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DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS: A CASE REPORT

***Safura Sultana, Dr. Shaheda Siddiqui, Uzma Parveen, Salva Fatima Heba, Sara Sana Ayesha Khanum and Rehab Rafi**

Department of Pharmacy Practice, Deccan School of Pharmacy (DSOP), OU, Hyderabad-500024, Telangana, India

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ABSTRACT

Systemic sclerosis (SSc, Scleroderma) is a rare connective tissue disorder that is highly diverse in its multisystem clinical presentation and follows a variable course. Systemic sclerosis is a disease that will lead to collagen diposition and fibrosis of skin and various internal organs like lungs, kidneys, blood vessels, and gastrointestinal tract. Systemic sclerosis can be clinically discerned into two subsets based on skin findings and serological data they are limited cutaneous systemic sclerosis (lSSc) and diffuse cutaneous systemic sclerosis (DcSSc). Diffuse cutaneous systemic sclerosis is governed by rapid progressive fibrosis of skin and other internal organs. Diffuse cutaneous systemic sclerosis is characterised by proximal skin sclerosis, Raynauds phenomenon, sclerodactyly, degenarative changes and vascular abnormalities. This case reports presents a case of diffuse cutaneous systemic sclerosis in 45yr old female patient with characteristic systemic and intraoral manifestations.

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INTRODUCTION

The word scleroderma is derived from two greek words "sclero" means hard and "derma" means skin owing to its characteristic hard skin (Rahul Srivastava *et al*., 2016). Systemic sclerosis is a rare, complex, systemic disease that will cause the fibroblast activation and increased collagen production which leads to desseminate vascular injury and sclerosis accompanying skin and various internal organs (Verrecchia *et al*., 2007). Sytemic sclerosis also affects facial and oral structures (Asokan *et al*., 2013). Systemic sclerosis is a heterogenous connective tissue disease in addition occurs as a result of characteristic auto immunity involvement demostrating disease specific auto antibodies and signs of vascular damage such as Raynauds phenomenon (Pia Moinzadeh *et al*., 2014). Raynauds phenomenon is an early indicator of systemic sclerosis, it is a medical condition occurs in response to cold exposure and exhibits vasospasm of fingers (Rahul Srivastava *et al*., 2016).

Corresponding author: **Safura Sultana,**
Department of pharmacy practice, Deccan school of pharmacy (DSOP), OU, India, Hyderabad-500024, Telangana, India.

Scleroderma (as a single condition) was first described by curzio naples in 1753. Following 100 years after this Gintrac gave the term scleroderma, as the skin is main organ involved. Systemic sclerosis patients has 3-5 years of survival rate following the onset of disease when associated with early pulmonary, cardiac or renal involvement (Verrecchia *et al*., 2007). Systemic sclerosis is present throughout the world and is represented in all ethnic groups. The peak age of onset is in between 30-50 years in females (Rahul Srivastava *et al*., 2016). The standard classification criteria for SSc are the 1980 preliminary criteria, developed by American college of Rheumatology (ACR) according to it (Clodoveo ferri *et al*., 2002).

Major criteria

- Proximal scleroderma

Minor criteria

- Sclerodactyly
- Digital pitting scars
- Bibasilar pulmonary fibrosis.

The patient should fulfill the major criterion or two of the three minor criteria. This classification is 98% specific and 97% sensitive for the diagnosis of systemic sclerosis (Clodoveo ferri *et al*., 2002). This disease is diagnosed based on clinical features and ANA profile, since this methods are not useful for early detection newer diagnostic techniques are being developed like in-vivo confocal laser scanning microscopy (Kirsten Sauermann *et al*., 2002). The following case report illustrates the clinical manifestations and treatment undertaken by heathcare professionals.

Case report

A 45yearold female patient was admitted into the general medicine department with complaints of continous of loss of weight approximately she lost around 20kg in 2 months, pain in lower limbs since three days and decrease in mouth opening. On Physical examination, the patient was conscious coherent and presented positive for muscle wasting of both upper and lower limbs and contractures of upper limb. Systemic examination (vitals) was normal.

History

On enquiring the patient had no history of Hypertension, diabetes or thyroid disorders. The family history of patient was also not significant and there was no past history of any surgeries, contact with any TB patient and Blood transfusion. The personal history of patient includes she is bed ridden since 2 months unable to perform daily life activities, sleep disturbed, unable to open mouth completely, tightness of face skin, clawing of fingers of hands. The general physician advised for dermatological opinion, The dermatological review disclosed the patient with following clinical manifestations sclerodactyly (Figure-1), microstomia, Raynauds phenomenon, clawing of fingers and few palmar pits.

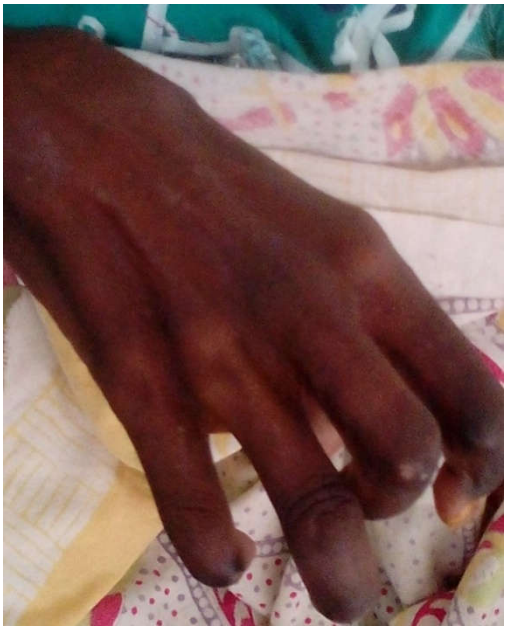

Figure 1. Sclerodactyly of fingers

The various lab investigations performed and treatment provided are listed below in – Table 1,2,3,4,5. The pathological impression of complete blood picture reveals Neutrophilia, Lymphopenia, Low haemoglobin and RBC count.

Table 1. a Complete Blood picture

Investigations	Observed values	Normal values
Neutrophils	82.8%	50-80%
Lymphocytes	12.7%	25-50%
RBC	3.65mill/mm^3	3.8-6.50mill/mm^3
Haemoglobin	9.1g/dl	11.5-17g/dl
Haematocrit	28.7%	37-54%

Table 2.b. Complete -Urine Examination

Investigations	Observed values	Normal values
Pus cells	3-4/HPF	0-4/HPF
Epithelial cells	2-3/HPF	0-5/HPF

Table 3.C. Liver function tests

Investigations	Observed values	Normal values
SGOT	168	5-45 U/l
SGPT	104	5-45 U/l
Alkaline phosphatase	173	28-88 U/l

Table 4.d. Erythrocyte sedimentation rate

ESR	Observed values	Normal values
ESR 1st hour	90mm	0-20mm/hr
ESR 2nd hour	140mm	0-20mm/hr

- e. Ultrasonography of the abdomen and pelvis indicated Grade III Fatty Liver.
- f. Thyroid profile was found to be normal
- g. 2D Echo indicated Mild LV Systolic Dysfunction.
- h. ANA profile indicated the presence of auto-nuclear antibodies.

The abnormal lab investigation and above clinical manifestations. Indicated that patient is suffering from diffuse cutaneous sytemic sclerosis

Table 5. Medication Chart

Drugs	Dose	Route	Frequency
Tablet prednisolone	10mg	Oral	Twice a day
Injection metrogyl	500mg	Intravenous	Thrice a day
Injection ceftriaxone	1 gram	Intravenous	Twice a day
Injection pantoprazole	40mg	Intravenous	Once a day
Tablet colchicine	0.5mg	Oral	Twice a day
Tablet Pentoxyphilline	400mg	Oral	Thrice a day
TabletHydroxyzine hydrochloride	25mg	Oral	Once a day
Capsule Calcitriol	1 capsule	Oral	Once a day
Tablet Ursodeoxycholic acid	300mg	Oral	Once a day
Tablet aspirin+clopidorgel+Rosuvastatin	20mg	Oral	Once a day
Tablet Cinitapride	1mg	Oral	Twice a day
Capsule Vitamin E	100 mg	Oral	Once a day
Aleo+vit E moistursing cream	-	Topical	Twice a day
Fluticasone cream	0.005%	Topical	Twice a day

DISCUSSION

Systemic sclerosis is a chronic disease of obscure etiology (Asokan *et al*., 2013). Although the exact pathogenesis of systemic sclerosis undetermined, the ascertained pathogenesis involves three characteristic features (a) Small vessel vasculopathy (b) Production of autoantibodies and (c)

Fibroblast disturbance leading to extensive deposition extracellular matrix (Frank van den Hoogen *et al*., 2013). Systemic sclerosis is sub classified into limited cutaneous systemic sclerosis and diffuse cutaneous systemic sclerosis this subtypes differ in their courses and prognosis (Chris and Derk, 2012; Kazuhiko Takehara, 2004). In diffuse cutaneous systemic sclerosis the fibrotic process is rapid and diffused to various internal organs like lungs, heart, gastrointestinal tract (John Varga and David Abraham, 2007). Swelling of skin is usually symmetric and progresses to induration in diffuse cutaneous systemic sclerosis, it may be confined to fingers (sclerodactyly) and hands or may affect most of the body (Asokan *et al*., 2013). Quick skin fibrotic progression and rapid visceral organ involvement is present in diffuse cutaneous systemic sclerosis (Chris and Derk, 2012). Auto antibody which is likely to be involved in diffuse cutaneous systemic sclerosis is Anti-topoisomerase-I (30% of patients). Survival rate in DcSSc is about 40-60% indicating the poor prognosis of disease (Chris and Derk, 2012). The various investigations used for diagnosis of sclerderma include nailfold capillaroscopy, ESR, test for auto antibodies (Chris and Derk, 2012). Sytemic sclerosis has no established treatment guidelines (Kazuhiko Takehara, 2004). The treatment should be directed towards organ specific therapies (Chris and Derk, 2012).

Conclusion

Lack of universally acceptable disease modifying agent makes systemic sclerosis a challenging disease for the treating doctors. Morbidity and mortality rates can be improved by early recognition, diagnosis, and aggressive management of affected organ systems. Long term follow up with screening tests performed at regular intervals is essential to reduce the risk of scleroderma complications. As Raynauds Phenomenon is hall mark feature of diffuse cutaneous systemic sclerosis, the patient should be adviced for conservative management which includes maintaining of core body temperature and avoiding peripheral cold exposure.

Abbreviation

SSc-systemic sclerosis, lSSc-limited cutaneous systemic sclerosis, DcSSc-diffuse cutaneous systemic sclerosis, ANA-Anti nuclear antibodies, ESR-Erythrocyte sedimentation rate.

Conflict of interest: There is no conflict of interest

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