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Alpha Tocopherol and Ascorbic Acid Treatment in a Child with Disseminated Jadassohn - Pellizzari Type Primary Anetoderma

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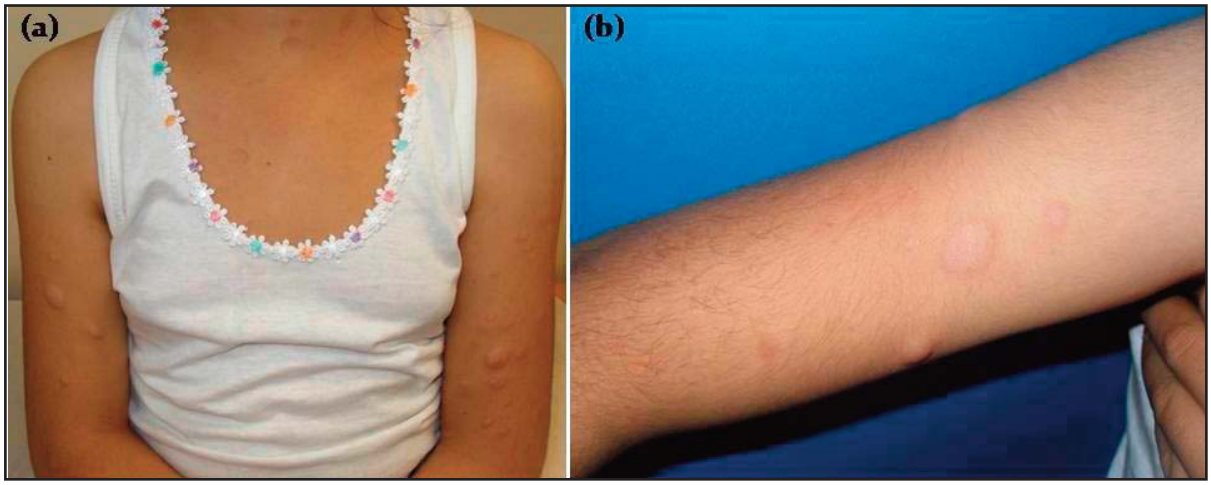
Abstract

Observation: Primary anetoderma is very uncommon in childhood. The exact pathogenesis and curative treatment of anetoderma are still unknown. We report a 12 -year-old girl presented with multiple elevated or herniated, circumscribed, skin-coloured papules which initially emerged as erythematous macules over her neck, trunk, upper and lower extremities. Histopathological examination of a papular lesion revealed mild perivascular lymphocytic inflammation in the mid-dermis, fragmented and loss of elastic fibres in dermis. No associated autoimmune disease was detected. Based on clinical and pathological findings, the patient was diagnosed as Jadassohn – Pellizzari type primary anetoderma. The patient was put on daily oral 500 mg ascorbic acid and 200IU alpha tocopherol. Lesions did not regressed; however, new lesions did not develop during 9-months of therapy. No adverse effect was detected. 3 months after ceasing antioxidant therapy, the patient had an upper respiratory tract infection preceding a few new anetoderma lesions. The antioxidants were re-administered promptly and no additional lesions were observed. Our case attracts attention to the possibility of oxidative stress accompanying systemic immunological responses that may cause anetoderma and to the efficacy of antioxidants in this disorder.

Introduction

Anetoderma is a rare elastolytic skin disorder characterized by typical loose macules and papules and subcutaneous tissue herniation due to focal loss of elastic fibres. When anetoderma develops without any associated underlying disease, it is referred to as primary anetoderma. Secondary anetoderma is related with eruptions of many dermatosis such as pilomatricoma, mastocytosis, generalized granuloma annulare, acne, or varicella. Primary anetoderma is classified into two subtypes as Jadassohn-Pellizzari type anetoderma following signs of inflammation preceding the anetoderma lesions and Schweninger-Buzzi type having no preceding inflammatory lesions. The exact pathogenesis of anetoderma is not elucidated [**1,2,3,4**]. Primary anetoderma is very uncommon in childhood. To the best of our knowledge, Jadassohn-Pellizzari type anetoderma in a child has not been reported yet. Herein, we reported a child with widespread Jadassohn-Pellizzari type primary anetoderma lesions and outcomes of oral alpha tocopherol and ascorbic acid treatment.



Figures 1a and b. (a) Multiple macules and papules on the neck and upper limbs, (b) close-up view of lesions

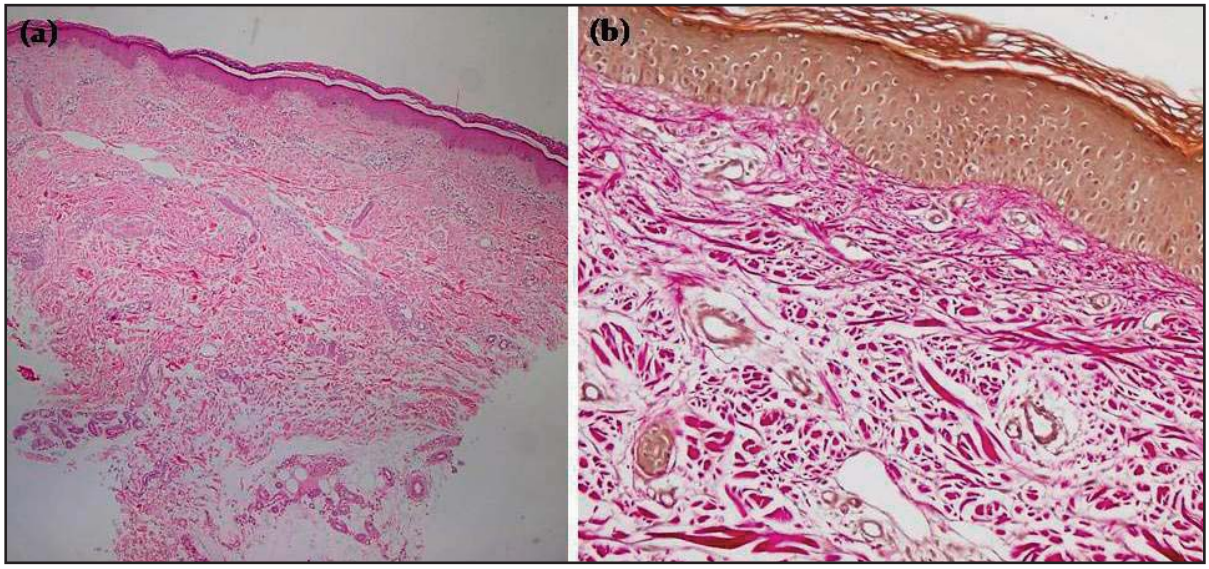


Figure 2a and b. (a) Mild perivascular lymphocytic infiltration in the lesional dermis (H&Ex40), (b) loss of elastic fibres in the superficial and mid-dermal lesional skin (EVGx200)

Case Report

A 12 -year-old girl who was complaining about soft skin puffinesses on her body lasting about 1 year admitted to our outpatient clinic. The lesions initially emerged as multiple erythematous macules over her neck, then spread over the trunk, upper and lower extremities. Most of them had become elevated or herniated, circumscribed, skin-coloured papules (**Figures 1a and b**). She denied any previous infection, vaccination, or trauma before the development of skin lesions. Her medical history was unremarkable. None of the family members had similar lesions.

Histopathological examination of a papular lesion revealed mild perivascular lymphocytic inflammation in the mid-dermis and fragmented and loss of elastic fibres in dermis (**Figures 2a and b**). Physical examinations of organ systems were normal. Echocardiographic measurements were within standard normal limits. Anti-RNP, anti-Sm, anti-SS-A, anti-SS-B, Scl-70, anti-Jo-1, anticardiolipin, antiphospholipid, and anti-Borrelia burgdorferi IgM and IgG antibodies, serology for HIV, hepatitis B and C were all negative. Based on clinical and pathological findings, the patient was diagnosed as Jadassohn – Pellizzari type primary anetoderma. The patient was put on daily oral 500 mg ascorbic acid and 200IU alpha tocopherol. Lesions did not regressed; however, new lesions did not develop during 9-months of therapy. No adverse effect was detected. 3 months after ceasing antioxidant therapy, the patient had an upper respiratory tract infection preceding a few new aneto-

derma lesions. The antioxidants were re-administered promptly and no additional lesions were observed.

Discussion

Anetoderma is a rare skin disorder, mainly defined in women aged 20-40 years. Anetoderma is very rare in children and age of onset of primary anetoderma is approximately 8 years among the child cases. Face, neck, trunk, upper and lower limbs are the most common body regions involved [**5**]. When the English dermatology literature was searched in Pubmed, there is no report about a child with Jadassohn-Pellizzari type. Since the preceding inflammation was clearly described before the anetoderma lesions developed, clinicopathological findings revealed this type of anetoderma in our case. Although classification of primary anetoderma depends on presence of signs of inflammation, definition of varying intensities of inflammatory infiltration in Schweninger-Buzzi type made some authors consider about a modified classification [**3**]. These authors subclassified primary anetoderma as idiopathic and secondary forms (associated with anti-phospholipid antibodies), irrespective of the presence of inflammation. We also agree with this kind of classification since inflammatory signs may be so subtle that patient or clinicians can easily miss out.

The etiopathogenesis of anetoderma is still unclear. Postinflammatory degradation of elastic fibres, increased extracellular matrix degeneration, decreased matrix production, and autoimmunity were suggested to involve in the pathogenesis of the disorder [**2, 3, 6**]. Activation of immunological mechanisms was accused as a causative factor when anetoderma lesions were observed after hepatitis B vaccination [**7**]. In our case, follow up of the patient showed us appearance of new lesions after upper respiratory tract infection although the patient denied any preceding infection in her first admission. This kind of disease progression suggested us that immune activation to microorganisms might have concurrently had effects to elastic fibres.

Many treatment strategies have been tried to control the disease activity and to make regression of skin lesions. Topical and intralesional steroids, oral penicillin G, phenytoin, dapsone, nicotinate were unsuccessful or provided only a little benefit in child cases [**1, 5**]. Colchicine with a dose of 1 mg/d restrained new lesions including neutrophilic infiltration in a 30-year-old male patient; however, ceasing the therapy resulted in progression [**8**]. Psoralen with ultraviolet A phototherapy was reported to provide excellent outcome in a 26-year-old patient with anetodermic mastocytosis [**4**]. It is well-known that oxidative stress involves in many systemic inflammatory conditions including skin and was suggested to contribute to the abnormal structure and function of elastic fibres in pathological conditions [**9, 10**]. Tocopherol has antioxidative and anti-inflammatory functions which may provide decreasing the frequency and severity of pathological events in the skin [**11**]. Vitamin C regulates elastin and collagen biosynthesis in skin and vascular structure [**12**]. Depending on this knowledge, we decided to prescribe an antioxidant combination therapy with ascorbic acid and alpha tocopherol to our patient. We think that disease control was achieved during the therapy since no new lesions developed. The occurrence of new lesions after a systemic infection suggested an increase of oxidative stress which could not have been balanced with antioxidants. The prompt use of antioxidant therapy might have provided control of anetoderma. Further researches about oxidative stress and treatment with antioxidants in anetoderma are needed.

Conclusion

Primary anetoderma may present as widespread lesions with preceding inflammation in children. Since primary anetoderma is very rare in childhood, it should be in the differential diagnosis of skin disorders demonstrating loss of elastic tissue. Our case attracts attention to the possibility of oxidative stress accompanying systemic immunological responses that may cause anetoderma and to the efficacy of antioxidants in this disorder.

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