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PUVA therapy in disabling pansclerotic morphea of children.

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Summary

We present a case of a 13-year-old girl with two years history of disease. The skin changes are initially characterized by unilateral brownish plaques developing gradually in well-demarcated white-colored sclerotic plaques and linear lesions involving the trunk and extremities. The pulmonary changes included fibrosis of the lung. Laboratory data are characterized by increased γ -globulin levels and anti-nuclear antibodies. The histopathological examination confirmed the diagnosis of morphea. We present her as a case of disabling pansclerotic morphea of children successfully treated with PUVA therapy.

Key words

Pansclerotic morphea, systemic sclerosis, PUVA therapy.

Disabling pansclerotic morphea of children is a rare variant of localised scleroderma. It is characterized by the appearance of polymorphous lesions with various levels of involvement of the skin and deep structures. The involvement of bones, tendons, fascia and muscles results in asymmetry, deformation of the limbs and severe disability (1, 3).

Case report

I.T.D., 13-year-old girl with 2-year history of the disease. The skin changes initially consisted of unilateral brownish plaques on the left side of the trunk and extremities. Later the lesions spread on the right part of the body and developed in deep solid masses involving the dermis and deepest tissues.

Examination of the skin revealed well-demarcated white-colored sclerotic plaques and linear lesions involving the trunk and extremities. The

left breast is deformed by a concentric sclerotic plaque (Fig. 1). In a period of two years the left extremities became thinner and the left leg gradually shortened of 3 cm.

Laboratory data were all within the normal range except for slight increase of erythrocyte sedimentation rate (20/58). Immunological tests: elevated γ -globulin levels: IgG 2002 mg% (n.v. 1160 \pm 305) and IgA 326 mg% (n.v. 200 \pm 60), positive α 1-antitripsin and rheumatoid factor, anti-nuclear antibodies 2.16 AI (n.v. <1) (ELISA, Alph Diagnostic, USA), anti-cardiolipid IgG antibodies: 3.35 AI (n.v. < 1) (ELISA, Inova Diagnostics, USA), positive ds-DNA, negative anti-Jo-1 (ELISA, Inova Diagnostics, USA) and negative anti-ss-B.

The capillaroscopy and the abdomen ultrasonography were normal.

Electromyography showed muscle changes localised in the sclerotic areas, as a result of the impairment of motor nerves by the sclerotic process.