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Original Research Paper

Radiology

INTERSTITIAL LUNG DISEASE IN YOUNG ADULTS – APPROACH AND WHAT NOT TO FORGET ?

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KEYWORDS :

CASE REPORT :

A 26 year old female patient presented on 19/01/2020 with complaints of Breathlessness on exertion since 2 years (with recent aggravation) , Cough with expectoration.

Past History – No history of DM/HTN or Tuberculosis
Family History – No similar complaints in the family
Personal History – Nothing significant

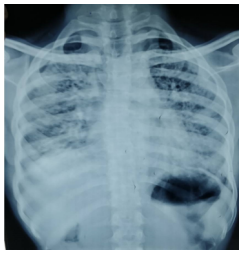
Systemic Examination

Respiratory System – BL BSE ; Creptitations in bilateral lower lung zone ; SpO2 – 77% on room air ; 90% on oxygen at 2L/min ; 2D ECHO – shows severe pulmonary artery hypertension ; Right atrium and right ventricle dilated ; NO LV RWMA ; No clots or vegetations ; LVEF = 56%

DATE	19-1-2020	20-1-2020	21-1-2020	24-1-2020	8-2-2020	10-2-2020
Total leucocyte count	19,580	20,800	16,600	21,500	16,500	17,000

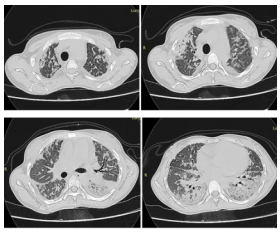
ESR is elevated (44/mm)
CRP is positive – 4.8mg/dl (Normal value = <0.6mg/dl)
Non-reactive for hepatitis A, B, C, E and HIV ; No AFB isolated in sputum sample

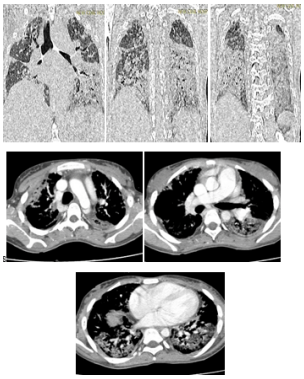
ADVICED: CXR and CT-THORAX



Chest radiograph showed bilateral dense reticulo-nodular opacities in bilateral lung fields , with multiple peripheral patches of soft tissue consolidative opacities in bilateral mid-lower zones.

P/o Interstitial lung disease with acute exacerbation , i/v/o clinical history;





CECT THORAX shows diffuse interlobular and intralobular septal thickening which show predominantly subpleural distribution with extensive GGOs forming dense patches of consolidation with tractional bronchiectasis and bronchiolectasis noted in bilateral lung parenchyma in an apicobasal gradient and anteroposterior gradient .

P/o Changes of interstitial lung disease (CT features consistent with non-IPF diagnosis)

Further work-up recommended to rule out underlying connective tissue disease Extensive GGOs seen in bilateral lower lobes, relatively sparing the anterobasal segment of bilateral lower lobes.Minimal effusion bilaterally with fissural extension into the oblique fissures

P/o changes of active infective etiology

Changes of pulmonary artery hypertension

APPROACH TO THE DIFFERENTIALS :

- GGOs with Inter- and intralobular septal thickening with apicobasal gradient ILD
- In keeping with the pulmonary hypertension, bilateral effusion and consolidation patches pulmonary edema ;

However NO signs of Volume overload , no pedal edema , no raised JVP.

- Bilateral effusion , with consolidation with leucocytosis , raised CRP Underlying inflammatory / infective pathology
- ILD in a 26 yrs old female Underlying connective tissue disease > Idiopathic
- Pulmonary artery hypertension in 26 yrs old Underlying connective tissue disease > Idiopathic

Follow up :

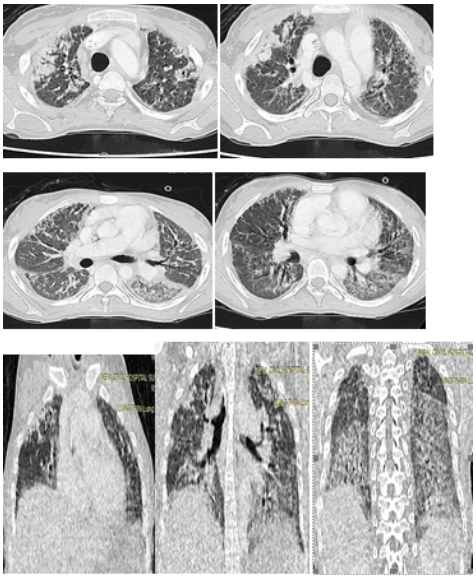
Subsequently done rheumatological reference , suggested work-up , which is as follows:

ANA – Negative ; cANCA/pANCA – Negative; no specific pattern

However other spectra which also shows ILD patterns : Anti-PR3, anti-MPO, Myositis antibody profile – Not done

Management with plasmapheresis , Immunosuppresants-Methylprednisolone and Antibiotics for active infections;

Followed up with CT after 2 weeks :



Changes of interstitial lung disease (CT features consistent with non-IPF diagnosis) remained the same in this scan as well . However as compared to previous CT , there is complete resolution of previous active infective etiology (Patches of consolidation significantly reduced , no effusion)

Further followed up with Lung biopsy , turned out of interstitial fibrosis.

DISCUSSION

The collagen vascular diseases that most commonly involve the lung are rheumatoid arthritis, progressive systemic sclerosis, systemic lupus erythematosus, polymyositis and dermatomyositis, mixed connective tissue disease, and Sjögren syndrome. The two thoracic manifestations with the greatest clinical importance in patients with collagen vascular diseases are interstitial lung disease and pulmonary arterial hypertension;[1]

Type of Collagen Vascular Disease	UIP	NSIP	COP	LIP	DAD	Hemorrhage	Airway Disease
Rheumatoid arthritis	+++	++	++	+	+	–	+++
Progressive systemic sclerosis	+	+++	+	–	+	–	–
Dermatomyositis/polymyositis	+	+++	+++	–	++	–	–
Sjögren syndrome	+	++	–	++	+	–	+
Mixed connective tissue disease	+	++	+	–	–	–	–
Systemic lupus erythematosus	+	++	+	+	++	+++	–

CTD-ILD VS IDIOPATHIC ILD – DOES IMAGING HELP?

Concentration of fibrosis within the anterior aspect of the upper lobes (with relative sparing of the other aspects of the upper lobes) and concomitant lower lobe involvement ("anterior upper lobe" sign) ; Exuberant honeycomb-like cyst formation within the lungs constituting greater than 70% of fibrotic portions of lung ("exuberant honeycombing" sign) ; Isolation of fibrosis to the lung bases with sharp demarcation in the craniocaudal plane without substantial extension along the lateral margins of the lungs on coronal images ("straight-edge" sign) [13]

All these signs were much more common in and highly specific for CTD UIP than for IPF UIP

THE MOST COMMON PATTERNS OF LUNG FIBROSIS
RA - UIP followed by NSIP [8,9]

SYSTEMIC SCLEROSIS - NSIP pattern ; UIP is much less common in systemic sclerosis than in rheumatoid arthritis. [10,11,12]

MYOSITIS often presents with a pattern of combined NSIP and organizing pneumonia ; Classically, the consolidation from organizing pneumonia rapidly resolves with corticosteroid therapy, leaving patients with CT patterns consistent with NSIP. This typical evolution of CT findings is highly suggestive of myositis Many patients with CTD may have no symptoms from a musculoskeletal standpoint, and the ILD may be the initial manifestation that leads to the eventual diagnosis [2-7]

Even in patients with a high-confidence UIP pattern, there are specific CT findings that should make one strongly consider a CTD-ILD diagnosis. In these patients, a more thorough clinical and serologic workup for underlying CTD may be necessary, especially in younger patients, black patients, and women

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