ERASMUS SYNDROME

A Rare Case Presentation

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Moderator : Dr.R.Ramakrisha Professor &HOD

Department of Respiratory Medicine

- A 45-Year-old female patient, who is stone crusher by occupation, resident of prakasham district came with chief complaints of
- -Difficulty in breathing since 15 days
- -Dry cough since 15 days

History of present illness

- Patient has on and off breathlessness from last 7 years, since 15 days it increased in severity which of grade 4 acc to MMRC. No postural and diurnal variations, no aggravating and relieving factors
  - No H/O of orthopnea, PND.
  - Dry Cough since 15 days .
  - No postural and diurnal variations present
  - No C/O chest pain,wheeze, palpitations, decreased urine output and swelling of feet, loss of weight/appetite

Past history

- Patient was treated for above complaints with ATT on clinicoradiological basis in 2018
- She was referred to rheumatologist and she was diagnosed to have scleroderma and treated with immunosuppressants.
- H/o usage of inhalers on and off since 2 years.
- Not a known case of Bronchial asthma ,COPD , COVID ,.
- Not a known case of Diabetes Mellitus , Hypertension, Thyroid, CAD, CKD, CVA,Epilepsy.

Treatment history

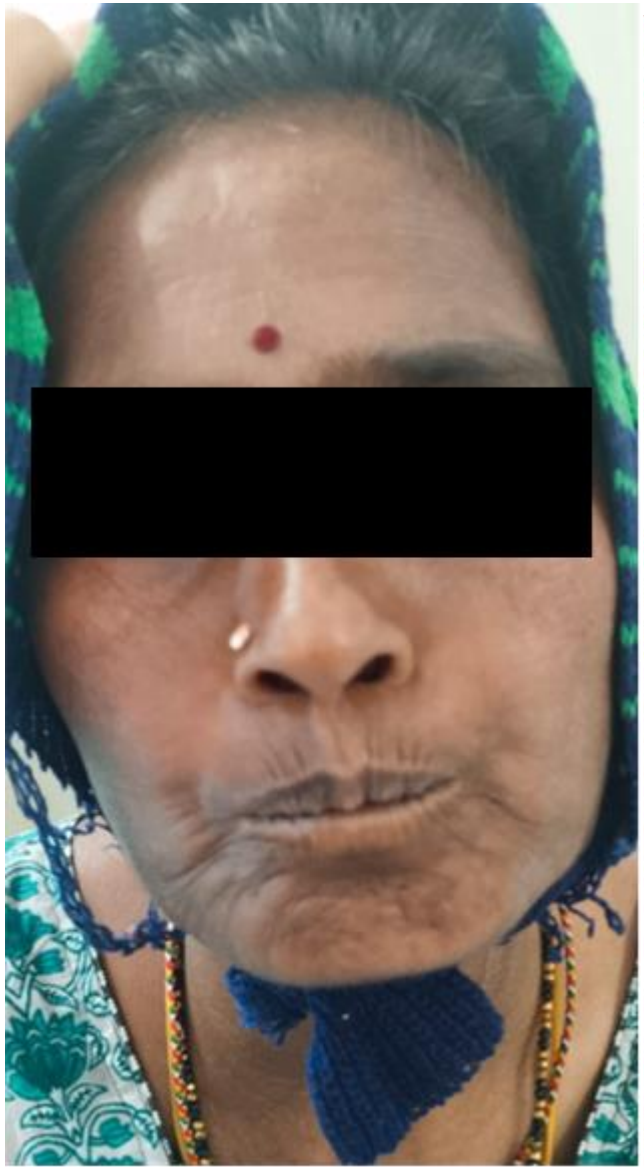
- History of usage of ATT FOR 6 months in 2018
- TAB HYDROCHLOROQUINONE 200 MG PO OD 0-0-1
- TAB PREDNISOLONE 5MG PO BD ½ - 0 - ½
- TAB METHOTREXATE 10MG PO ONCE A WEEK

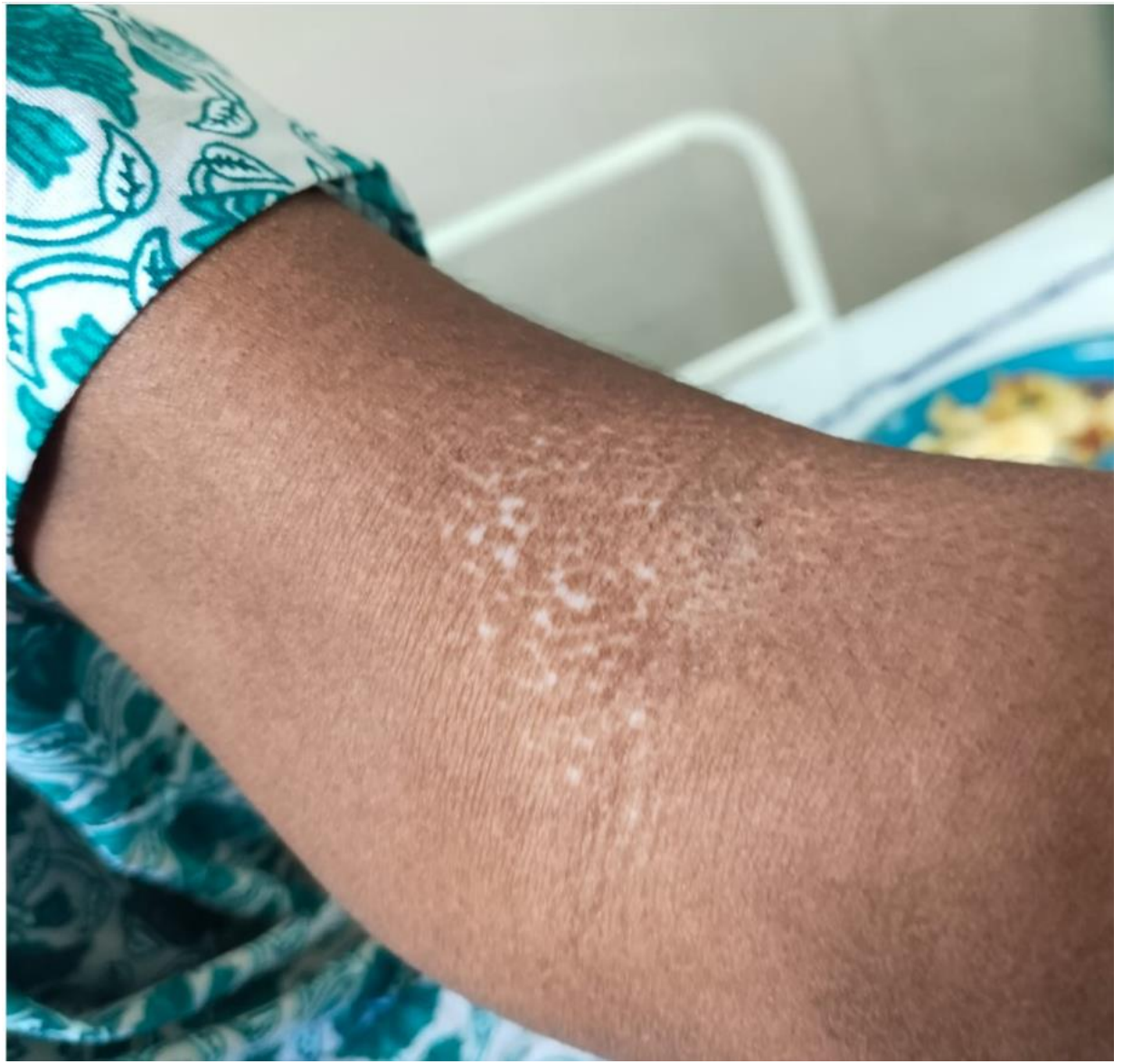
Personal history

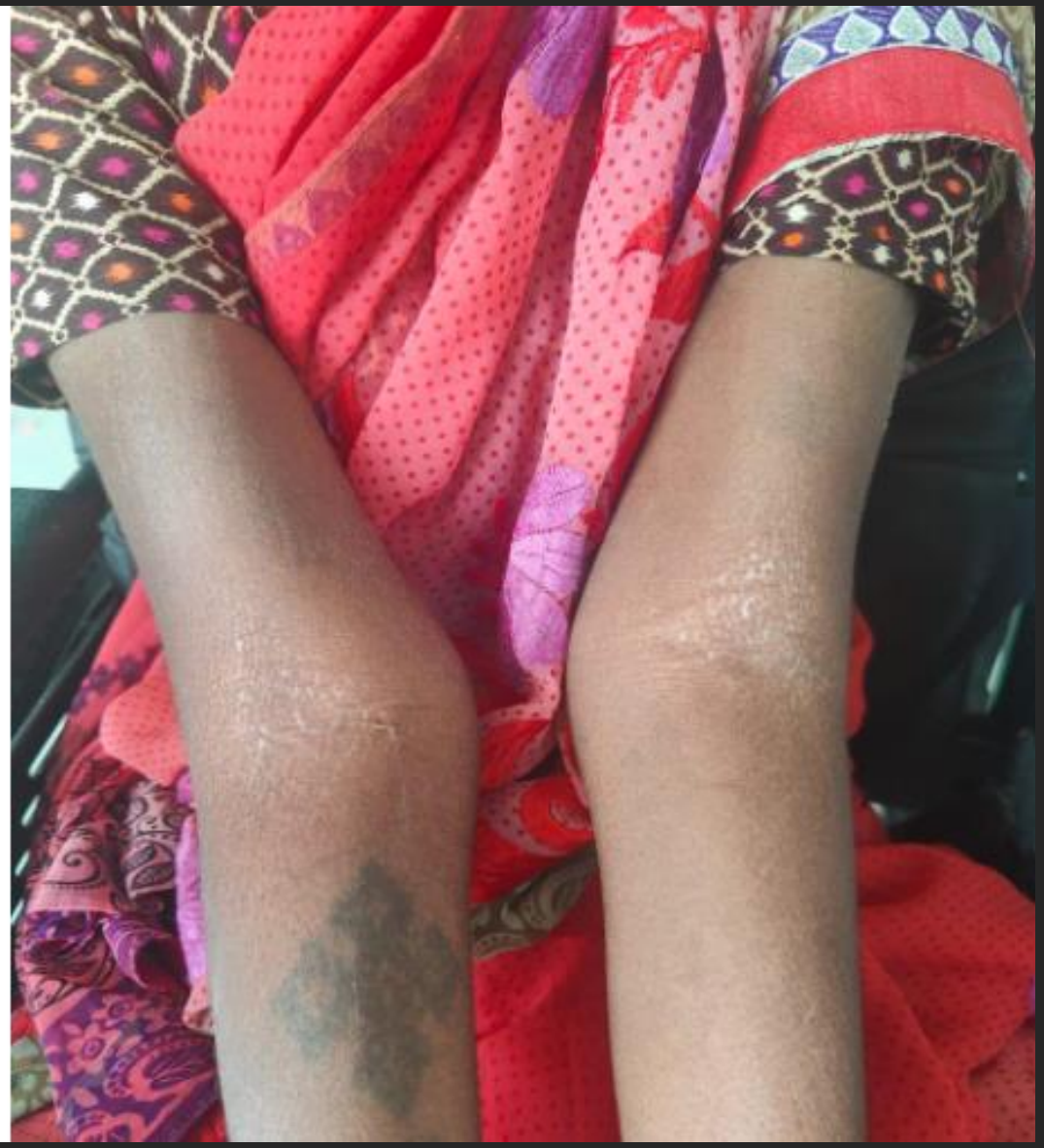
- She takes mixed diet.
- Normal sleep and appetite
- Regular bowel and bladder habits
- No H/o Smoking and alcohol
- Patient is married and having 2 children.

General Examination

- Patient is Conscious , coherent , well oriented to time , place ,person.
- Patient is thin built and ill nourished.
- No pallor, icterus, cyanosis, clubbing, pedal odema , lymphadenopathy
- Skin tightness over the face, upper limbs proximal to elbow, thighs ,legs.
- Perioral furrowing
- Hyperpigmented and hypopigmented patches over both elbow flexures - suggestive of Salt and Pepper appearance.
- Digital pits were seen on left index finger







Vitals

- PR : 74 beats per minute.
- BP : 110/70mmHg .
- RR : 19 cycles per minute.
- SPO2 : 96% at room air
- Temp : Afebrile.

On Auscultation

- B/L AE present
- Normal vesicular breath sounds
- B/L fine late inspiratory crepitations in mammary, infraaxillary,infrascapular and interscapular areas

Investigations

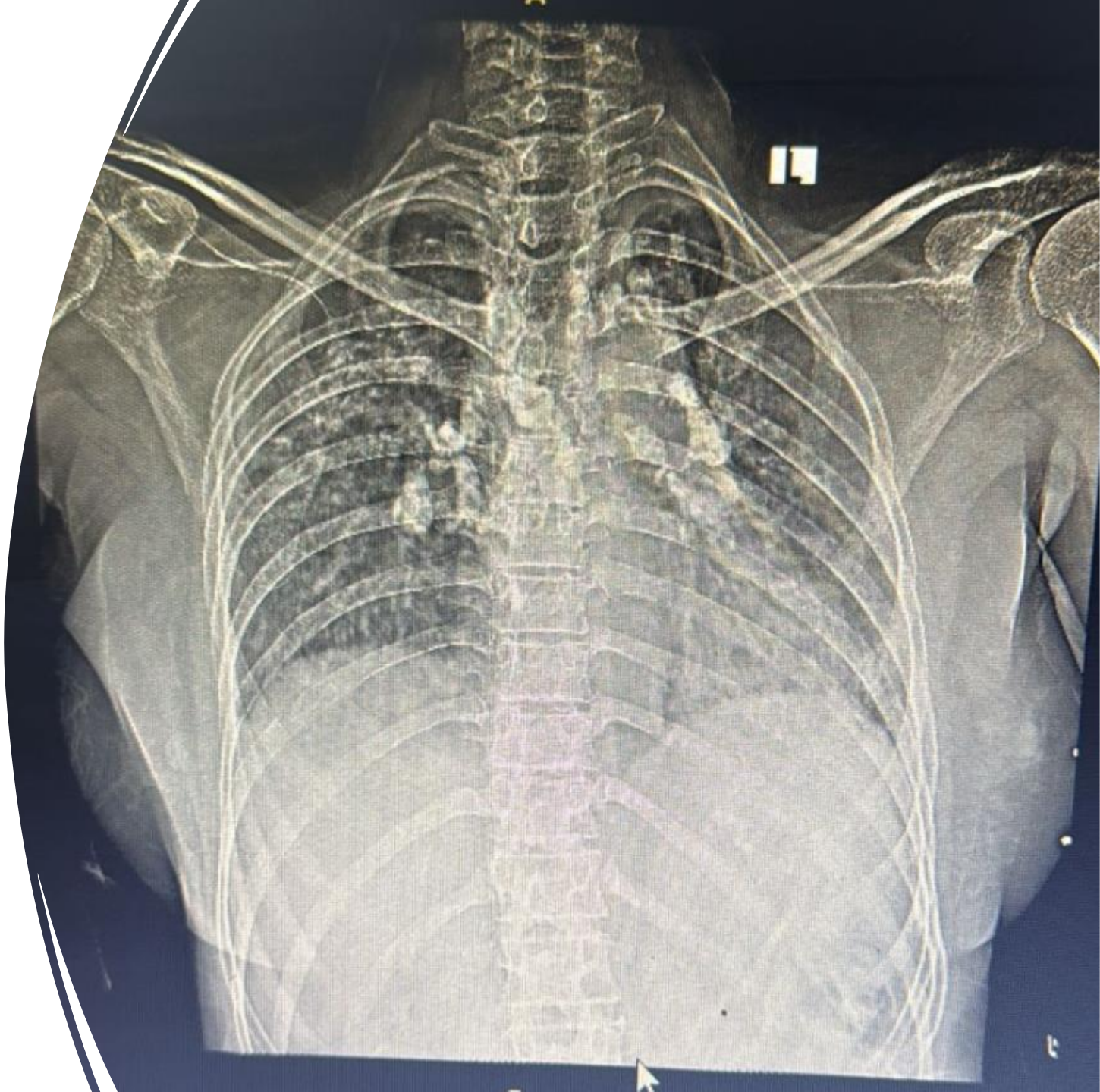
- **CBP:**
  - Hb-11.5gm/dl
  - Total count-12,400 cells/m3
  - RBC count-4.72 million/m3
  - Platelets-3.3lakh/m3
  - Differential count N-90% E-01% L-07% M-02%
  - LFT,RFT are with in normal limits.

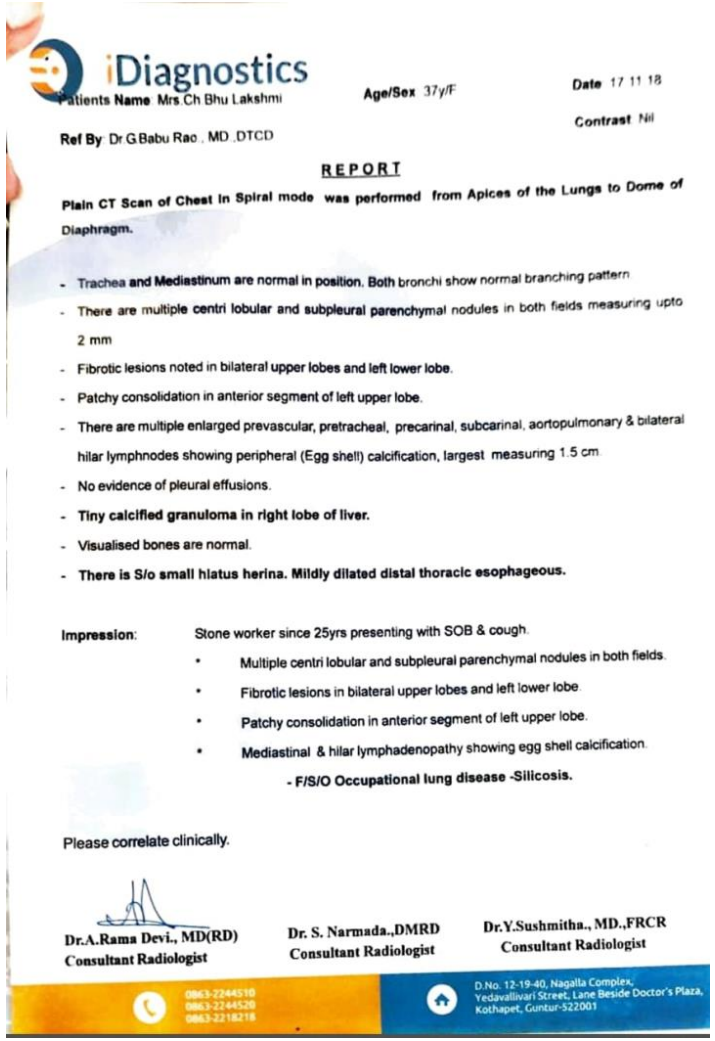
Investigations

- VIRALS:NON REACTIVE
- ECG:NORMAL
- 2D ECHO:
  - NO RWMA,Dilated RA,RV
  - MODERATE TR,SEVERE PAH
  - MILD MR, MILD RV DYSFUNCTION
  - NRML LV SYSTOLIC FUNCTION,
  - GRADE 1 DIASTOLIC DYSFUNCTION,
  - NO CLOTS,NO PE
- RF : NEGATIVE
- ANTI SCL 70 : STRONGLY POSITIVE

Radiological Investigations

CXR S/O BILATERAL HILAR CALCIFIED LYMPHNODES.





iDiagnostics

Patients Name Mrs.Ch Bhu Lakshmi Age/Sex 37y/F Date 17.11.18

Ref By Dr.G.Babu Rao., MD.,DTCD Contrast Nil

REPORT

Plain CT Scan of Chest in Spiral mode was performed from Apices of the Lungs to Dome of Diaphragm.

- Trachea and Mediastinum are normal in position. Both bronchi show normal branching pattern.
- There are multiple centri lobular and subpleural parenchymal nodules in both fields measuring upto 2 mm.
- Fibrotic lesions noted in bilateral upper lobes and left lower lobe.
- Patchy consolidation in anterior segment of left upper lobe.
- There are multiple enlarged prevascular, pretracheal, precarinal, subcarinal, aortopulmonary & bilateral hilar lymphnodes showing peripheral (Egg shell) calcification, largest measuring 1.5 cm.
- No evidence of pleural effusions.
- **Tiny calcified granuloma in right lobe of liver.**
- Visualised bones are normal.
- **There is S/o small hiatus herina. Mildly dilated distal thoracic esophageous.**

Impression: Stone worker since 25yrs presenting with SOB & cough.

- Multiple centri lobular and subpleural parenchymal nodules in both fields.
- Fibrotic lesions in bilateral upper lobes and left lower lobe.
- Patchy consolidation in anterior segment of left upper lobe.
- Mediastinal & hilar lymphadenopathy showing egg shell calcification.

- F/S/O Occupational lung disease -Silicosis.

Please correlate clinically.

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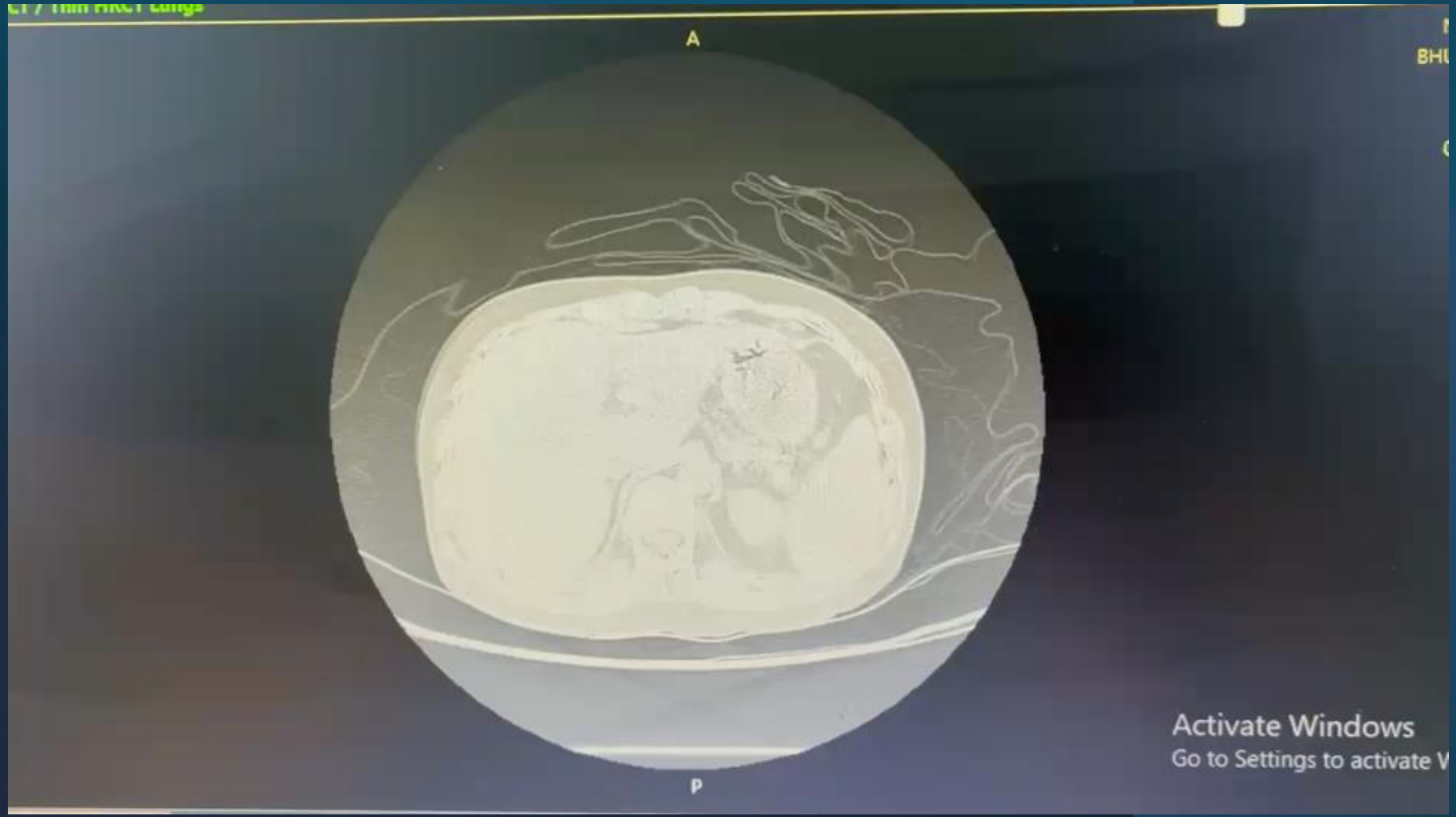
HRCT Chest

HRCT CHEST shows egg shell calcification of hilar and mediastinal lymphnodes s/o SILICOSIS.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- Few patchy areas of GGO in the anterior segment of bilateral upper lobes likely **infective etiology.**
- Multiple fibrocalcific nodules in the apical,posterior segment of right upper lobe ,apicoposterior segment of left upper lobe.Multiple calcified mediastinal lymphnodes suggestive of **post infective sequelae.**
- Multiple fibrotic strands with adjacent traction bronchiectactic changes on apical, posterior segments of right upper lobe, and apicoposterior segment of left upper lobe.
- Honeycombing ,fibrotic strands and adjacent bronchiectatic changes in right middle lobe, inferior lingular segment and bilateral lower lobes predominantly in basal segments with apicobasal gradient likely **ILD WITH UIP PATTERN.**

ERASMUS SYNDROME

- **DEFINITION**:

  - Erasmus syndrome is a rare occupational rheumatological disorder characterized by the development of systemic sclerosis following exposure to silica with or without associated silicosis.
  - Its incidence is 0.9 % among systemic sclerosis patients as per brazil study.

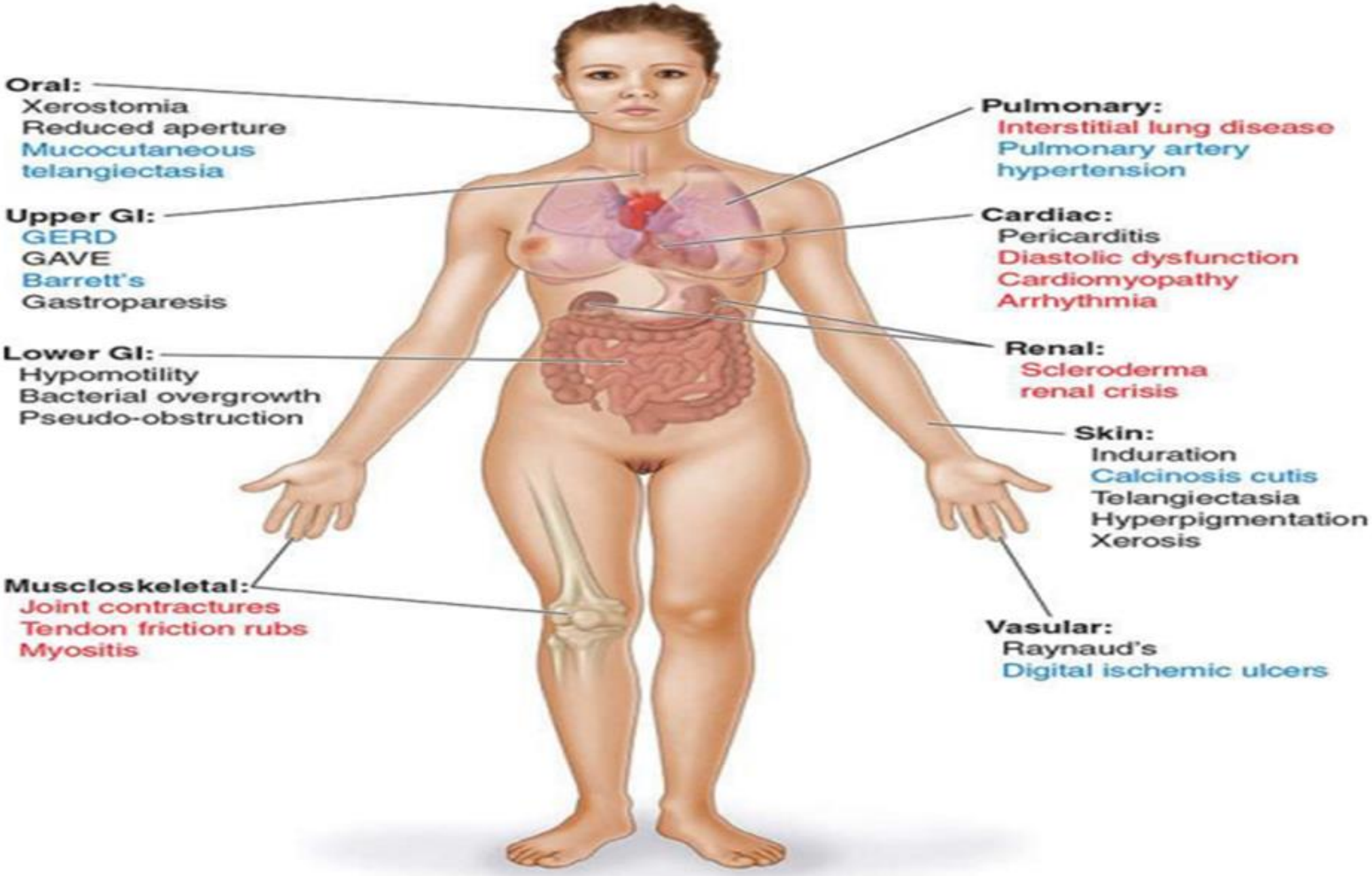
- Erasmus in 1957 demonstrated a notable occurrence of progressive systemic sclerosis (PSS) among gold miners exposed to dust containing a significant proportion of free silica
- In India, the initial instance of "Erasmus syndrome" (silicosis-induced systemic sclerosis) was documented by Khanna et al. in 1997.
- A meta-analysis conducted between 2012 and 2022 revealed that Erasmus syndrome most commonly affects men, with an average onset age of 51.7 years, following chronic silica exposure ranging from six to 47 years.

PATHOPHYSIOLOGY

- The exact pathophysiology involves the silica induced activation of macrophages,which release inflammatory cytokines such as interleukin -1,IL-2,and tumor necrosis factor -alpha.
- These cytokines stimulate T-helper cells and promote fibroblast activation resulting in excessive collagen deposition which is the hallmark of systemic sclerosis.
- Chronic silica exposure is associated with immune dysregulation which includes increase in soluble IL-2 receptor levels which further exacerbates the autoimmune response.

Systemic sclerosis

- Systemic sclerosis is a multisystem autoimmune disorder of idiopathic aetiology manifested vasculopathic changes, diffuse tissue fibrosis in multiple organs such as lung, heart, kidney gastrointestinal tract and skin.
- It is predominantly found in females.
- However, males have a more severe expression of disease and higher mortality.
- It may be caused by environmental and occupational exposures to vinyl chloride, organic solvent, and seldom by silica.


Oral:
Xerostomia
Reduced aperture
Mucocutaneous telangiectasia
Upper GI:
GERD
GAVE
Barrett's
Gastroparesis
Lower GI:
Hypomotility
Bacterial overgrowth
Pseudo-obstruction
Muscloskeletal:
Joint contractures
Tendon friction rubs
Myositis
Pulmonary:
Interstitial lung disease
Pulmonary artery hypertension
Cardiac:
Pericarditis
Diastolic dysfunction
Cardiomyopathy
Arrhythmia
Renal:
Scleroderma renal crisis
Skin:
Induration
Calcinosis cutis
Telangiectasia
Hyperpigmentation
Xerosis
Vasular:
Raynaud's
Digital ischemic ulcers

Classification criteria for diagnosis of systemic sclerosis

ITEM	SUBITEM	WEIGHT/SCORE
Skin thickening (bilateral); fingers extending proximal to MCP joints		9
Skin thickening of fingers only	Puffy fingers Sclerodactyly (skin thickened distal to MCP joints)	2 4
Fingertip lesions	Digital tip ulcer Fingertip pitting scars	2 3
Mucocutaneous telangiectasia		2
Abnormal nailfold capillary pattern		2
Lung involvement	PAH Interstitial lung disease	2 2
Raynaud's phenomenon		3
SSc-specific autoantibodies	ACA Scl-70 RNA polymerase III	3

Abbreviations: ACA, anticentromere antibodies; MCP, metacarpophalangeal joint; PAH, pulmonary arterial hypertension.

Features of LIMITED VS DIFFUSED Systemic sclerosis

CHARACTERISTIC FEATURE	LIMITED CUTANEOUS SSc	DIFFUSE CUTANEOUS SSc
Skin involvement	Indolent onset. Limited to fingers, distal to elbows, face; slow progression	Rapid onset. Diffuse: fingers, extremities, face, trunk; rapid progression
Raynaud's phenomenon	Antedates skin involvement, sometimes by years; may be associated with critical ischemia in the digits	Onset coincident with skin involvement; critical ischemia less common
Musculoskeletal	Mild arthralgia	Severe arthralgia, carpal tunnel syndrome, tendon friction rubs
Interstitial lung disease	Slowly progressive, generally mild	Frequent, early onset and progression, can be severe
Pulmonary arterial hypertension	Frequent, late, may occur as an isolated complication	Often occurs in association with interstitial lung disease
Scleroderma renal crisis	Very rare	Occurs in 15%; generally early (<4 years from disease onset)
Calcinosis cutis	Frequent, prominent	Less common, mild
Characteristic autoantibodies	Anti-centromere	Anti–topoisomerase I (Scl-70), anti-RNA polymerase III

Key Findings of Intrathoracic Disease in SSc

- (Fibrotic) ILD
  - *NSIP >> UIP*
  - *Peripheral/subpleural predominance*
  - *Lower zone predilection*
  - *Ground-glass opacification, traction bronchiectasis/bronchiolectasis, lower zone volume loss, architectural distortion, and/or honeycombing*
- Esophageal dilatation
- PA enlargement (↑PA:Ao diameter)

Management

Current guidelines for managing systemic sclerosis, including those associated with occupational exposure, emphasize a multidisciplinary approach. This includes symptomatic management, immunosuppressive therapy, and preventive measures such as occupational safety protocols.

Early intervention is crucial to mitigate complications, particularly pulmonary hypertension and ILD, which are major contributors to morbidity and mortality.

DISEASE-MODIFYING THERAPY IMMUNOSUPPRESSIVE AGENTS :

Cyclophosphamide

Methotrexate

Mycophenolate mofetil

Tocilizumab, a monoclonal antibody that blocks IL-6 receptor signaling, also showed benefit on both skin and lung involvement in randomized SSc trials

ANTIFIBROTIC THERAPY:

- Tyrosine kinase inhibitor like nintedanib, alone or in combination with mycophenolate, in patients with SSc-ILD, with significant slowing of the loss of lung function.

VASCULAR THERAPY:

- The goal of Raynaud's therapy is to control episodes, prevent and enhance the healing of ischemic complications, and slow the progression of obliterative vasculopathy.
- Calcium channel blockers – AMLODIPINE,NIFEDIPINE
- PDE 5 inhibitors – SILDENAFIL

SILICOSIS

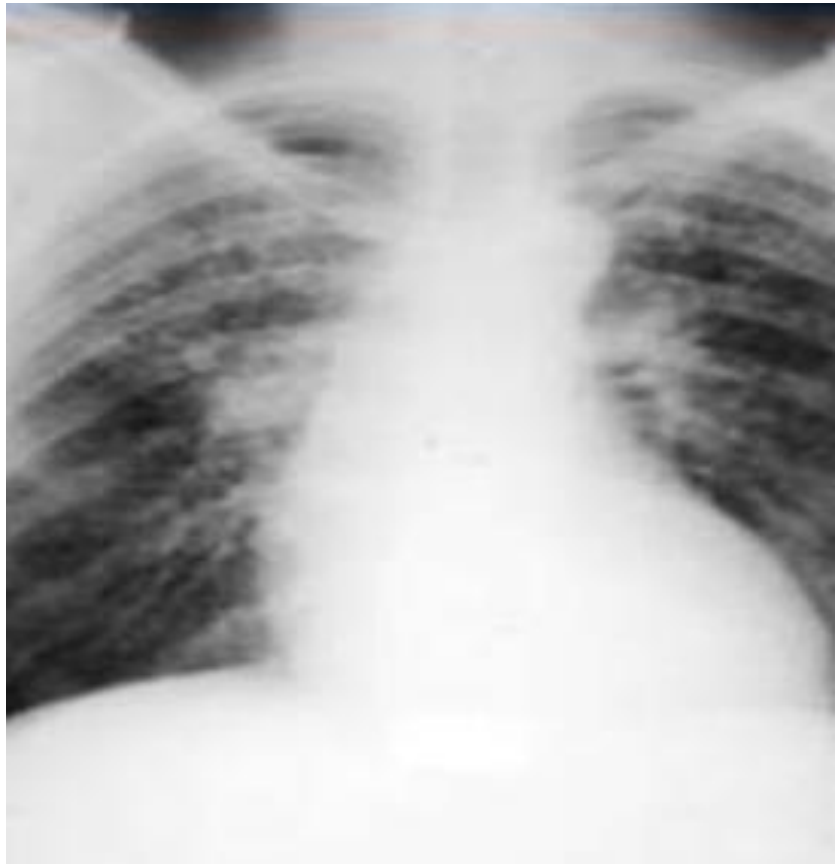
- Silicosis is a type of pneumoconiosis. It is an occupational disease characterised by irreversible lung fibrosis because of crystalline silica inhalation, retention, and pulmonary reaction among workers in mining, quarrying, masonry and sand blasting sectors.
- Silicosis is frequently linked with tuberculosis (silico-tuberculosis), lung carcinoma and occasionally related with systemic sclerosis (silica associated systemic sclerosis -SA-SS), systemic lupus erythematosus and rheumatoid arthritis.

Types of silicosis

- Chronic Or Classic
- Accelerated
- Acute

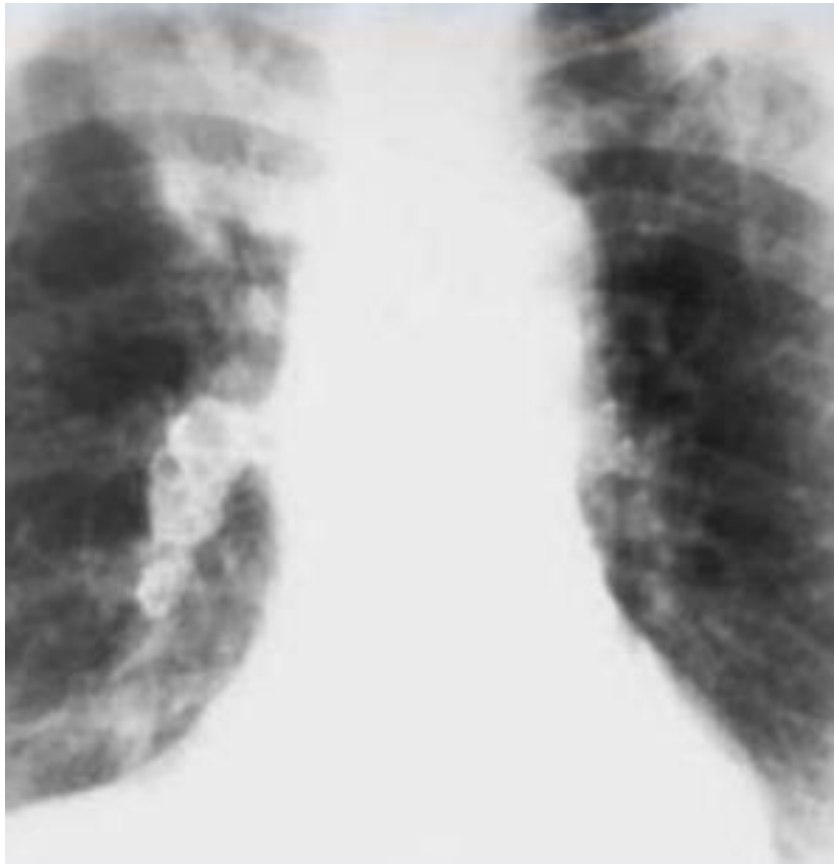
Chronic or classic silicosis:

- It is the most common form of SILICOSIS usually follows 15 or more Years of exposures to respirable dust containing quartz. in the later stages patient experience fatigue, Extreme Shortness of Breath, Chronic Cough , Chest pain or Respiratory Failure
- The pathologic hallmark in the lungs of patients with the chronic form is silicotic nodule



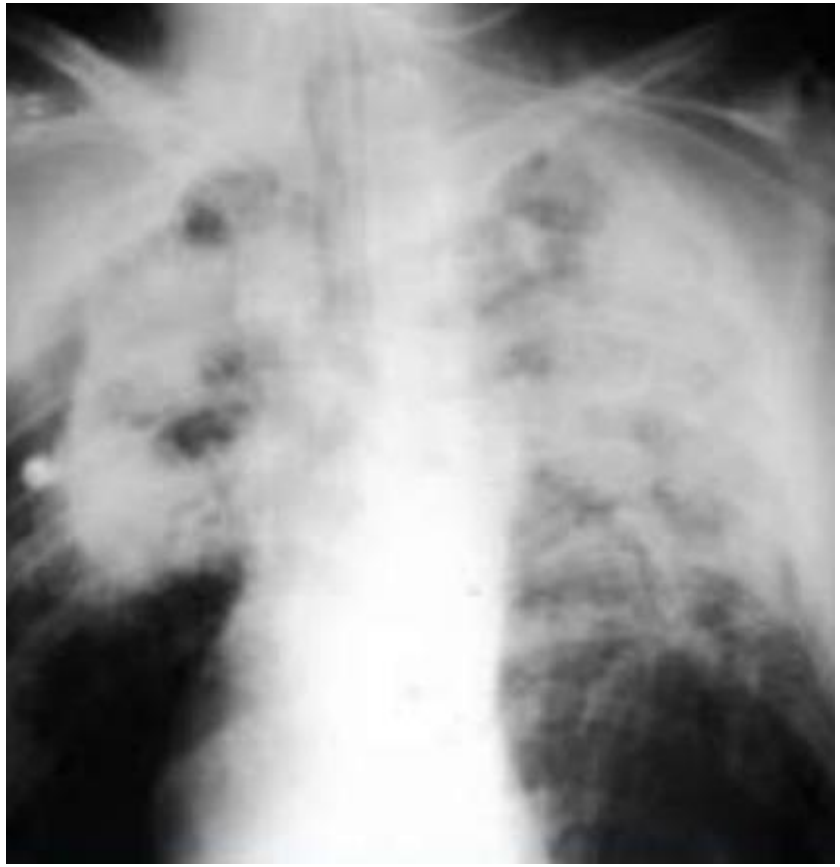
Accelerated silicosis.

- Accelerated silicosis can occur after 5-10 years following to high concentrations of the respirable crystalline silica. Symptoms include, severe breathing trouble, cough, generalized weakness and weight loss



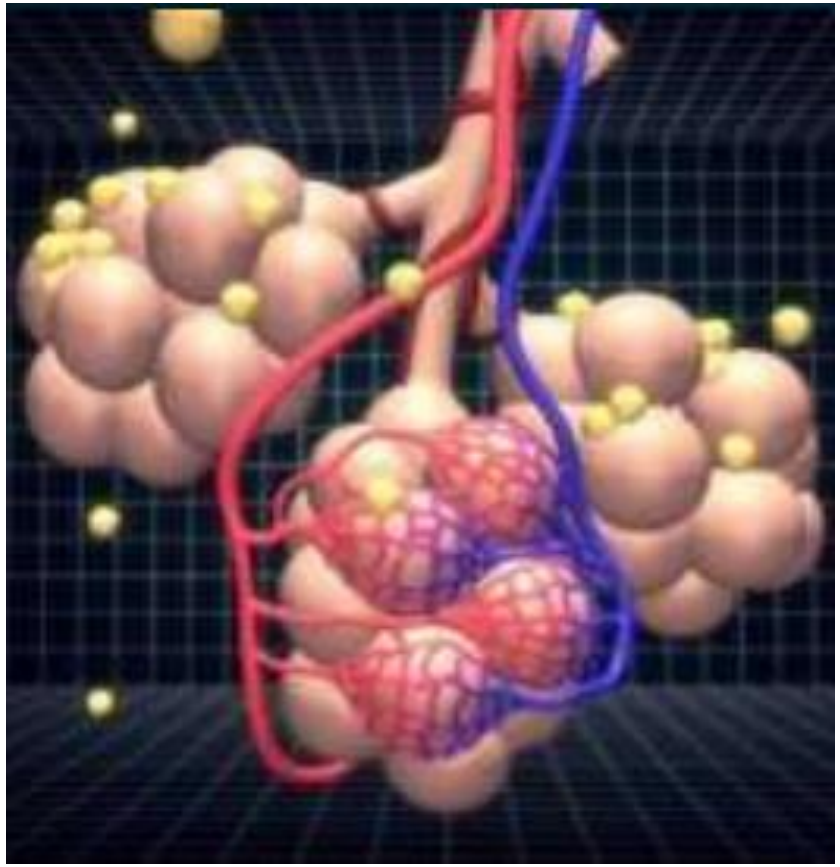
Acute silicosis.

- Acute silicosis develops within a few months upto about 5 years after massive inhalation of silica. symptoms include dramatic dyspnea, weakness and weight loss and this may lead to death



Pathophysiology

- silicosis is characterized by granulomatous and fibrotic lesions, occurring due to accumulation of respirable silica mineral particles.
- Crystalline silica dust causes fluid accumulation and scar tissue in the lungs and reduces the ability to breathe.
- When small silica particles are inhaled, they can embed themselves deeply into the tiny alveolar sacs and ducts in the lungs when oxygen and carbon dioxide are exchanged



Management

The control of silicosis in both the developed and developing world requires comprehensive prevention strategies, including exposure control, medical screening and surveillance, research, and education

Prophylactically ISONIAZID (INH) therapy is advised in silicotic patients with tuberculin positive skin test .

CONCLUSION

Erasmus syndrome is a sporadic progressive disease caused by inhalation of crystalline silica, where there is development of progressive systemic sclerosis, which leads to progressive dyspnoea and multiple symptoms. There might be associated interstitial lung disease or severe restrictive lung disease over silicotic lung. Sometimes, pulmonary hypertension could be developed which can complicate the morbidity of silicosis.

Thank you