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Case Report

A Case Report on Systemic Sclerosis

P Vyshnavi*, K Geethanjali, M Prasanth, NS Kavya and S Swetha

Pharm D Intern, Department of Pharmacy Practice, Sri Padmavathi School of Pharmacy, Tiruchanoor, Tirupati, Andhra Pradesh, India

***Corresponding Author:** P Vyshnavi, Pharm D Intern, Department of Pharmacy Practice, Sri Padmavathi School of Pharmacy, Tiruchanoor, Tirupati, Andhra Pradesh, India.

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Abstract

Scleroderma, or systemic sclerosis (SSc), is a chronic multisystem autoimmune disease characterized by a vasculopathy, diffuse fibrosis of skin and various internal organs, and immune abnormalities. Scleroderma occurs in two forms, localized and systemic forms. Major pathogenic mechanisms involved in scleroderma were immune system activation, enhanced fibroblast activity and endothelial dysfunction. We report a case of systemic sclerosis with cutaneous and musculo skeletal manifestations. There is no complete cure for systemic sclerosis, treatment was given only for symptomatic relief and to manage complications. Early diagnosis and treatment plays an important role to increase the life expectancy of the patient.

Keywords: *Scleroderma; Fibrosis; Autoimmunity; Antinuclear Antibodies; Corticosteroids*

Introduction

Scleroderma, or systemic sclerosis (SSc), is a chronic multisystem autoimmune disease characterized by a vasculopathy, diffuse fibrosis of skin and various internal organs, and immune abnormalities. The clinical manifestations of this disease are extremely heterogeneous and depend on the presence and degree of various internal organ involvement. Scleroderma occurs in two forms, localized and systemic forms [1].

Localised scleroderma: Involvement of the skin and subcutaneous tissue.

Systemic sclerosis: Diffuse fibrosis of the skin and internal organs, primarily involving the blood vessels, gastrointestinal tract, lungs, heart, and kidneys. It was further classified as

- **Limited cutaneous systemic sclerosis (lcSSc):** CREST syndrome refers to Calcinosis, Raynauds phenomenon, Oesophageal dysmotility, Sclerodactyly, Telangiectasia.
- **Diffuse systemic sclerosis (dcSSc):** Sclerosis of proximal extremities, trunk, and face
- **Systemic sclerosis sine scleroderma (ssSSc):** Organ fibrosis only; no skin thickening.

It was mostly observed in children and young adults. Females were diagnosed twice as frequently as males. Major pathogenic mechanisms involved in scleroderma were immune system activation, enhanced fibroblast activity and endothelial dysfunction [2]. Diagnosis of systemic sclerosis was made based on Physical examination i.e. cutaneous and systemic signs and symptoms or American college of rheumatology diagnostic criteria for systemic sclerosis, Complete blood count, Urinalysis, Complete metabolic panel, ANA, ACA, Antibodies for topoisomerase [3]. Current treatment of systemic sclerosis is directed toward managing complications and providing symptomatic relief.

Case Report

A 31 year old female patient was admitted in the department of dermatology in a tertiary care teaching hospital with chief complaints of tightness of skin since 1 year, joint pains gradual in onset, progressive in nature associated with joint movement restriction, ulcer over both feet, swelling of both hands, feet, blackish discolouration of skin all over the body, involuntary passage of urine and stools since 1 month, difficulty in breathing on exertion, difficulty in mouth opening, head ache, visual disturbances since 3 months. She was diagnosed with Systemic sclerosis five years back and on irregular treatment with prednisolone, chloroquine. Her menstrual history reveals she attained menarche at 12 years and menopause 1 year back. personal history discloses that her father was also died with similar complaints 10 years back.

Cutaneous examination shows tight, taut, shiny skin on face, Microstomia, Tautness of skin on hands, legs, non-pitting edema of fingers, Flexion deformities of hands with hyperpigmentation of skin over hands, Non-healing ulcers of varying sizes over both feet with pale granulation tissue. Autoamputation of right little toe was also seen. Her hair and mucosa was normal. Nails show Onycholysis.

Her HRCT Thorax reveals Interstitial lung disease. Her blood profile indicates anti-nuclear antibodies (ANA) positive, C – reactive protein (CRP) 12.8 mg/l and RA factor 128. Ultrasonography of abdomen and liver function tests doesn't show significant abnormality.

Treatment given includes Prednisolone 20 mg, Nifedipine 10 mg, Amoxiclav 625 mg, topical emollients and vitamin supplements. Patient had previously used chloroquine but now it was stopped as she develops visual disturbance due to chloroquine.

Discussion

Systemic sclerosis is a chronic disease of unknown etiology and involves multiple organ systems. Though the exact nature of the disease was not known, presence of antibodies makes this disease to categorise under auto immune disorders. Many studies reveals that the alteration of fibroblast function which leads to excessive fibrosis of several organs play a key role in the progression of systemic sclerosis. The pathological alterations in systemic sclerosis started with endothelial cell activation and vascular damage of unknown etiology leads to Lymphocyte and monocyte infiltration. Extravasation of these inflammatory cells activates fibroblasts the key for excessive fibrotic pathology of scleroderma. This may also causes imbalance between endothelium derived vasoactive substances i.e. increased endothelin and decreased nitric oxide, prostacyclin leads to vasospasm, muscular hypertrophy and ischemia. Other key growth factors that have also been implicated in the pathogenesis are connective tissue growth factor (CTGF), platelet-derived growth factor (PDGF) and the beta chemokines monocyte chemoattractant protein (MCP)-1 and MCP-3, TNF alpha.

The pathogenesis was summarised as Increased permeability, enhanced vasoreactivity, enhanced expression of adhesion molecules, imbalance between hemostatic and fibrinolytic factors, platelet activation, altered vascular wall growth bring about altered functional state of the endothelium and fibroproliferative changes in the vasculature [3,4].

Typical signs and symptoms of systemic sclerosis includes Cutaneous: Diffuse edema of hands and feet (early stages) Progressive skin tightening, Sclerodactyly, Digital ulcers and pits, Contractures, Hyperpigmentation, hypopigmentation, salt and pepper skin, Calcinosis, Telangiectasias, microstomia. Vascular: Raynaud's phenomenon, onycholysis, Digital ischemia and ulcers. Pulmonary: Interstitial lung disease, including alveolitis and interstitial fibrosis, Pulmonary hypertension, Recurrent aspiration pneumonitis caused by esophageal reflux and dysmotility, Respiratory muscle weakness. Cardiac: Cardiomyopathy, Congestive heart failure, Heart blocks, Pericarditis. Gastrointestinal: Gastroesophageal reflux, Esophageal dysmotility, aperistaltic esophagus, Esophageal stricture, Barrett's esophagus, Colonic wide-mouth diverticuli, Primary biliary cirrhosis, Anal incontinence, Orodental manifestations [2,3,5].

Early stages of disease often mimic other autoimmune disorders like systemic lupus erythematosus but later differential diagnosis was done based on symptoms like cutaneous manifestations, organ involvement etc. Blood profile was done to known the presence of antibodies and other organs involved. Once the diagnosis was confirmed the patient need to be evaluated that to which type of scleroderma he/she belongs to. Then, the patient was treated based on the symptoms like CCBs, Alpha blockers for Raynauds phenomenon, endothelial antagonists – bosantan, Prostanoids analogues–alprostadil, Corticosteroids, Immunosuppressants methotrexate, rituximab etc. Proton pump inhibitors, antimotility agents, for GI symptoms. Antihistamines, emollients, antibiotics for cutaneous features. NSAIDs, for muscular abnormalities [6,7]. The patient must be treated based on severity, drug tolerance, possibility of adverse drug reactions etc. There is no complete cure for systemic sclerosis, treatment was given only for symptomatic relief and to manage complications.

Conclusion

Scleroderma presents great challenges to various medical professionals as it involves several organ systems. Physicians and patients should be more attentive to the potential risk factors for organ damage, in the early stages of disease. Early diagnosis and treatment plays an important role in reducing complications and to increase the life expectancy of the patient.

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