Multiple flesh-colored asymptomatic plaques on the trunk: a case of primary anetoderma in a young adolescent

© D. LINDER1, E. MARINELLO2, G. BIOLO2, R. SALMASO3, S. PIASERICO2

1Medical University of Graz, Graz, Austria;
2Dermatology Unit, University of Padua, Padua, Italy;
3Pathology Unit, University of Padua, Padua, Italy

ABSTRACT
Our case concerns an asymptomatic Anetoderma in a 14 year old boy. Because both of the slightly uncommon clinical presentation (Anetoderma presents, to the best of our knowledge, much more often as depressed sharply delimited slack skin areas rather than as protruding papules/plaques, as in the present case) as well as of the possible need for follow-up or even for prophylaxis for thrombophilic states (possible association of Anetoderma with Antiphospholipid Syndrome) we feel that the presented case may be of interest to the readers.

Keywords: primary anetoderma, elastolytic disorders, case report.

Dennis Linder — MD, MSc, Privatdozent (Adjunct Professor), Medical University of Graz, Graz, Austria. https://orcid.org/0000-0002-0305-3146
Elena Marinello — Dermatology Unit, University of Padua, Padua, Italy
Giulia Biolo — Dermatology Unit, University of Padua, Padua, Italy
Roberto Salmaso — Pathology Unit, University of Padua, Padua, Italy
Stefano Piaserico — Dermatology Unit, University of Padua, Padua, Italy

TO CITE THIS ARTICLE:
https://doi.org/10.17116/kliderms20181706142

A 14-year-old boy presented to our clinic with multiple asymptomatic lesions, which had started appearing on his trunk 3 years before. The physical examination showed about ten sharply demarcated, flesh-colored, oval shaped plaques of about 4×8 mm on the abdomen and lumbar area (Fig. 1). The papules were slightly protruded with overlying wrinkled skin, and revealed a soft-elastic consistency on palpation. No other skin lesions or accompanying systemic symptoms were observed, and no previous trauma on the affected area was reported. The past medical history and family history were unremarkable, except for a thrombophilic status affecting the father and two siblings of the father, who all reported previous episodes of deep venous thromboses.

A biopsy was performed from one lesion on the lumbar area for histopathological examination, revealing a slight superficial perivascular lymphocytic infiltrate (Fig. 2). The elastic Verhoeff—Van Gieson stain showed a reduction and fragmentation of elastic fibers within the superficial and mid dermis (Fig. 3). Based on clinical and histological features, a diagnosis of primary anetoderma was made. These histopathological features were fundamental to rule out the differential diagnosis of popular elastorrhesis, a rare cutaneous disorder characterized by asymptomatic indurated white or flesh papules, that occurs predominantly during adolescence; in popular elastorrhesis the reduction in number and the fragmentation of elastic fibers involves only the mild and/or reticular dermis, sparing the superficial dermis [1].

We performed a screening for antiphospholipid antibodies and other thrombophilic factors, that revealed a slightly positivity of lupus anticoagulant (LAC) (1.4 ratio). Other risk factors were within normal range, antinuclear antibodies, extractable nuclear antigens and VDRL were negative.

Anetoderma is a rare elastolytic disorder associated with a loss of substance on palpation, as a result of focal destruction of elastic tissue in the superficial and mid-dermis. Predilection sites are the trunk and proximal portions of the extremities and, less commonly, neck and face. Anetoderma occurs mainly in women aged 20—40 years but is occasionally reported in younger patients of both sexes, mostly children and rarely in infants [2]. Its clinical presentation consists of small, sometimes atrophic papules, macules or plaques, i.e. sharply demarcated macular, papular or depressed areas of slack skin.

Primary anetoderma (PA) has traditionally been considered an idiopathic disorder to be distinguished from secondary anetoderma, in which elastolysis is associated with use of medications such as penicillamine and different systemic diseases such as infections and malignant disorders. Although the underlying pathogenesis of anetoderma is not fully established, it has been suggested that loss of elastic fibres was due to either degradation by elastase enzymes, such as proteases and matrix metalloproteinases, or reduced synthesis of elastic fibres [3].
In recent years, there has been a growing evidence linking PA with a wide range of immunological abnormalities. The first association described was between primary anetoderma and different forms of lupus, including discoid lupus erythematosus, systemic lupus erythematosus, and lupus panniculitis, but the relation has not been clearly established [4].

Moreover, there are isolated reports of PA and autoimmune diseases like primary hypothyroidism, Grave’s disease, Addison’s disease, Sjogren’s syndrome, alopecia areata, vitiligo, and multiple sclerosis, suggesting the presence anetoderma as a possible marker of autoimmunity [5, 6].

The most stressed association is between primary anetoderma and the presence of antiphospholipid antibodies (aPL), with or without antiphospholipid syndrome (APS) [4]. In the course of last decades many cases of primary anetoderma combined with positive aPL were reported, either as part of APS, or in conjunction with other clinical and/or laboratory signs of autoimmunity [4]. Nevertheless, the actual prevalence of aPL in PA remains controversial and unclear. In the study by E. Hodak et al. [7] comprising nine unselected PA patients, positive aPL were found in all, mostly of IgA isotype (anti-cardiolipin and/or anti-beta-2-glycoprotein). In two of these patients the PA presented as the first clinical sign, preceding by many years the classical signs of APS. The authors suggested that the work-up of these patients should include screening for antiphospholipid antibodies (anti-cardiolipin profile, anti-beta-2-glycoprotein, and LAC, and emphasized the need for long-term follow-up in PA patients; autoimmune diseases may develop later in the course of the disease, even years after the onset of the skin lesions. The pathophysiologic role of aPL in PA is still unknown. Some authors hypothesized that microthrombi, occasionally observed in the dermal vessels of PA lesion of patients with aPL, could provoke local ischemia and consequent degeneration of elastic tissue characteristic of anetoderma [8].

A. Sparsa et al. [9] suggested that, even in absence of microthrombi, the alternation of hypoxia-reoxygenation typical of this prothrombotic condition may trigger a local imbalance between metalloproteinases and their inhibitors, leading to the destruction of elastic tissue. According to other authors the destruction of elastic fibres may be immunologically mediated by aPL, that bind directly to an antigen epitope that is phospholipids-related, possibly apolipoprotein H [8].

Considering these growing evidences, periodic hematological follow-up should be considered to assess the potential risk of developing an APS in patients affected by anetoderma.
The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

Dennis Linder and Elena Marinello — conceived the idea and wrote the manuscript (they contributed equally)

Giulia Biolo — helped with implementation
Roberto Salmaso — histologic evaluation
Stefano Piaserico — revised the work critically

REFERENCES


Received 19.09.18
Accepted 14.11.18