Simultaneous Occurrence of Anetoderma in Premature Identical Twins

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Anetoderma, or macular atrophy, is a benign condition characterized by focal loss of dermal elastic tissue, resulting in localized areas of flaccid or herniated sac-like skin. Lesions appear on the upper arms, trunk and thighs. Anetoderma was traditionally classified into two types. In primary anetoderma atrophic lesions appear on previously normal skin, whereas secondary anetoderma is preceded by an inflammatory dermatosis in the same location in association with a variety of skin conditions or penicillamine use. This classification is of historical interest only because neither the prognosis of the disease nor its possible association with other medical problems was related to the presence or absence of inflammation. The exact cause of anetoderma is unknown.

We describe here the simultaneous occurrence of anetoderma on the abdomen of premature identical twins, supporting the hypothesis that genetic factors are involved in the development of the disease.

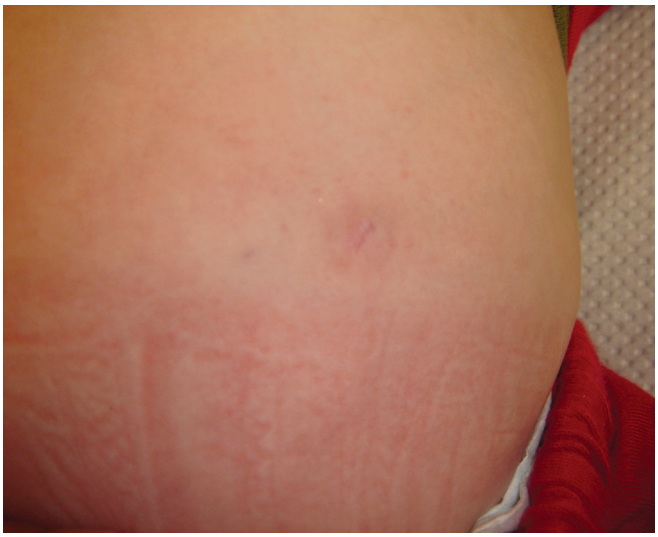
Patient Description

Identical male twins presented at age 3 months with a similar atrophic patch on the lower abdomen. The babies were born at 26 gestational weeks weighing 1200 and 1050 g. Their postnatal course was complicated by grade I intraventricular hemorrhage, respiratory distress syndrome, urinary tract infection, and retinopathy of prematurity. The family history was not contributory.

On clinical examination the babies appeared otherwise healthy. A well-circumscribed, round to oval-shaped depression measuring 20 mm in diameter was noted below the level of the normal skin on the abdomen of both babies [Figure]. There were no other signs of cutaneous disease, and no neurological, orthopedic or ophthalmological abnormalities.

Histopathological examination revealed mild hyperkeratosis, spongiosis, and mild interstitial infiltrate with lymphocytes in the mid-dermis. In addition, a decreased number of elastic fibers were visualized in the superficial and mid-dermis.

Screening for associated ocular, gastrointestinal, cardiac, pulmonary, endocrine and bone disorders revealed no abnormalities. Screening for systemic disorders such as systemic lupus erythematosus and antiphospholipid syndrome was negative. No new skin lesions were detected during follow-up of a year.



Well-circumscribed atrophic area just lateral to the umbilicus.

Comment

In 1992 Jadassohn was the first to describe anetoderma as a clinicohistopathologic entity. Clinically, anetoderma presents as a circumscribed area of slack skin and herniation. The histological changes consist of a severe reduction in skin elastic fibers with loss of amorphous elastin and persistence of microfibrils.

The exact cause is unknown. Possible explanations for the loss of dermal elastic tissue include defective elastin synthesis, uncontrolled production of elastolytic enzymes, elastophagocytosis, or loss of elastolytic enzyme inhibitors.

The old literature contains numerous reports of anetoderma in patients with lupus erythematosus, although the relationship between the disorders has not been clearly established. More recently, a growing body of literature has linked anetoderma with a wide range of immunological abnormalities, most commonly high levels of antiphospholipid antibodies with or without antiphospholipid syndrome.

Anetoderma can present at any age, although the majority of patients are in their teens and twenties and are usually female. Cases in children have also been reported. The occurrence of an-

etoderma in infancy is extremely rare; our review of the literature revealed only 12 cases [1-3].

Prizant et al. [1] described a previously unrecognized type of anetoderma in nine extremely premature neonates born between 24 and 29 weeks gestation (594–1531 g birth weight). In eight, the anetoderma developed during their stay in the neonatal intensive care unit, in the absence of any known preceding inflammatory lesions. All the anetodermas were located on the ventral surface of the trunk and the proximal limbs. All the infants had many of the complications associated with prematurity, and eight required ventilatory support for bronchopulmonary dysplasia. The authors suggested that the placement of adhesive monitoring leads may have caused the anetoderma and that the poor condition of the infants could have been a contributing factor. Colditz et al. [2] described two female infants who presented with anetodermas on the forehead that were associated with the use of electrocardiographic electrodes. Both had been born at 27 gestational weeks (birth weight 630 and 529 g), and both were growth-retarded (5th centile and < 3rd centile, respectively). Another 48 premature infants with higher birth weight hospitalized in the same unit at the same time did not have anetodermal lesions.

Todd [3] reported a finding of isolated chest anetoderma in the smaller (794 g) of a pair of monozygotic dichorionic twins born at 32 weeks gestation. The larger twin (1600 g) had septicemia and required ventilatory support but did not have anetoderma. The author speculated that birth weight may be more important than prematurity or general condition as a risk factor for anetoderma of prematurity [3]. It is possible that the reduced growth and thickness of the epidermis associated with intrauterine growth retardation contributes to the formation of anetodermas. However, since anetoderma of prematurity is rare compared to prematurity and low birth weight, we cannot draw any conclusions regarding the risk posed by these factors.

Familial anetoderma is uncommon. To the best of our knowledge, 10 families with at least 2 immediate family members with anetoderma have been reported in the literature. Peterman and colleagues [4] described a Caucasian family with four affected members. These reports suggested an autosomal dominant trait, an autosomal recessive trait, autosomal dominant transmission with incomplete penetrance of the gene, and undefined traits.

Zellman and Moise [5] reported congenital anetoderma in monozygotic twins. Because both patients lacked evidence of either clinical or histopathological inflammation, the authors felt it unlikely that an autoimmune or inflammatory cell mediation process was involved in the pathogenesis.

Our findings support abnormalities in identical twins, reinforcing the possible importance of genetic factors in the development of anetoderma. However, since only two pairs of monozygotic twins concordant for anetoderma have been reported, genetic factors are probably not definitive by themselves.

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