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

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Published: J Turk Acad Dermatol 2016; 10 (3) 16103c8
This article is available from: http://www.jtad.org/2016/3/jtad16103c8.pdf
Keywords: Antioxidant, elastic fibers, Jadassohn - Pellizzari type, primary anetoderma

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 pilomatrixoma, 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 Schwenninger-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.
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, anticytodioplin, 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 regress; 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-
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tered promptly and no additional lesions were ob-
served.

Discussion

Anetoderma is a rare skin disorder, mainly
defined in women aged 20-40 years. Aneto-
derma 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 pre-
ceding inflammation was clearly described be-
fore the anetoderma lesions developed,
clinopathological findings revealed this type
of anetoderma in our case. Although classifi-
cation of primary anetoderma depends on
presence of signs of inflammation, definition
of varying intensities of inflammatory infiltra-
tion in Schweninger-Buzzi type made some
authors consider about a modified classifica-
tion [3]. These authors subclassified primary
anetoderma as idiopathic and secondary
forms (associated with anti-phospholipid an-
tibodies), irrespective of the presence of in-
flammation. We also agree with this kind of
classification since inflammatory signs may
be so subtle that patient or clinicians can ea-
sily 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 aneto-
derma 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 altho-
ugh the patient denied any preceding infec-
tion in her first admission. This kind of
disease progression suggested us that im-
mune 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 reg-
ression of skin lesions. Topical and intralesio-
nal steroids, oral penicillin G, phenytoin,
dapsone, nicotinate were unsuccessful or pro-
vided 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, cea-
sing 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 mastocyto-
sis [4]. It is well-known that oxidative stress
involves in many systemic inflammatory con-
ditions including skin and was suggested to
contribute to the abnormal structure and
function of elastic fibres in pathological con-
ditions [9, 10]. Tocopherol has antioxidative
and anti-inflammatory functions which may
provide decreasing the frequency and severity
of pathological events in the skin [11]. Vit-
amin C regulates elastin and collagen biosynt-
thesis 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 le-
sions developed. The occurrence of new lesi-
ions 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 rese-
arches about oxidative stress and treatment
with antioxidants in anetoderma are needed.

Conclusion

Primary anetoderma may present as wides-
pread lesions with preceding inflammation in
children. Since primary anetoderma is very
rare in childhood, it should be in the differen-
tial diagnosis of skin disorders demonstrating
loss of elastic tissue. Our case attracts atten-
tion to the possibility of oxidative stress ac-
companying systemic immunological
responses that may cause anetoderma and to
the efficacy of antioxidants in this disorder.

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