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A CASE OF SERO-NEGATIVE SCLERODERMA WITH ILD

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ABSTRACT
Systemic sclerosis is an uncommon connective tissue disorder characterized by multisystem involvement, heterogeneous clinical manifestations, a chronic and often progressive course, and significant disability and mortality.[1] It is diagnosed in the presence of characteristic clinical findings and is supported by specific auto antibodies in 90% of cases.[2] The lung is involved in the great majority of cases of scleroderma, with post-mortem series indicating a 70% to 100% incidence. Prior to the use of vasodilator therapy, the mean survival following a diagnosis of pulmonary hypertension was approximately 2 years.[3] Several case reports were published as Seronegative Scleroderma.[4] We report a case of systemic sclerosis who presented as interstitial lung disease where all specific auto antibodies were negative.

CASE REPORT
A 35 years old male, non smoker, Farmer by occupation, referred to our department as a case of ILD with complaints of breathlessness for the past 1 year which was insidious onset progressive nature. Patient also had dry cough and mild hemoptysis and low grade fever for past one week. Patient denied the H/O chest pain, abdominal pain and other constitutional symptoms.Patient had h/o Raynaud's phenomenon for past 2years. He took ATT 6years ago for 6 months. No other significant exposure history, drug intake history in the past. On General examination patient had clubbing, pitting pedal edema. On examination of the respiratory system B/L fine end inspiratory creps present. On examination of CVS loud P2 and Mid Systolic Murmur were present over the pulmonary area. On examination of the skin following findings were present sclerodactyly upto DIP, stretched shiny skin over the face, decreased mouth opening, pinched beak nose (Figure - 1), finger tip pitting scars, fixed flexion contraction of fingers (Figure - 2). And his saturation often not recorded with puseoximetry.

Investigations
6MWT

	SpO2	PR	Distance	BORG scale
Before	90%	86 bpm		0
After	84%	120 bpm	300 meters	7

Patient was unable to complete the test due to dyspnea.

PFT

	Pre (%pred)	Post (%pred)
FVC	47.4%	49.6%
FEV1	49.1%	50.1%

CECT THORAX: B/L reticular thickening& B/L honeycombing (Figure - 3) with pulmonary hypertension (Figure - 4).

2D echo: EF 56%, Dilated RA, RV, moderate PH, RVSP – 57 mmHg.

Serologic Tests: RA factor, ANA, Anti ds DNA Ab, Anti Scl70, Centromere, Sm, Jo –1, SSA, SSB, RNP, ANCA, Anti GBM Ab ALL WERE NEGATIVE.

Diagnosis
According to ACR / EULAR criteria.[5]

Item	Score	
Skin thickening of fingers upto DIP	-	4
Finger tip pitting scars	-	3
Pulmonary hypertension and/or ILD	-	2
Raynaud's phenomenon	-	3
		12

Our patient had score of 12 (more than 9). This is consistent with diagnosis of Systemic Sclerosis.

Our patient improved with following drugs - cyclophosphamide, nifedipine, bosentan, diuretics & is doing well on our regular follow up.

Figure 1: Pinched up nose.

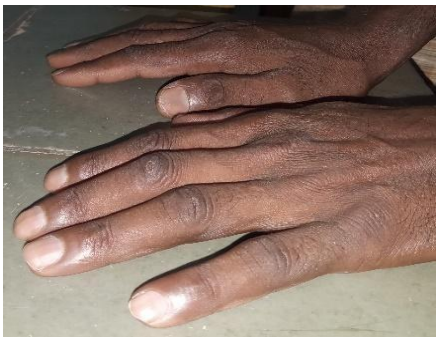

Figure 2: Fixed flexion deformity.

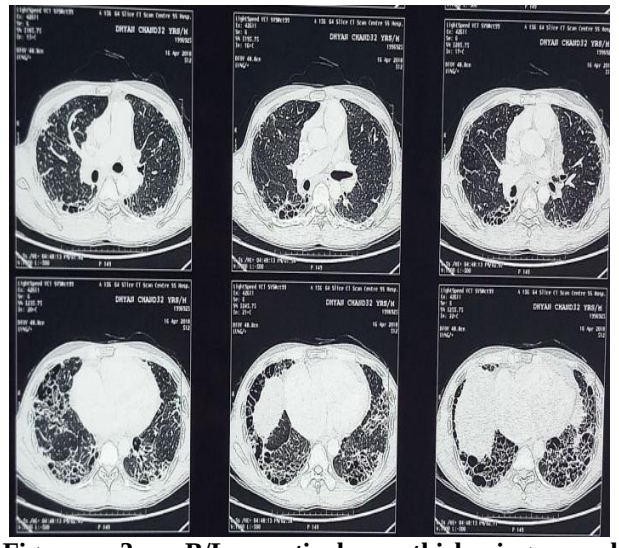

Figure 3: B/L reticular thickening and honeycombing.

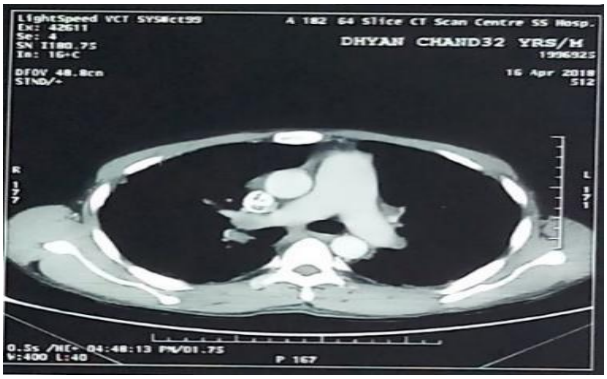

Figure 4: Enlarged pulmonary artery.

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