Case Report

Scleroderma: A Case Report

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ABSTRACT

Scleroderma is a connective tissue disorder, mainly involving skin, blood vessel, and visceral organs. The term "sclera" meaning hard "derma" meaning skin in Greek. The manifestations are due to diffuse deposition of collagen in the skin and internal organs along with vascular injury and immunologic abnormalities. The skin becomes shiny and taut giving a mask like appearance of the face and claw like appearance of fingers. Orofacial changes are quite characteristic apart from mask like appearance of face there will be decrease in facial profile and rigidity of tongue. In this article, we report a case of scleroderma with brief review of literature emphasis on orofacial manifestation.

Keywords: Fibrosis of the skin, Mask like face, Oral manifestation, Reynaud's phenomenon, Scleroderma

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INTRODUCTION

Scleroderma is a disorder of the connective tissue characterized by fibrosis of the skin, blood vessels, and visceral organs.[1] Scleroderma occurs in two forms, localized and systemic forms. In the localized form (morphea), there will be involvement of the skin and subcutaneous tissue occasionally deeper tissues. In the systemic form, diffuse fibrosis of the skin and internal organs, primarily involving the blood vessels, gastrointestinal tract, lungs, heart, and kidneys.

The chief manifestations include thickening and induration of the skin, digital pitting, sclerodactyly, pigmentations, telangiectasia, and Raynaud's phenomenon. Raynaud's phenomenon is defined as a paroxysmal vasospasm that is induced by exposure to cold or emotional stress.[2,3] It may begin in children and young adults with females being twice as frequently affected as males.

The oral manifestations of scleroderma include fibrosis and rigidity of facial skin (mask-like appearance face), tongue, soft palate, larynx, salivary glands, and buccal mucous membrane leading to microstomia, dysarthria, dysphagia, and xerostomia. Periodontal manifestations such as loss of attached gingiva and multiple foci of recession have been reported.[4]

Radiographically, there will be widening of the periodontal ligament (PDL) space without mobility of teeth and resorption of the coronoid process, condyle, and posterior aspect of the ascending ramus of the mandible. These changes in the mandible and temporomandibular joint cause impaired growth and development as well as functional problems such as subluxation and abnormal motion.[4,5]

The mechanism of the fibrotic changes is unknown, but hyperplasic changes of collagen have been documented and also inflammatory changes and globulin deposits were found in blood vessel walls, which apparently explain the basis for altered collagen. The pathological findings indicate that fibroblasts are activated to produce excessive amounts of collagen and other components of the cellular matrix.[6]

Diagnosis of scleroderma is clinical and is made by the presence of Raynaud's phenomenon, typical skin thickening and visceral involvement. Laboratory investigations are supportive. For classifying the subtypes of disease and excluding other scleroderma mimicking condition serology test for antibody profile is helpful. To determine the extent and the stage of visceral involvement due to the disease process organ specific investigation are helpful.[7]

Treatment of scleroderma is a challenging because, disease manifestations are varied and are a cumulative effect of progressive fibrosis, obliterative vascular changes and immune system activation and autoimmunity.

The goal of therapy is to improve quality of life by minimizing specific organ involvement and subsequent life-threatening disease. Hence, multiple drug therapy targeting the different pathogenetic mechanisms required.

CASE REPORT

A 14-year-old female patient reported to the department, with a complaint of small mouth since birth. Patient also gives the history of inability to close mouth but no difficulty in tongue movement, speech, and swallowing. No history of joint pain. Past medical, dental, and family history was non-contributory. On general examination patient was conscious, co-operative moderately built with signs of pallor.

Extra oral examination revealed stiffening (fibrosis) of facial skin with smooth, taut, and mask like appearance of face. Lips were rigid, incompetent with loss of vermillion border of upper lip, producing microstomia with fish beak appearance. Pinched appearance of nose resembling mouse. Blanching of hand and hyper pigmented areas on face and feet were noticed. Surface of the skin was smooth and shiny (Figure 1).

On intra oral examination, full complement of teeth with macrodontia and angles Class I molar relation with crowding was noted. Maxillary palatal arch was narrow due to microstomia. Buccal mucosa was thick with stiffness of the mucosa. Yellowish stain was present on all teeth surface with frosted appearance (Figure 2).

Hence, considering the history and clinical examination provisional diagnosis was given as scleroderma.

Further investigations were carried out, intraoral periapical radiograph and orthopantomogram revealed there was widening of PDL space in relation to 34 and thickening of PDL. There was no resorption bone (Figure 3).

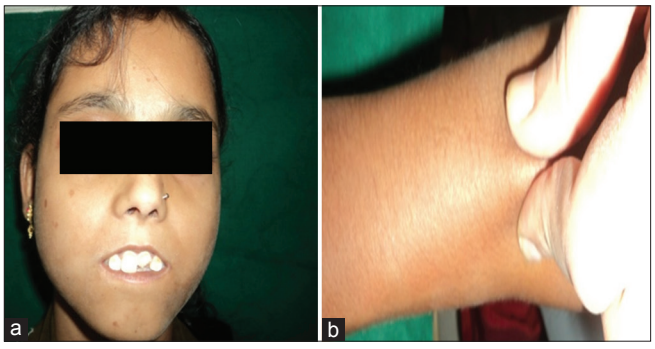

Figure 1: (a) Extra oral view of patient, (b) stiffening of the skin showing difficulty in pinching

Reynaud's phenomenon was checked, there was discoloration of extremities after exposure to cold (Figure 4). On hematologic investigations neutrophils was high, hemoglobin was 9.6 g%. Chest X-ray and ultrasound of pelvis and abdomen were done which was normal. By taking consent from patient punch biopsy was done on the shin area (leg). Histopathology report showed atrophic epidermis and flattened ridges. Papillary dermis showed thickened collagen fibers, mild perivascular lymphocytic infiltrate. Reticular dermis showed focally homogenized collagenized fibers, thickened wall of small blood vessels with narrowed lumen. Considering all the above features we came to final diagnosis of scleroderma. Patient was prescribed

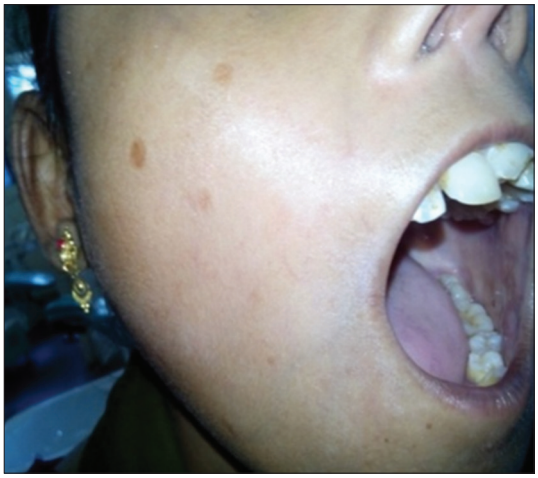

Figure 2: Intra oral view

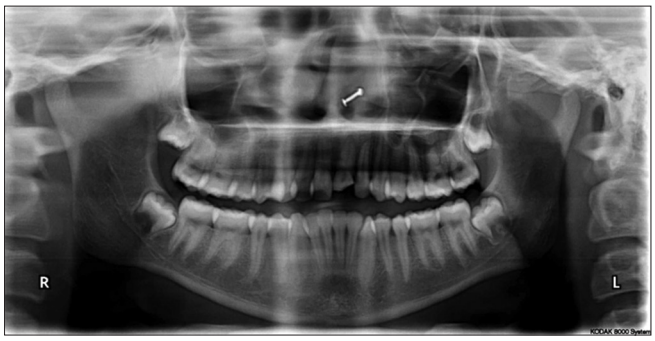

Figure 3: Orthopantomogram showing widening of periodontal ligament space

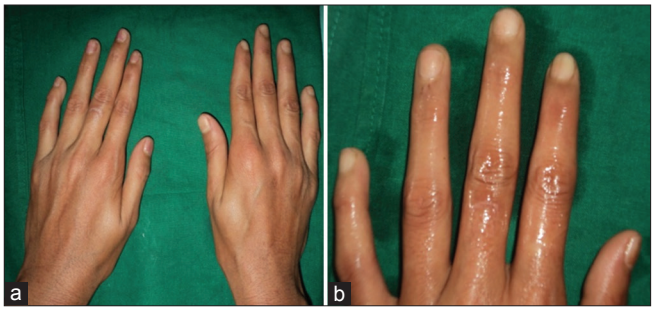

Figure 4: Reynaud's phenomenon showing discolored finger nail

with tablet nicordia once daily for 2 weeks. Patient was referred to Department of Dermatology and Pediatrics, rheumatology for further management.

DISCUSSION

Scleroderma is an autoimmune disorder involving multiple systems thus making the course of this disease unpredictable. Though the correct nature of the disease is not known, high prevalence of circulating auto antibodies in serological investigations points it toward autoimmune mechanism.[1]

The pathogenesis of scleroderma remains unclear but it is characterized by endothelial activation, immune system dysfunction and enhanced fibroblast activity. The endothelium controls the contraction and relaxation of vascular smooth muscle cells, leading to vasospasm and smooth muscle hypertrophy. Eventually it leads to obliteration of the lumen of small arteries and capillaries which leads to ischemia. There is extravasation of inflammatory cells initially predominated by monocytic lineage and later by lymphocytes.[8,9] There is enhanced fibroblast activity which stimulates fibroblast to produce excessive extracellular matrix, the main hallmark in scleroderma.

Scleroderma clinically presents as two major forms as localized form and systemic form. Localized form affects the skin, without involving the internal organs.[10]

Tuffanelli and Winkelmann classified localized scleroderma (LSc) into three types:

- Circumscribed sclerotic plaques (morphia)
- Streak on the skin (linear Sc)
- Generalize morphia widespread skin involvement with multiple plaque and frequent muscle atrophy.

Systemic scleroderma (SS) was classified by Lepoy *et al*. as,

- Limited cutaneous SS formally called calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia syndrome
- Diffused cutaneous SS: Sclerosis of face, trunk, proximal extremities
- SS sine scleroderma: No skin thickening only organ fibrosis.

Reynaud's phenomenon is one of the earliest manifestations of the disease though it may not show the typical triphatic color changes in affected individual when exposed to cold or stress but does present as pallor of the digits.

Differential Diagnosis

Early stages of disease often share clinical and laboratory features of other connective tissue diseases such as systemic lupus erythematosus and rheumatoid arthritis. Once skin thickening has developed, the differential diagnosis includes a wide array of other disorders in which tightening and thickening of the skin prominent features. In diabetic digital sclerosis, vinyl chloride disease, vibration syndrome, bleomycin induced scleroderma, chronic reflex sympathetic dystrophy, amyloidosis, and acrodermatitis are characterized mainly by skin thickening and involvement of digits. Disorders characterized by skin thickening and sparing the fingers include eosinophilic fasciitis, eosinophilic-myalgia syndrome, generalized subcutaneous morphea, amyloid, carcinoid syndrome.[7,11,12]

Management

In general, disease heterogeneity (stage, severity, and pace of pattern) is an important factor in the therapeutic plan. With the manifestation of Reynaud's phenomenon calcium channel blockers, angiotensin Type II receptor blocker, surgical sympathectomy will be the choice of treatment.[13] Cases with digital ulcer, skin fibrosis, arthritis, myositis, drugs like immunosuppressive, non-steroidal anti-inflammatory drugs, low dose of steroids given. In severe cases were their is renal crisis and pulmonary hypertension angiotensin-converting enzyme inhibitors given as a treatment modalities.[14]

CONCLUSION

Scleroderma is a multisystem organ involvement disorder with oral and cutaneous manifestation. Autoantibody profile will help in identifying disease severity and organ involvement, differentiating scleroderma from other disease. Physicians and patients should be more attentive to the potential risk factors for organ damage, particularly very early in the disease, even when the patients may not be symptomatic. Treatment should be initiated as soon as problems are identified.

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How to cite this article: Kotian R, Shruthi M, Kini R, Dsouza LC. Scleroderma: A Case Report. Int J Adv Health Sci 2015;1(12):6-9.

Source of Support: Nil, **Conflict of Interest:** None declared.