SCLERODERMA IN PEDIATRIC AGE GROUP: REPORT OF 25 CASES

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Abstract - Scleroderma is a multisystem autoimmune connective tissue disease, characterized by hardening of the skin. We had 25 patients in the Iranian Pediatric Unit of Rheumatology over 10 years. There was female predominance (F.M = 4/1, 80% female, 20% male). The age of onest was from 6 months to 16 years, the mean age being 10 years. The symptoms and signs were: nonpitting edema in 16%, hard skin in 80%, sclerosis of distal limbs in 72%, facial in 20%, atrophy, and hypopigmentation or hyperpigmentation in 40%, telangiectasis (15%), subcutaneous calcification (10%), Raynaud's phenomenon in 60%, digital ulceration (16%), muscle pain and weakness, arthralgia (30%). Rarer manifestantion included, dysphagia (16%), dyspnea (8%), cardiovascular disease, pericarditis, cardiomegaly, CHF (16%). Laboratory abnormalities included: anemia (80%), positive FANA. > 40 (88%) ESR > 60 (40%) CRP +++ (40%). Skin biopsy documented increase in the collagen content of dermis in 75%.

Acta Medica Iranica 37(2): 106 - 109; 1999.

Key Words: *(Scleroderma), FANA (Fluorescent Antinuclear Antibodies), ESR (Erythrocyte sedimentation rate), CRP (C-reactive protein)*

INTRODUCTION

Scleroderma is a chronic multisystem connective tissue disease characterized by symmetric fibrous thickening and hardening (sclerosis) of skin (hard-skin), and degenerative changes in synovium and widespread abnormalities of the viscera, such as esophagus, intestinal tract, heart, lungs, and kidneys (1, 2, 4). Scleroderma has been described worldwide and in all races. Childhood onset is very uncommon. The incidence is 4.5 to 12 cases per one million. High incidence is reported in the 30-50 years age group. There are several small published reports of pediatric patients with scleroderma. This study is important because we have 25 cases diagosed as scleroderma in Pediatric Unit of Rheumatology in Rheumatology Research Center in Tehran University of Medical Sciences.

PATIENTS AND METHODS

Between 1987 to 1997 we have observed 25 children with symptoms of scleroderma in Pediatric Rheumatology Unit in Rheumatology Research Center. All patients were examined in the outpatient clinic of Shariati Hospital and Children Hospital Medical center by different groups of pediatric rheumatologists, pediatric cardiologists and pediatric gastroenterologists.

Data was collected on a standardized protocol comprising 20 items. All patients were examined once a month for one year, then every two months for about ten years. Diagnosis was based on clinical manifestations of the disease.

RESULTS

We had 25 cases of scleroderma. The age at onset was from 6 months to 16 years. Female to male ratio was 4:1 (80% female, 20% female) (Table 1).

The clinical manifestations of scleroderma in the pediatric age group consists of two different groups:

1- Constitutional sings comprised of: fever (70%), malaise (50%), poor appetite (32%), weight loss (25%) and general pain (40%) (Table 2.).

Table 1. Age & Sex distribution

Age(y)	Total	%	Male	%	Female	%
0-2	2	8	--	--	2	8
2-4	1	4	--	--	1	4
4-6	2	8	--	--	2	8
6-8	3	12	--	--	3	12
8-10	5	20	2	8	3	12
10-12	6	24	1	4	5	20
12-14	4	16	2	8	2	8
14-16	2	8	--	--	2	8

2- Specific organ involvements are summarized in Table 3. Skin manifestations included non-pitting edema in 4 cases (16%), hard skin (80%), sclerosis in distal limb in 18 cases (72%), facial sclerosis in 5 cases (20%), atrophy of the skin in 10 cases (40%), digital pitting and ulceration in 4 cases (16%), telangiectasis in 3 cases (12%) and subcutaneous calcinosis in 2 cases (80%). Gastrointestinal involvement: dysphagia, and esophageal dysfunction, on esophagoscopy were seen in (16%), esophagitis in 10%. Cardiomegaly without CHF was seen in three cases (12%). Abnormal ECG, and CHF were reported in one patient. Raynaud's phenomenon was seen in 15 cases (60%). Pulmonary manifestations: such as dyspnea, dry cough, and decreased vital capacity on spirometry were seen in 2 cases (8%). Musculoskeletal manifestations included: morning stiffness, arthralgia, muscle pain, myositis, and joint contracture seen in 10 patients (40%). Central nervous system abnormality was seen in one case. Renal disease was not seen in this study.

DISCUSSION

Scleroderma is characterized by progressive sclerosis of skin, Raynaud's phenomenon, arthritis and arthralgia, muscle weakness and pain, dysphagia, dyspnea, and also visceral involvement. The onset of the disease is usually insidious with low grade fever, malaise and profound fatigue. The course of disease is progressive. Weight loss is common even in the absence of gastroinestinal involvement. The skin initially appears shiny and taut and may be erythematous at early stages. The face and neck creases are obscured and hair growth is usually involved next. In later stages, atrophy occurs. Digital skin is reduced. Internal organ involvement occurs in the systemic progressive form of disease.

Table 2. Clinical manifestations of scleroderma

Constitutional sign:	Total	%	Female	%	Male	%
Fever	18	70%	16	64%	2	8%
Malaise	12	50%	12	50%	0	0%
Fatigue	12	50%	8	30%	4	20%
Poor appetite	8	32%	6	24%	2	8%
Weight loss	6	25%	5	20%	1	4%

Table 3. Presenting signs and symptoms in children with scleroderma

Major manifestations (organ involvelment)	Number	%
Non-pitting edema	4	16%
Hard-skin (Tightening)	20	80%
Sclerosis of distal limb (dorsum of the digits)	18	72%
Sclerosis of facial skin	5	20%
Skin atrophy	10	40%
Raynaud's phenomenon	15	86%
Digital pitting ulceration	4	16%
Telangiectasis	3	12%
Subcutaneous calcinosis	2	8%
Arthralgia & arthritis	10	40%
Musculoskeletal disease myositis	8	32%
Gastrointestinal involvement	4	16%
Dysphagia	4	16%
Esophagitis	2	8%
Cardiac manifestation	3	12%
Cardiomegaly without CHF	3	12%
Abnormal ECG with CHF	1	4%
Pulmonary disease	2	8%
Central nervous system	1	4%
Renal disease	0	0%

Table 4. Laboratory finding

Lab exam	age	number	%
Hb	<12	20	80%
ESR	>40	22	88%
ESR	>60	10	40%
CRP	+++	10	40%
FANA	+	5	20%

We studied 25 patients over a period of 10 years. The demographic profile of these patient was similar to those in western reports. The constitutional signs (fever 70%, malaise 50%, fatigue 50% poor appetite 32% weight loss 25%) were less frequent than those in western reports. The percentage of patients having Raynauds'phenomenon (86%) is similar to those in western reports (90%). Gastrointestinal involvement including dysphagia and regurgitation was seen in (16%) compared to (35%) of western patients. Cardiac and pulmonary involvement, a primary cause of morbidity among children with SSc, was seen in 12% similar to western reports. Musculoskeletal symptoms are relatively common in SSc: arthritis, morning stiffness and joint pain were seen in 10 cases (40%). In conclusion, the present study shows that SSc is not common in children. Comparison between western and Iranian patients demonstrates some differences: higher incidence of constitutional signs, skin manifestations, gastrointestinal manifestations cardiac and pulmonary manifestations, musculoskeletal manifestations, arthralgia, neurological manifestations, and Raynaud's phenomenon.

Table 5. SSc in children compared with western reports

Author	N	Male	Female	Age at onset(Y)
Cassidy et al	15	0	15	3-15
Komreich et al	13	4	9	3-12
Szymanski-Jagiello et al	12	3	10	6-14
Schlesinger and Schaller	11	2	8	6-10
Moradi-Nejad et al	25	5	20	1-16

Table 6. Clinical manifestations compared with western reports. 10-14

Symptoms & sign	Cassidy et al (n=15)	Moradi-nejad et al (n=25)
Skin tightening (hand & face)	15	20
Raynaud's phenomenon	11	15
Arthralgia & arthritis	9	10
Subcutaneous calcification	3	2
Telangiectasis	4	3
Dysphagia	3	4
Dyspnea	3	2
Cardac involvement	3	3

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