III. PEDIATRICS

CONSIDERATIONS ON A CASE OF SYSTEMIC SCLERODERMA IN CHILD

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Abstract

The authors present a patient who was diagnosed with scleroderma at the age of 7 years, sustained by the typical aspect of the cutaneous modifications (hard edema at the level of the face, neck and limbs), difficulties in opening the mouth and histological modifications (the atrophy of the epidermis and the increase in thickness of the derm through the proliferation of the dermic conjunctive). In evolution, until the age of 16 years, the cutaneous lesions became general and complications appeared: oesophagitis of reflux, articular modifications, hypacusia, muscular tiredness and systolic breath in the mitral focus. The case was appointed in scleroderma, systemic clinical form. Comments were made on the differential diagnosis, on the treatment, the evolution and prediction. The presentation of the case is motivated by the rarity of scleroderma and especially of the systemic clinical form at child. The hypacusia was considered as a particularity of the case.

Key words: scleroderma, systemic clinical form, child.

Introduction

Systemic scleroderma (Ss) is a chronic, multi – systemic disease, characterized by the hardening, increasing in thickness and the rigidity of the teguments and by modifications of fibrotic type, inflammatory and vascular of certain organs. (1, 10) The etiology is unknown. It is however sustained the role of heredity and of certain environment factors. (2, 10) The pathogenesis of this disease is not clear. (1) From the pathogenic point of view it is characterized by anomalies of the metabolism of the collagen (excessive proliferation of mesenchymatous cells, fibroblasts, myocytes, endothelial cells, which, activated by unknown factors determine an increased synthesis of collagen I, III, X and other components of the conjunctive tissue with excessive deposit inside the skin, systems and organs), associated with vascular anomalies (hyperplasia of intimate, of small arteries and vasospasm). (2, 10) A vascular lesion, possibly caused by deposition of immune complexes or by release of citotoxic factors, seems to be at the origin of the disease. As a consequence, platelet adhesion and activasion might occur in sclerodermic patients. The observation that platelet might release, upon aggregation, a potent mitogenic factor, named Platelet Derived Growth Factor has focused interest on platelets as the potential mediators of the fibrotic process characteristic of SS. (1) The skin of patients with SD is characterised by an excess accumulation of collagen in the extracellular matrix of the fibrotic reticular derms. (4) Abnormalities in newly formed collagen structures as well as splitting of newly formed collagen fibrillae into microfibrillae were observed. (8) Elastic fibers are also disrupted in this disease, however, in contrast to collagen relatively few studies have provided information concerning the changes that occur to elastic fiber components in SD. (4, 9) Eosinophilia as a possible heart damaging factor in Ss in children. It was discovered that associated Ss and eosinophilia ran a course marked by more well – defined exudative reactions, with the heart being injured more frequently and gravely. A correlation was noted between the "sclerodermic" heart and the eosinophil count in the peripheral blood. (5) The disease is accompanied by immunologic cellular and humoral anomalies. (2, 10) As spreading, the SD is met all over the world, but the frequency of the disease is relatively small. (10) At the child the frequency is smaller than at the adult. (2, 3) At the girls the frequency is more than at the boys. (3)

Clinical observation

The patient M.C. is presented, who was hospitalized for the first time in January 1998, at the age of 7 years, for edemas and discrete myalgias. At the clinical examination was noticed a hard edema at the level of the face, neck and limbs and a slight difficulty in speaking. The diagnosis of SD was suspected which was confirmed by cutaneous biopsy (made at IOMC Bucharest) which showed: hyperkeratosis, formation of infundibulate corneous corks, the epidermis moderately atrophic, the derm increased in thickness with approximately 3 times the normal thickness, the dermic conjunctive increased in thickness disposed with horizontal stripes, fragmented, for the rest, the clinical examination on apparatuses and the paraclinical one did not show any pathologic modifications. For 7 years the child was lost from evidence. At the age of 14 years, in March 2005, she is hospitalized again for epigastric pains, dysphagia, pyrosis, hematemesis and melaena. The barium transit showed hypokinesia of the esophagus. (figure 1) For the rest, the digestive tube was normal.

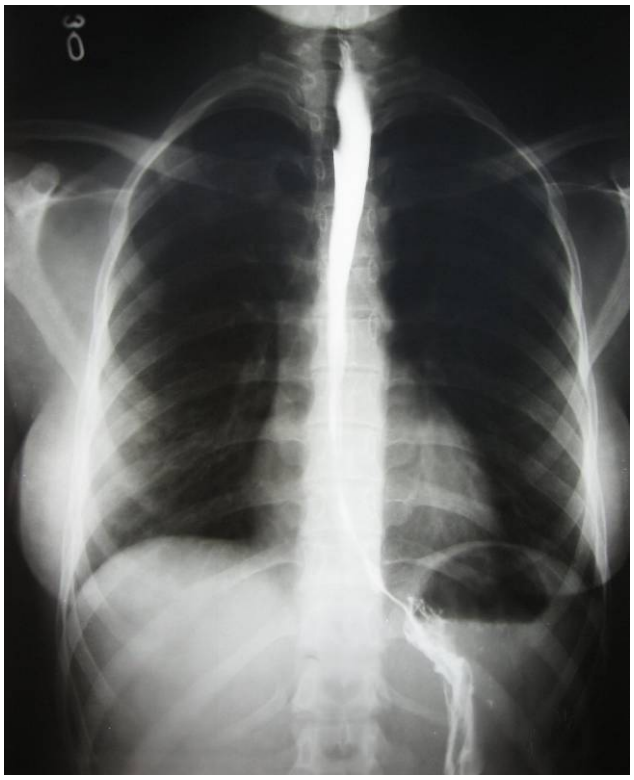

Fig. 1 SD. Barium radiography. Hypokinesia of the esophagus.

The esophagoscopy distinguished oesophagitis of reflux and esophageal cankers in the inferior third. After a year, in January 2006, she returns for: vertigo, headache, hypacusia. In January 2007 she is hospitalized again (F.O. 2452 / 15.01.2007) for the clinical and paraclinical revaluation accusing headache and tiredness to effort. The clinical examination finds a 16 year old teenager, with a weight of 41 Kg, waist 150 cm, with hardened, rigid, infiltrated teguments. At the face level the teguments were glossy, spread, with the cutaneous pleats erased and lack of expression of the mimics. (figure 2) She has difficulties in moving her mandible and opening the mouth.

At the level of the hands, a slight fixing in semiflexion of the fingers was ascertained. (figure 3).

Fig. 2 Ss. Edemas and the rigidity of the teguments of the face.

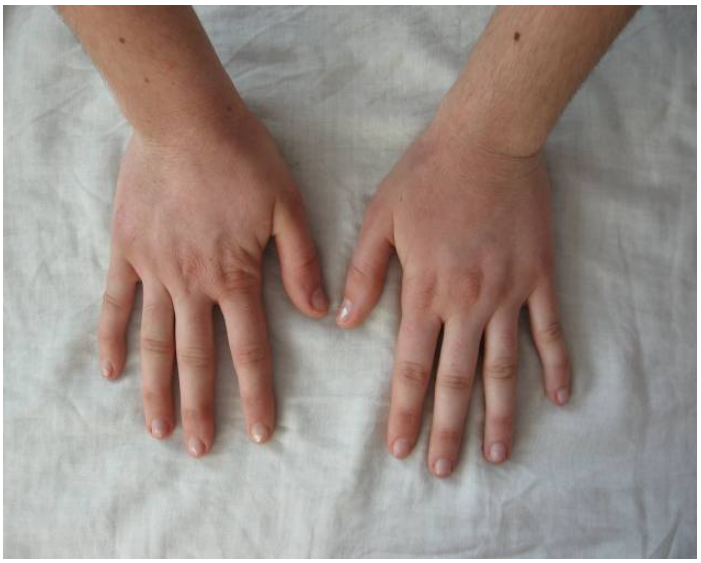

Fig. 3 Ss. Fixation in semiflexion of the fingers of the hand by increasing the thickness and by sclerosis of the teguments.

More than the previous hospitalizations a systolic breath is noticed, degree II at the top of the heart.

Paraclinical explorations: HLG, serum iron, the inflammatory tests (VSH, fibrinogen, CRP), summary urine exam, urea, creatinine, uric acid, glycaemia, calcemia, blood ionogram, electrophoresis of proteins, transaminase, CP-K, aldolase were normal. FR and ANA absent. Immunelectrophoresis showed a raise of IgM (300ui/ml, N=100-200 ui/ml), FO normal, IDR 2u PPD = 0 mm.

Functional explorations : EEG, EKG, EMG – normal range; Normal breathing functional tests .

Imagistic explorations: abdominal echography – normal, cardiac lung radiography without pathologic modifications, hands radiography – normal, sinuses radiography – bi-jaw sinusitis.

Corroborating the case history data with the actual clinical and paraclinical exam the following diagnosis was reached: Systemic scleroderma (diffuse cutaneous form with late visceral affection). Oesophagitis. Hypacusia. Jaw sinusitis. Cardiopathy?

Discussions

The positive diagnosis was sustained by the typical aspect for SD of the cutaneous modifications (confirmed by histological examination), to which the visceral affection was added in time (oesophagitis, muscular affection expressed by tiredness to effort and possibly heart affection). Lung and renal sufferance was not distinguished, Raynaud syndrome (met at 95% of the ill) (10) and neither visceral touches more rare (Sicca syndrome). From the point of view of the classification, the case was framed in Ss with diffuse cutaneous fibrosis and particularly with late visceral affection (non characteristic Ss diffuse which presents precocious visceral affection). (10) The differential diagnosis was made with:

a) The localized sclerodermias (in plates and in band) which are also frequent at the child and present cutaneous lesions limited to the skin and subcutaneous tissue. (2, 3, 10)
b) Eosinophilic fasciitis (Schulmann syndrom) similar SD from which differs through characteristic eosinophilia in the blood and in the inflamed areas (at the bioptic) and good response to cortisone.
c) Mixed disease of the conjunctive tissue (Scharp syndrome), characterized by common elements LES, DM and SD and good response to cortisone
d) Secondary forms of SD (after medications, chemical substances, medulla transplant).
e) Pseudosclerodermias (sclerema, scleromixedem, cutaneous amyloidosis).

The treatment is deceptive (7) and it has not been definitively standardized. (6, 11) The administration of immunodepressives, agents that diminish the collagen production or factor XIII do not have noticeable therapeutic results. (2, 3, 7) The presented case was initially treated for the cutaneous affection in the stage of edema with prednisone having an unsatisfying effect. Subsequently, the treatment of oesophagitis was made with rest with the bed raised at the head, diet regime, Ranitidine, Metoclopramid and Dicarbocalm, with a very good evolution. A recovery program was applied for the increase of the elasticity of the tegument and articular mobilities, by hydrothermal treatment and kinetotherapy. At the same time, we tried to diminish the hardening of the teguments by administration of vitamin E and application of ointments for the prevention of skin dryness and for the anti-inflammatory effect (ointment with hydrocortisone and emollient substances) in the areas more affected with modest benefits.

SD has a chronic evolution with progression more or less rapid. The systemic forms have a severe evolution, potentially fatal by renal, heart and lung lesions and not validating by cutaneous lesions. (6) The duration of survival after the age of 5 years is met in 50-70% of cases and of 10 years in 40-60% of cases. (10) The case presented, after the

last revaluation was framed in the forms with a less severe evolution (not characteristic for Ss), motivated by the fact that 9 years after the diagnosis was put, it surely presents only esophagial and articular affection and possibly heart and muscular affection, but the evolution is not validating by the generalized cutaneous lesions and hypacusia.

Conclusions

The case was presented due to the very small frequency of SD at child (20 cases at 1000000 general population of which less than 8-10% at child) (2) and also because of the fact that the clinical form of Ss described in that particular case is not characteristic to the child.

Taking into consideration the continuous evolution of the disease, it is imposed that the ill person to be periodically revaluated and the medicine and recovery treatment should be adapted to the functional modifications found.

Particularity of the case

Is constituted by the signalling of the hypacusia as a complication not registered in the specialty literature studied.

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