10. Reiss F, Linn E. The therapeutic effect of epsilon-aminocaproic acid on anetoderma Jadassohn. *Dermatologica*. 1973;146:357-360.
11. Venencie PY, Winkelmann RK, Moore BA. Anetoderma. clinical findings, associations, and long-term follow-up evaluations. *Arch Dermatol*. 1984;120: 1032-1039.