Original Research Paper

Volume - 13 | Issue - 04 | April - 2023 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

General Medicine

A CASE OF SYSTEMIC SCLEROSIS SINE SCLERODERMA

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ABSTRACT Systemic sclerosis is an uncommon connective tissue disorder with multisystem involvement, heterogenous clinical manifestations and have a chronic, progressive course. Autoimmune damage, vasculopathy and extensive fibrosis are the key etiopathogenic factors. Patients have clinical manifestations ranging from skin thickening limited to distal extremities/face to widespread multiorgan involvement. All typical features of systemic sclerosis without skin thickening is defined as SYSTEMIC SCLEROSIS SINE SCLREODERMA Case presentation: We report a case of A 40 year old female patient presented to us with the chief complaints of ulcers at the tip of right index and middle fingers associated with pain and swelling over a period of 10 days.Past history of amputation of left distal phalanx 1year back for similar complaint. She had a history suggestive of Reynaud's phenomenon. She was diagnosed as hypothyroid 5 years back with irregular treatment and with GERD 2ys back. CT CHEST-bilateral ground glass opacities,Paratracheal lymphadenopathy,Mild thyromegaly ,Mild dilatation of oesophagus,COLOUR DOPPLER-Right upper limb -normal study ,ANA PROFILE -Anti scl -70 –positive ,Anti jo 1-borderline positive **CONCLUSION :** Systemic sclerosis sine scleroderma (ssSSc) is an infrequent SSc variant characterized by visceral and immunological manifestations of SSc in the absence of clinically detectable skin involvement with good prognosis compared to other variants

KEYWORDS : SYSTEMIC SCLEROSIS SINE SCLREODERMA , Anti scl -70 –positive Anti jo 1-borderline positive

INTRODUCTION :

Systemic sclerosis is an uncommon connective tissue disorder with multisystem involvement, heterogenous clinical manifestations and have a chronic, progressive course. Autoimmune damage, vasculopathy and extensive fibrosis are the key etiopathogenic factors. Patients have clinical manifestations ranging from skin thickening limited to distal extremities/face to widespread multiorgan involvement. All typical features of systemic sclerosis without skin thickening is defined as SYSTEMIC SCLEROSIS SINE SCLREODERMA

CASE REPORT :

A 40 year old female patient presented to us with the chief complaints of ulcers at the tip of right index and middle fingers associated with pain and swelling over a period of 10 days.

Past history of amputation of left distal phalanx 1year back for similar complaint. She had a history suggestive of Reynaud's phenomenon. She was diagnosed as hypothyroid 5 years back with irregular treatment and with GERD 2ys back.

GENERAL EXAMINATION :

Normal except for the presence of ulcers over right fingers and amputat ed distal phalanx of left finger

SYSTEMIC EXAMINATION:

RS -Bilateral basal crepts on auscultation
CVS- Normal
CNS – Norml
GIT – Normal

INVESTIGATIONS :

Haemogram-normal
LFT - normal
RFT-Blood urea -34mg/dl,
Sr.creatinine - 1mg/dl
THYROID PROFILE –
T3-0.8ng/ml
T4-7.1ug/ml
TSH-10.63MIU/ml
URINE ANALYSIS –normal
ECG-normal
2D ECHO –normal

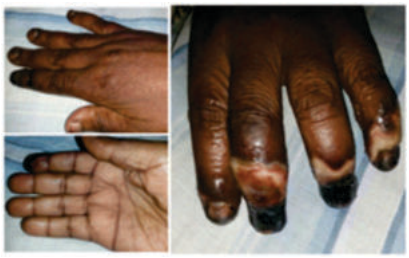
CT CHEST-bilateral ground glass opacities
Paratracheal lymphadenopathy
Mild thyromegaly
Mild dilatation of oesophagus

COLOUR DOPPLER-Right upper limb -normal study

ANA PROFILE :

Anti scl -70 –positive
Anti jo 1-borderline positive

CHARACTERISTICS :



Digital necrosis : Sharply demarcated necrosis of the fingertip secondary to ischemia in a patient with limited cutaneous systemic sclerosis (SSc) associated with severe Raynaud's phenomenon.

DISCUSSION:

An elderly female who presented with painful digital ulcers with amputated distal phalanx without any skin thickening was investigated and to have hypothyroidism, GERD, Interstitial lung disease with scl-70 positive antibodies comes under variant of SSC -SYSTEMIC SCLRESOSIS SINE SCLREODERMA. Patient was started on treatment with cyclophosphamide and bosentan. There was improvement in the condition with decreased pain.

CONCLUSION :

Systemic sclerosis sine scleroderma (ssSSc) is an infrequent SSc variant characterized by visceral and immunological manifestations of SSc in the absence of clinically detectable skin involvement with good prognosis compared to other variants.

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