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Limited Sclerosis- A Rare Connective Tissue Disorder- A Case Report

Authors
Rohit Ram Kumar[1], Madhu G Kandathil[2], Arathi N[3], Suresh Raghavan[4]
[1]Junior Resident, [2]Lecturer, [3]Assistant Professor, [4]Professor
Dept of General Medicine, TD Medical College, Alappuzha, Kerala, India – 688005

ABSTRACT
Limited cutaneous scleroderma is a rare connective tissue disorder characterised by thickening of skin limited to the distal aspect of limbs and face and the basic pathology is functional and structural vascular alterations, inflammation, sustained fibroblast activation, fibrogenesis and impaired matrix degradation.
They often describe long standing Raynaud's phenomenon, GERD, may have telengectasias, skin calcifications and sclerodactyly.
Here we present a case of limited scleroderma presenting in the early phase of the disease with skin involvement alone.

INTRODUCTION

Scleroderma is a connective tissue disorder with multisystem involvement, and follows a chronic progressive course, with thickening of the skin being the hallmark of the disease.

It is of diffuse and limited variety.

In the diffuse variety, thickening and in duration of skin progresses from the distal limbs to the proximal aspect and may also involve the trunk and face

The limited variety usually spares the trunk and is confined to the distal limbs and face. Renal crisis is exceedingly rare in LcSSc.

Among those with the limited variety anti-Scl70 positivity is associated with a high risk of developing interstitial lung disease and anticentromere antibody positivity is associated with increased risk of developing PAH.

Interestingly, ANA positivity and anti Scl70 negativity are at high risk of developing ILD and PAH.

CLINICAL HISTORY

60 yr old female farmer with no comorbidities hailing from Alappuzha district of Kerala presented with gradually progressive and symmetric thickening of skin of both the upper limbs below the elbow and both lower limbs below the knee joint for 8 months. She also had:

- Uncomfortable feeling on the fingertips on coming into contact with cold water for 6 ½ months.
- Thickening of facial skin and appearance of white patches on the forehead and tip of nose for 5 months.
- No history of thickening of skin over the trunk
- No history of discolouration of fingers on exposure to cold
- No history of dysphagia/ dyspeptic symptoms/ symptoms of thyroid dysfunction

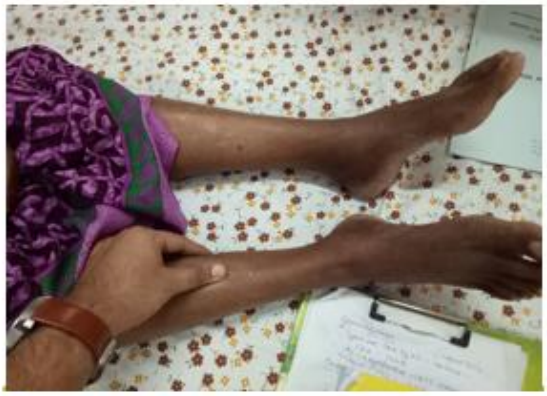
- No history of frothing of urine/oliguria/ periorbital puffiness early in the morning
- No history of small joint pain/deformity/ morning stiffness/ oral ulcers
- No history of dyspnoea on exertion/angina/orthopnoea

CLINICAL POSSIBILITIES

- Scleroderma (Limited Variety As Trunk Is Spared)
- Hypothyroidism
- Eosinophilic Fascitis
- Dermatomyositis
- Scleredema

ON EXAMINATION

- Shiny stretched skin of face
- Beaked nose with Vitiligo on the tip of nose
- Forehead shows salt and pepper pigmentation
- Stretched and shiny skin of fingers and the forearm
- Bulbous appearance of the tip of fingers
- Thick skin below the level of elbows and knees
- No microstomia/radial furrowing/ telengectasias
- Vitals were normal
- All systems were within normal limits



- Blood routine examination with ESR, ECG, RBS, Lipid profile, URE, Chest Xray, liver and renal functions were normal
- We also did S.TSH, which was normal.
- S.electrophoresis, S. calcium, phosphate, S.uric acid were normal.
- Rheumatoid factor, viral markers and ANA profile were negative
- We then proceeded for the autoantibodies in scleroderma and it was found that anti SCl-70 was strongly positive and anti centromere antibody was negative
- Subsequently we did an HRCT chest and an echocardiography to see if there was

ILD and PAH respectively. Pulmonary function tests were also normal

- Interestingly both came out to be normal. We also did an Xray of the hand to see for acralosteolysis and it was absent
- Finally we proceeded towards the confirmatory step of skin biopsy and it showed a normal epidermis
- And Dermis showed a *perivascular lymphocytic infiltrates* which is a sign of early disease
- Thus we came to a conclusion of a case of limited systemic sclerosis with no organ involvement presenting at an early stage

TREATMENT

- We started the patient on Hydroxychloroquine and tab nifedipine also as we suspected the onset of Raynaud's phenomenon
- She started feeling better and cold intolerance of the fingertips has improved
- Now she is on close follow up from our side and she is better

DISCUSSION

- Thus we have a rare case of limited scleroderma presenting to us at an early stage with thickening of skin alone at present
- The hallmark of the disease being excessive production of normal type -1 collagen with accumulation of fibronectin and glycosamino-glycan in the extracellular matrix.
- There is loss of sweat glands and hairloss on the areas of tight skin. Skin thickening virtually begins on the hands in all cases of scleroderma and if it occurs elsewhere, the possibility of *morphea, eosinophilic fasciitis or other mimics should be considered*
- Early diagnosis improves the outcome and morbidity of the patient due to early introduction of treatment and follow up

DIFFUSE VS LIMITED SCLEROSIS

	LIMITED VARIETY	DIFFUSE VARIETY
Skin involvement	90%	100 %
Raynaud's phenomenon	99	98
Oesophageal involvement	90	80
Pulmonary fibrosis	35	65
PAH	15	15
Myopathy	11	23
Cardiac involvement	9	12
Scleroderma renal crisis	2	15

- The Modified Rodnan skin score has been used clinically and on trials to document skin involvement
- The score is calculated by examining *17* areas and each area graded from 0 to 3(severe involvement) for a total possible score of 51. Scores of 15-20 and rapid progression in the 1styr of the disease are associated with more severe disease
- Internal organ involvement does not mimic skin involvement and may worsen with time.

CONCLUSION

- We decided to choose this case as limited scleroderma is a very rare entity and we could diagnose the patient with the disease at the earliest. Now she can be put on close follow up to see for the development of any symptoms of organ involvement.
- Thus we can reduce the mortality and morbidity associated with the disease

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