

SYSTEMIC SCLEROSIS: A CASE STUDY IN AYURVEDIC SETTINGS**Mahesh MP¹, Niranjan Y^{2*}, Mayya Jagadeesha A³**

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ABSTRACT

Systemic sclerosis (SSc) is a multisystem disorder of unknown cause characterized by fibrosis of skin, blood vessels, and visceral organs including the gastrointestinal tract, lungs, heart, and kidneys. There are two distinct clinical presentations viz., Diffuse and limited forms. We are herewith reporting a case of diffuse cutaneous scleroderma in a 21 year old female student. The possible understanding of the case in terms of Ayurveda and a therapeutic protocol with promising result has been discussed.

KEYWORDS: Systemic Sclerosis, Scleroderma, Ayurveda, *Vatavyadhi*

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INTRODUCTION:

Scleroderma is derived from the Greek words *Skleros* (hard or indurated) and *derma* (skin). It was initially defined by Hippocrates, but a detailed description was given by Carlo Curzio (Sapadin AN, Fleischmajer R. 2002). Systemic sclerosis (SSc) is a multisystem disorder of unknown cause characterized by fibrosis of skin, blood vessels, and visceral organs including the gastrointestinal tract, lungs, heart, and kidneys (Fauci AS *et.al.*, 1998). There are two overlapping forms. Limited cutaneous scleroderma is limited to the skin on the face, hands and feet. Diffuse cutaneous scleroderma covers more of the skin, and is at risk of progressing to the visceral organs, including the kidneys; heart, lungs and gastrointestinal tract are affected.

Annual incidence is 19 per million, and prevalence is 19–75 per 100,000. Women are roughly four times more likely than men to develop systemic scleroderma (<http://www.clevelandclinicmeded.com>). Incidence is twice as high among African Americans, and the Choctow Native Americans in Oklahoma have the highest prevalence in the world (469/100,000). The interval of peak onset starts at age 30–35 and ends at age 50–55 (Bermas BL, 2009 and www.umm.edu) coinciding with the greatest changes in hormone levels (Chiffot H *et.al.*, 2009). There is some hereditary association, some suggestion of immune reaction (molecular mimicry) to a virus, and some cases caused by toxins (Kasper *et.al.*, 2005).

The outstanding feature of SSc is overproduction and accumulation of collagen and other extracellular matrix proteins in the skin and other organs. While the pathogenesis of SSc remains to be further elucidated, the disease process involves immunologic mechanisms, vascular damage, and activation of fibroblasts (Fauci AS *et.al.*, 1998). Clinical manifestations of Reynaud's phenomenon, skin thickening, telangiectasia are most commonly encountered. Arthralgias, myopathy, esophageal dysmotility, pulmonary fibrosis,

pulmonary hypertension are also observed (Fauci AS *et.al.*, 1998).

At present there are no specific diagnostic tests for SSc and the disorder is diagnosed primarily based on the collective appearance of a cluster of clinical symptoms, such as Reynaud's phenomenon, telangiectasias, esophageal dysfunction with gastro-esophageal reflux, characteristic pigmentary changes, or presence of digital ulcers or calcinotic lesions accompanying clinically detectable skin induration. It is well recognized that the presence of specific autoantibodies is one of the most common manifestations of SSc and greater than 90% of SSc patients harbor antinuclear antibodies in their serum. Numerous autoantibodies have been described in SSc patients, some of these are highly specific for SSc, including anti-Scl-70 and anticentromere antibodies, and these have, therefore, been used as diagnostic biomarkers to support or confirm the clinical diagnosis of SSc. Anti-Scl-70 antibodies are directed against DNA topoisomerase I and are almost exclusively present in the sera of patients with the diffuse form of SSc (Castro SV, Jimenez SA, 2010 and Jimenez SA, Derk CT 2004).

The many complications of scleroderma can have a major impact on a person's sense of well-being. Patients are greatly concerned about changes in their appearance, particularly those changes caused by tightening of the facial skin. Depression has great impact, along with pain, on reducing patients' ability to function socially (<http://www.umm.edu>).

Prognosis is difficult to predict until the disease differentiates into recognizable subsets. Patients with limited cutaneous scleroderma have a good prognosis, with 10-year survival of 75%, although <10% develop pulmonary arterial hypertension after 10–20 years. Patients with diffuse cutaneous scleroderma have a 10-year survival of 55%. Death is most often from pulmonary, heart and kidney involvement, although survival has greatly improved with effective treatments for kidney failure. Immunosuppressive drugs are used, although

glucocorticoids have limited application (www.en.wikipedia.org). Various therapeutic modalities are being used for the treatment of localized scleroderma. There is no precise treatment scheme for this disease. A majority of patients can be successfully treated with topical therapy and phototherapy, but the progressive forms of the disease with intensely expressed skin sclerosis sometimes may need even systemic treatment (Braun-Falco O *et.al.*, 2004). Hence an attempt is made to explain the condition in Ayurveda and to derive a treatment strategy.

CASE STUDY:

A 21 year old college student was referred from peripheral outpatient department to Kayachikitsa department of the central hospital on December 26, 2012 with complaints of gradually progressive feeling of tightness of the

skin and face with skin thickening. She also had joint pain involving small joints of hands & wrists, elbows, shoulders and knee. There was h/o difficulty in squatting, getting up from squatting position. She also had Raynaud’s phenomenon, digital calcinosis, weight loss since onset of illness with discoloration of the neck region, (Fig. No.1) hair fall and general weakness were also seen. Her ANA Profile for Scl 70 was ++ (strong positive). Pulmonary function tests were technically poor (restriction) and poor patient effort. Hemoglobin 11.4g%, Total leucocyte count 5950/mm³, Differential count was 59% of neutrophils, 38% of lymphocytes, 02% monocytes and 01% eosinophils, ESR was 10 mm/1st hr, routine urine examination was within the normal range.

Fig. No. 1: Clinical features



1.1 Shiny, taut facial skin, 1.2 Digital calcinosis, 1.3 calcinosis in the sole, 1.4 Skin induration, 1.5 Sclerodactyly, 1.6 & 1.7 Discoloration at elbow

Her vitals were stable with Height - 160 cm, Weight - 38 kg, B.P. - 100/70 mm of Hg. On examination, she demonstrated *srotodushti* (morbidly of channels) of *Annava* (food transportation) as *aruchi* (anorexia), *Rasavaha srotas* (channels carrying food) as *Aruchi* (anorexia), *pandutwa* (paleness) and *krushangata* (leanness). *Raktavaha srotas* (blood carrying channel) with Skin lesions, *Asthivaha srotas* (channels carrying osseous tissue) as *Vaivarnya* (discolouration), *asthisoola* (pain in bone), *Majjavahasrotas* (channels carrying components of bone marrow) as *parvaruk* (pain in joints) and *Swedavahasrotas* (channels carrying sweat) as *Aswedana* (absence of perspiration) and *parushyam* (roughness).

DISCUSSION:

Looking in to the clinical presentation of the case, the following probable diagnoses were contemplated; *Vatarakta*, *Amavata*, *Kushta*

(*obstinate skin diseases*) and *Twakgatavata* (*Vayu* located in skin).

The presentation of Reynaud's phenomenon on exposure to cold, shiny taut skin over extensor surfaces, sclerodactyly and digital calcinosis, the involvement of vitiated *Vata* (bio-humour) is obvious in terms of *shyavaarunavarna* (bluish discoloration) (Acharya YT, 1994) over extremities, an increase of *kharatwa* (roughness) and *rookshatwa* (dryness) of *Vata*. The salt and pepper discoloration pointing towards *Charmakushta* (Skin disorder characterized by thickening of skin) (Acharya YT, 1994) (Fig.No. 2), resembling the skin of an elephant. The thinning of skin is a feature in *TwakgataVata* (*vayu* located in skin) (Acharya YT, 1994). The digital calcinosis progressing in to rat-bite necrosis (Haustein UF, 2002) typically draws attention to the *akhuvisha* (rat bite poisoning) (Acharya YT, 2003 and Khunte AM, 2002) simile in *Vatarakta*.

Fig. No.2 Salt and pepper discoloration in the nape (image 2.1) resembling the skin of elephant



Salt and pepper discoloration resembling the skin of elephant
(actual unedited patient photograph)

A complex multifactorial etiopathogenesis was finally accepted based on the following logical sequence; the disease has hereditary role, supporting a genetic mutation and auto-immunity, similarly *Kushta* has *beejadosha* (genetic) as a factor. The peripheral features of scleroderma resembles the *laxanas* (signs and symptoms) of *Vatarakta* (Acharya YT, 1994); of both *uttana* and *gambhira* nature at various

instances of disease progression. The cutaneous manifestations mimic a particular variety of *Kushta* especially *Charmakhya* of *Vataja* type. The joint manifestations point towards *Amavata* (Murthy PHC, 2006) or *Sandhivata* (Khunte AM, 2002) features. However systemic involvement may denote various *srotodushti* (vitiating of channels) features. Hence, a complex pathological process duly

constituted by vitiated *Vata* by means of affliction of *twak* (skin) with the following *samprapti ghataka* (factors of pathogenesis); *Vatapradhana Kapha* are the involved *Dosha* (bodily humor), *Dushya* (vitiating bodily tissue) being *Twak* (skin), *Snayu* (ligaments), *Sira* (veins), *Kandara* (tendons) and *Jatharagni* (digestive fire) in *mandavastha* (diminished) and *Ama* (unprocessed food) born out of it. Many *srotas* (channels) are involved in the later stages with primary *Rasavaha Srotodusti* (morbid food carrying channels) of *Sanga* (obstruction) type followed by *Vimargagamana* (diversion of flow of the contents to improper channels). The *Doshakopa* (aggravated bodily humor) begins at *Amapakwashaya*, moving through *Rasayani* (circulatory channel) and settling at *Twak* (skin). Scleroderma being a physical illness, *adhithana* (substratum) is *Shareera* (physical body). The disease has *Bahyarogamarga* (external pathological route) involved extending in to *Madhyama* (median route of pathogenesis). Oil application slightly improving the dryness and tightness of skin may be considered as *Upashaya* (symptomatic relief). *Upadrava* like Pulmonary hypertension, involvement of *Sandhi* (joints), *Sira* (blood vessels), *Snayu* (ligaments) are observed as complications.

The conclusion regarding the diagnosis was derived as a complex syndrome dominated by *prakupita Vata* (vitiating vata) taking *ashraya* (base) in *rasa* (circulating fluid tissue), later the *samprapti* (pathogenesis) extends to deeper tissues especially *sira* (blood vessels), *kandara* (tendons), *snayu* (ligaments) and *asthisandhi* (bony joints). In therapeutic perspective, the condition is managed with *Vataprasamana* (pacifying *Vata*) and *raktaprasadana* (pacifying *Rakta*) line of treatment in accordance with the *chikitsa* (treatment) advocated for *Vatavyadhi* (Acharya YT, 1994 and Khunte AM, 2002) and

Vatarakta (Acharya YT, 1994) in classical Ayurvedic literatures.

TREATMENT PROTOCOL:

On the day of admission, *Agnideepana* (improving digestive fire), *amapachana* line was adopted using *Hingvashtaka choorna*. *Abhyanga* (oil massage) was done in the next two days with *Yashtimadhu taila* and *sweda* (sudation) using the *Nadee sweda* apparatus. On the third day, *Koshtashodhana* was done with *Gandharvahastadi eranda taila* 30 ml in the morning at 6.30 AM. *Avarasuddhi* was observed. *Samsarjana* advised for three *annakala* (time of food intake).

Yogavasti (medicated enema schedule) is planned considering the *Samprapti Ghataka* with *Anuvasana* (oil enema) of *Yashtimadhu taila* and *Ksheerabala taila* 60 ml with 5 g of *saindhava lavana* *Erandamuladi nirooha* was carried out as *Asthapanavasti* (decoction enema) using *Erandamula kwatha*-500ml, *Ksheerabala taila* + *yashtimadhu taila*-75ml, *Madhu* (honey)-25ml, *Saindhava* (rock salt)-10g and *Sathapushpa kalka* (*Anethum sowa* powder)- 20 g.

On discharge, she was prescribed with *Manjishtadi Kwatha* 25ml twice daily, Tablets of *Kaishora guggulu* 2 with *Kwatha*, *Kamadugha* rasa tablet twice daily. *Guggulutikthaka ghrita* 10 ml after food as *shaman sneha*. *Yashtimadhu taila* was advised for external application.

RESULTS AND CONCLUSION

After 3 months of active treatment in the above lines, the patient was happy with relieved tightness over skin of face and limbs, improved ability to perform normal activities and an enhanced overall quality of life. This example may be a silver lining in the horizon for the mankind with Scleroderma.

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