

CASE STUDY

Management of Acrosclerosis through *Ayurveda* – A case Report

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ABSTRACT

Acrosclerosis is a generalized disorder of connective tissue in which fibrosis and degenerative changes predominate. A 45 year old female suffering from Acrosclerosis having complains of pain and stiffness in proximal and distal interphalangeal joints and tightening of the skin over dorsum of hands, blackish patches on nose, lips and face, flexed fingers was treated on the line of treatment of *Gambhir Vatarakta*. *Vatavidhwamsana Rasa* 125mg, *Yogendra Rasa* 125mg, *Amritadi Guggulu* 250mg, *Tiktaka Ghrita* 10ml, all twice a day and *Kaala Basti* of *Dasamoola Taila* and *Kwatha* were administered. She was also underwent *Raktamokshana* (Leech therapy). Patient got symptomatic relief in pain and swelling. The skin became smooth. She performs routine activities much better after the treatment. This case study demonstrates that Ayurvedic principle of *Gambhir Vatarakta* is effective in the management of Acrosclerosis.

Keywords : *Ayurveda*, *Kaala Basti*, *Gambhir Vatarakta*, Acrosclerosis, *Raktamokshana*

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Introduction:

Acrosclerosis causes stiffness of the skin of the fingers, atrophy of the soft tissue of the hands and feet, and osteoporosis of the distal phalanges, along with Reynaud's phenomenon. The term scleroderma is derived from a Greek term 'scleros', which means hard and 'derma' which means skin. It is a generalized disorder of connective tissue in which fibrosis and degenerative changes predominate. The most important features of the disease are the thickening and hardening of the skin and distinctive involvement of multiple internal organs, most notably the lungs, gastrointestinal tract, heart, and kidneys^[1]. Scleroderma is mainly classified into diffuse and limited form based on the extent of skin involvement and presence of lesions in internal organs. The prevalence rate of this disease is around 5/100,000^[2] The disease is commoner in females. The male to female ratio is 1:4.

It's usually found in the fourth and fifth decades. The initial symptoms include tightness of the extremities and arthralgia. Initial manifestations are edema of the hands and feet, which may extend more proximally. The edema is followed by thickening and tightening of the skin. Skin loses its normal pliability and ultimately becomes "hide bound" and non-pinchable. Later, skin of face gets involved and gives rise to the 'mouse head' appearance with microstomia, beaked nose, mask like expression, and difficulty in opening the mouth. There will be pigmentary changes developing over the extremities and anterior chest wall. These are hyper-pigmentation or a combination of spotty hyper and hypo-pigmentation (salt and pepper skin), and telangiectasia over the fingers, palms, face, lips, and tongue. Painful, non-healing, chronic, indolent ulcers are common over the fingers and toe tips^[3]

Diagnosis is mainly done clinically. The most important finding is the hard skin, which is not pinchable and devoid of hair. Laboratory investigations help to rule out other conditions. Erythrocyte sedimentation rate (ESR) is moderately raised but it may be normal in early cases and in cases where only the skin is affected. IgG may be elevated during the active phase. Rheumatoid factor is positive in 20-25% and Antinuclear factor (ANF) in about 40% cases in low titers. ANA subset tests can help distinguish two forms of the disease, limited versus diffuse. The limited form is most closely associated with the anti-centromere pattern of ANA staining, while the diffuse form is associated with autoantibodies to Scl-70. Progressive Acrosclerosis can occur with other connective tissue disorders like systemic lupus erythematosus (14%), rheumatoid arthritis (8.4%), and mixed connective tissue disease (2.8%). It has to be distinguished from other collagen disorders like dermatomyositis and mixed connective tissue disease. In other disorders like eosinophilic fasciitis, scleredema adultorum Bushke, myxedema, and acromegaly there may be thickening and tightness of the skin.^[4]

Presently there is no treatment in modern medicine to halt the progression of the disease. The intervention are mainly done to relieve the symptoms like arthralgia and

to slow the progression of organ damage.^[5]

This is a case report of a patient of progressive Acrosclerosis which was managed according to the line of management of *Gambhir Vatarakta*.

Case Report:-

A 45 year old female came to OPD on dated 06.12.2016. The patient had complains of pain and deformities of proximal and distal inter-phalangeal joints of both hands for last three years. Fingers of both hands of the patient become bluish in winter season as a first symptom 10 years before. It started with the index finger of right hand and later spread to other fingers. Later pain started in the proximal and distal inter-phalangeal joints. She also noticed swelling and stiffness of the joints. Her fingers were stucked in flexed position and she was unable to extend fingers due to pain. There were pus pockets on the fingers. (Figure 3) Later on, red spots appeared on her face and lips, which became darkened after some days. Family history was negative for similar condition. Laboratory investigations were as follows- Hemoglobin (12.8%), ESR (8mm/hr) and serum uric acid (3.9mg/dl) were in normal range and qualitative RA Factor was negative. IgG (1329 mg/dl) was also under reference range. In ANA profile, Proliferative cell nuclear antigen (PCNA) was positive (SLE-3-7%), Scl-70 was positive (Progressive Acrosclerosis (diffuse form-65%), AMA-M2 was positive (PSS-7-25%, Chronic liver disease -7-25%). (figure 1) Above explained laboratory parameters confirmed the diagnosis as progressive Acrosclerosis. X Ray of hand was also performed. (Figure 2) The Patient was admitted in indoor patient department of the hospital on dated 20/1/17. She was advised *Vataavidwamana Rasa* 125mg, *Yogendra Rasa* 125mg, *Amritadi Guggulu* 250mg and *Panchathiktaka Ghritha* 10ml orally. At the same time *Kaala Basti* was administered with *Dasamoolataila* and *Dasamoola Kwatha*. *Raktamokshana* was done once in month by leech application. (Figure 5) Patient got symptomatic relief of pain and swelling disappeared. The skin of the fingers which was tight became smooth. She was able to do daily activities normally and was also able to extend her fingers more. The patient was on follow up for every fifteen days for next two months and thereafter

gradually had symptomatic relief.

Table no.I Showing comparison of signs and symptoms of patient before and after treatment

Symptoms	Normal	Before treatment	After treatment
Range of motion at time of Flexion at proximal interphalangeal joint	100°	30 °to 60 °	60 °to70 °
Range of motion at time of Extension at proximal interphalangeal joint	0 °	-45 °to -30 °	-30°to -15 °
Range of motion at time of Flexion at Distal interphalangeal joint	80 °	30 °to 45 °	45 °to 60 °
Range of motion at time of Extension at Distal interphalangeal joint	0 °	-30 °to-15 °	-20 °to -15 °
Tightness of skin of fingers	-	+++++	+

Figure 1 showing scanned reports of ANA profile of the patient

ANA BY IMMUNOBLOT (LINE IMMUNO ASSAY)*		
The assay detects ANA of IgG class against 17 different antigens :-		
Test Description	Observation	Disease Association & Prevalence
dsDNA	Negative	SLE (40-95%)
Nucleosomes	Negative	SLE(40-70%)
Histones	Negative	Disseminated Lupus Erythematosus (30-70%), Rheumatoid arthritis (15 - 50%)
SmD1	Negative	SLE (15- 40%)-highly specific (> 95%)
PCNA (Proliferating Cell Nuclear Antigen)	Positive	SLE (3-7%)
Ribosomal P0-Protein	Negative	SLE (10-35%)
SS-A/Ro60	Negative	Sjogren syndrome(Upto 96%), Neonatal lupus erythematosus (100%)
SSA/Ro-52	Negative	Congenital Heart Block in Neonatal Lupus and with cutaneous lupus manifestations.
SS-B/La	Negative	Sjogren Syndrome(50-70%)
CENP-B	Negative	Systemic Sclerosis (CREST syndrome: Calcinosis, Raynauds disease, Esophageal dysfunction,Sclerodactylia,Teleangiectasis), Scleroderma (95%)
Scl-70	Positive	Systemic sclerosis (100%), Progressive systemic sclerosis -65%, (diffuse form)
U1- snRNP	Negative	Diagnostic marker of mixed connective tissue disease (MCTD),

Figure 2 showing X-Ray of hand showing flexion of proximal and distal interphalangeal joint of second, third and fifth digit of right hand and fifth digit of left hand



Figure 3 showing pus pocket on proximal inter-phalngeal joint of third digit of right hand



Figure 4 showing pus pocket on proximal inter-phalngeal joint of third digit of right hand Before treatment



Figure 5 Showing leech application on affected region



Figure 6. showing dark black spots on the face and lips in photographs A and B before and after treatment respectively.



Figure 7 showing after treatment :- Digits of right hand are in relaxed position as compared to before treatment condition



7. Discussion

The main features of progressive Acrosclerosis are stiffness of the skin of the fingers, atrophy of the soft tissue of the hands and feet, and osteoporosis of the distal phalanges, tightness of the extremities and arthralgia and distinctive involvement of multiple internal organs, most notably the lungs, gastrointestinal tract, heart, and kidneys. The tight atrophic skin is particularly vulnerable to trauma. Edema, ulcers are common over the fingers and toe tips.

Vatarakta is a variety of 80 *Vata Roga*. Ingestion of foods which are predominantly *Lavana*, *Amla*, *Katu* and *Kshara*, having irregular diet habits, sleeping during day time, lack of physical exercise etc. are cause of *Vata* and *Rakta Dushti*.^[6] *Vata* vitiated by its own etiological factors, while circulating through its own *Srotases*, is blocked or encircled (*Avarnam*) by vitiated *Rakta* and

gets more vitiated. *Vata* in this state in combination with vitiated *Rakta*, produces the disease *Vatarakta*.^[7] Vitiated *Vayu* and *Rakta* spreads all the body and it gets accumulated in the phalanges of hands and legs producing inflammation, pain warm touch, redness like symptoms. Afterwards it produces these symptoms in other joints.^[8] The disease is stated to be of two varieties on basis of its site of pathogenesis, *Uttana* in which disease is confined to *Twak* and *Mamsa Dhat* and *Gambhir* in which disease is situated in remaining *Dhatus*.^[9]

The signs & symptoms of *Gambhira Vatarakta* are stated as following – swelling like nodules which are hard and painful, covered by dark, copper colored skin. There is burning sensation, pricking pain, throbbing sensation and these swellings are likely to inflame and suppurate also.^[10]

Thus, signs and symptoms of progressive Acrosclerosis

have the resemblance with the manifestation of *Gambhir Vatarakta*.

In this case the patient was presented with blackish patches on upper limbs and face having arthralgia and deformities of interphalangeal joints which was very similar to Rheumatoid Arthritis, but RA factor of this patient was negative. These symptoms were very similar to *Gambhiravatarakta*. So, the management was based on the line of treatment of *Vatarakta*. *Basti* and *Raktamokshan* is the best treatment for vitiated *Vata* and vitiated *Rakta* respectively. *Shaman* treatment was also given along with *Basti* and *Raktamokshan*. *Vatavidhwamsana*, *Yogendra Rasa*, *Amritadi Guggulu*, *Tiktaka Ghrita*, *Dasamoola Taila* & *Dasamoola Kwatha* are *Vatashamaka* and *Amritadi Guggulu*, *Tiktaka Ghrita* are *Pittashamaka* & *Raktashodhaka*. Combined effect of these drugs has capabilities to address all the manifestation of Progressive Acrosclerosis. Patient had got symptomatic relief in pain and swelling. The skin became smooth and she does her routine activities normally.

8. Conclusion:-

Progressive Acrosclerosis has close resemblance with *Gambhira Vatarakta*. This case of erythroderma was successfully managed in the line of treatment of *Gambhira Vatarakta*. This case study demonstrates that Ayurvedic management is effective in progressive Acrosclerosis symptomatically.

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सारांश:

अक्रोस्केलेरोसिस एक सामान्यीकृत विकार है जिसमें विशेषतया त्वचा अधिक मोटी और सख्त हो जाती है, हाथ और पैर की संधियों में जकड़ाहट और कई आंतरिक अंग भी प्रभावित होते हैं। इस रोग की व्यापकता दर लगभग 5/100,000 है। अक्रोस्केलेरोसिस से पीड़ित 45 वर्षीय एक महिला जिसकी अंगुलियों की संधियों में दर्द और जकड़न, दोनों हाथों की त्वचा सख्त, उसकी नाक, होंठ और चेहरे पर काले धब्बे, दर्द के कारण अपनी अंगुलियों का विस्तार करने में असमर्थता थी छ उसका उपचार गंभीर वातरक्त के समान किया गया। इस रोगी में वातविध्वनसन रस 9२५ मिलीग्राम, योगेंद्र रस 125 मिलीग्राम, अमृतादि गुग्गुलु २५० मिलीग्राम, तिक्तक घृत 9० मिलिलीटर की मात्रा में दिन में दो बार दिया गया और दशमूल तैल और क्वाथ की काल बस्ति, जलौका से रक्त विस्त्रावन करवाया गया | इस उपचार से रोगी को दर्द और सूजन में राहत मिली। त्वचा मुलायम हो गई। वह अपनी दिनचर्या एवं अन्य गतिविधियों को सामान्य रूप से करने में समर्थ हो गई | गम्भीर वातरक्त का आयुर्वेदिक सिद्धांत अक्रोस्केलेरोसिस के उपचार में प्रभावी है।