Original Research Article

Primary Thoracic Manifestations in Connective Tissue Diseases on High Resolution Computed Tomography: A Prospective Study

Ritu Dhawan Galhotra¹, Gurkirandeep Singh², Karan Saggar³, Kavita Saggar⁴, Kamini Gupta⁵

¹Associate Proffesor, Department of Radiodiagnosis, DMCH, Ludhiana, Punjab, ²PG resident, DMCH, Ludhiana, Punjab, ³BDS, Private Practitioner, Ludhiana, Punjab, ⁴Professor and Head, Department of Radiodiagnosis, DMCH, Ludhiana, Punjab, ⁵Associate Professor, Department of Radiodiagnosis, DMCH, Ludhiana, Punjab, India

Corresponding author: Dr Gurkirandeep Singh, PG Resident, Department of Radiodiagnosis, Dayanand Medical College and Hospital, Ludhiana, India

How to cite this article: Ritu Dhawan Galhotra, Gurkirandeep Singh, Karan Saggar, Kavita Saggar, Kamini Gupta. Primary thoracic manifestations in connective tissue diseases on high resolution computed tomography: a prospective study. International Journal of Contemporary Medicine Surgery and Radiology. 2018;3(1):28-35.

ABSTRACT

Introduction: The aim of this paper was to evaluate the thoracic manifestations associated with the Connective tissue disorders, with an emphasis on interstitial and airway disease pattern on the High Resolution computed tomography (HRCT) findings.

Material and methods: The present study was conducted for a period of one year. A total of 70 patients with various connective tissue disorders having respiratory complaints were evaluated.

Results: During the study period 70 patients (58 females and 12 males) underwent evaluation. Cough and dyspnea were the most common presenting symptoms. Variety of thoracic abnormalities were in detected in 67 (95%) cases. Most common abnormality detected on HRCT was interstitial fibrosis/interstitial lung disease present in 42(60%) cases. Most common parenchymal abnormalities seen were reticulations (61.4%), ground glass opacification (40%), mosaic attenuation (32.8%) and honeycombing (24.3%). Airway abnormalities seen were bronchiectasis (48.5%), emphysema (12.8%), and ground glass nodules (2.8%).

Conclusion Interstitial lung disease is the most common pulmonary manifestation among patients with connective tissue disorders, and early detection and prompt treatment is expected to improve the outcome.

Keywords: Connective Tissue Disorders, Thoracic Manifestations, Interstitial Lung Disease

INTRODUCTION

Connective tissue disorders (CTDs) or collagen disease are a heterogeneous group of systemic inflammatorydiseases of autoimmune origin that affect a wide range oforgans and systems. These include rheumatoid arthritis, systemic lupus erythematous, systemic sclerosis, sjogren's syndrome, polymyositis/dermatomyositis, mixed connective tissue disorders. These are often associated with a wide range of lung conditions The frequency and wayin which the respiratory system is affected depend on each type of collagen disease with a specific entity.¹⁻³ The most common lung condition in every case is pneumonitis⁴ which appears in all the histopathological patterns of idiopathic interstitial pneumonitis (IIP).⁵ Since these patients are immunocompromised, infections are one of the most common causes of respiratory disease.⁶ Lastly, drug-related adverse reactions should also be included in the differential diagnosis.

Therefore, the diagnosis and clinical management of these patients is complex and should be based on the combination of different diagnostic approaches: clinical symptoms, laboratory and imaging findings. Sensitivity of chest radiography for the detection of interstitial lung disease is very low. Conversely, HRCT provides good sensitivity and specificity and is able to provide a confident diagnosis of interstitial lung disease and fibrosis. In addition, It allows multiplanar reformatted images and has the advantage of being an affordable and non-invasive technique.⁷⁻⁸ The aim of this paper is to evaluate the pulmonary manifestations associated with the CTDs on the high-resolution computed tomography (HRCT), which facilitate early diagnosis of these conditions.

MATERIAL AND METHODS

The present study was conducted in the department of radiodiagnosis of the Dayanand Medical College and Hospital, Ludhiana and included assessment of Primary thoracic manifestations in connective tissue diseases on high resolution computed tomography (CT). All patients with connective tissue disorders presenting with respiratory symptoms over a period of one year were included in the study. Consent was obtained from each patient. HRCT or CECT chest (on Somatom definition AS+128 slice CT machine by Siemens Germany Ltd.) was done. All the results were recorded on Microsoft excel sheet. Analysis was done by using SPSS software. Statistical analysis was done using tests of significance.

RESULTS

Study was done for a period of one year to evaluate the

	Total no. of patients	Percentage				
RA	39	55.7				
SLE	8	11.1				
Systemic sclerosis	8	11.1				
Sjogrens	5	7.4				
Polymyositis	3	4.3				
MCTD	3	4.3				
Others	4	5.7				
Total	70					
Table-1: Distribution of cases for the different disease entities						

thoracic imaging findings in patients with various CTDs (Rheumatoid arthritis (RA), Systemic lupus erythematous (SLE), Systemic sclerosis, Sjogrens syndrome, Polymyositis, Mixed connective tissue disorder (MCTD)). Total of 70 patients were evaluated.

Out of total 70 patients Thirty nine cases were of RA, eight cases each of SLE and systemic sclerosis, five cases were of sjogrens, fourcases were of polymyositis and three cases were of MCTD. Four cases were autoantibody positive and were placed in others category(table1)

Most commonly affected age group was mostly between 51-70 years with mean age of presentation being 56.8+/-13.5 years. Mean age of presentation in RA was 60.6, in SLEwas 49.7, in systemic sclerosis was 47, in sjogrens was 57.6, in

	Connective tissue	RA	SLE	Systemic sclerosis	Sjogrens	Polymyositis	MCTD	Others	Total
	disorder associated								
Age group	18-30	1	1	2	0	0	0	0	4
	31-40	0	1	1	0	0	1	1	4
	41-50	6	2	2	1	0	0	1	12
	51-60	11	2	1	2	2	0	0	18
	61-70	17	1	1	2	1	2	1	25
	More than 70	4	1	1	0	0	0	1	7
Total		39	8	8	5	3	3	4	70
			Table-2	Distribution of age o	f patients				

	Connective tissue disorder associated								
	RA	SLE	Sjogren	SYS Sclerosis	Polymyositis	MCTD	Others		
	(n=39)	(n=8)	(n=5)	(n=8)	(n=3)	(n=3)	(n=4)		
Cough	30	6	1	6	1	2	2	48	
	76.90%	75.00%	20.00%	75.00%	33.30%	66.70%	50.00%	68.57%	
Dyspnea	35	8	5	8	3	3	3	65	
	89.70%	100.00%	100.00%	100.00%	100.00%	100.00%	75.00%	92.86%	
Hemoptysis	1	1	0	0	0	0	0	2	
	2.60%	12.50%	0.00%	0.00%	0.00%	0.00%	0.00%	2.86%	
Fever	21	7	1	3	1	2	4	39	
	53.80%	87.50%	20.00%	37.50%	33.30%	66.70%	100.00%	55.71%	
			Table-3: Clin	ical symptoms di	stribution				

	Connective tissue disorder associated							Total
	RA (n=39)	SLE (n=8)	Sjogren (n=5)	SYS sclerosis (n=8)	Polymyositis (n=3)	MCTD (n=3)	Others (n=4)	
Reticular	26	2	2	7	3	1	2	43
	66.70%	25.00%	40.00%	87.50%	100.00%	33.30%	50.00%	61.43%
Honeycombing	12	0	0	4	0	0	1	17
	30.80%	0.00%	0.00%	50.00%	0.00%	0.00%	25.00%	24.29%
Ground glass opacity (GGO)	17	4	0	1	3	0	3	28
	43.60%	50.00%	0.00%	12.50%	100.00%	0.00%	75.00%	40.00%
Mosaic attenuation	14	2	3	2	1	1	0	23
	35.90%	25.00%	60.00%	25.00%	33.30%	33.30%	0.00%	32.86%
Nodular	8	2	1	2	0	1	1	15
	20.50%	25.00%	20.00%	25.00%	0.00%	33.30%	25.00%	21.43%
Consolidation	7	2	0	0	0	1	0	10
	17.90%	25.00%	0.00%	0.00%	0.00%	33.30%	0.00%	14.29%
Cysts	6	0	1	1	0	0	1	9
	15.40%	0.00%	20.00%	12.50%	0.00%	0.00%	25.00%	12.86%
	Table	-4: Parench	ymal abnor	malities on HRC	in various CTDs			

International Journal of Contemporary Medicine Surgery and Radiology

29

polymyositis was 55.3, and in MCTD was 52 years. Mean age of presentation in females was 55.9 years and in males was 60.75 years.(table2)

In our study 82 percent of the cases were females indicating the female predilection of connective tissue disorders. Most common presenting symptom was dyspnea. Cough

Airway abnormality	Connective tissue disorder associated								
	RA (n=39)	SLE (n=8)	Sjogren (n=5)	SYS Sclerosis (n=8)	Polymyositis (n=3)	MCTD (n=3)	Others (n=4)	Total	
BE	26	0	2	3	0	2	1	34	
	66.7%	0.0%	40.0%	37.5%	0.0%	66.7%	25.0%	48.57%	
Emphysema	7	1	0	0	0	0	1	9	
	17.9%	12.5%	0.0%	0.0%	0.0%	0.0%	25.0%	12.86%	
Ground glass nodules (GGN)	2	0	0	0	0	0	0	2	
	5.1%	0.0%	0.0%	0.0%	0.0%	0.0%	0.0%	2.86%	
Table-5: Airway abnormalities on HRCT in various CTDs									

Lobes affected	Right upper lobe	Right middle lobe	Right lower lobe	Left upper lobe	Left lower lobe	Total				
	(ROL)	(RIVIL)	(RLL)	(LOL)	(LLL)					
RA	23	24	32	19	33	39				
	59.0%	61.5%	82.1%	48.7%	84.6%					
SLE	2	3	4	3	4	8				
	25.0%	37.5%	50.0%	37.5%	50.0%					
Sjogren	4	4	4	3	3	5				
	80.0%	80.0%	80.0%	60.0%	60.0%					
SYS sclerosis	6	6	6	7	6	8				
	75.0%	75.0%	75.0%	87.5%	75.0%					
Polymyositis	2	3	2	2	2	3				
	66.7%	100.0%	66.7%	66.7%	66.7%					
MCTD	3	3	3	3	2	3				
	100.0%	100.0%	100.0%	100.0%	66.7%					
Others	3	3	3	3	4	4				
	75.0%	75.0%	75.0%	75.0%	100.0%					
Total	43	46	54	40	54	70				
	61.4%	65.7%	77.1%	57.1%	77.1%					
	Table-6: Affection of different lung zones in different diseases									

	Connective tissue disorder associated									
Distribution	RA	SLE(n=8)	Sjogren	SYS sclerosis	Polymyositis	MCTD	Others			
	(n=39)		(n=5)	(n=8)	(n=3)	(n=3)	(n=4)			
Apicobasal gradient	22	0	0	4	0	0	0			
	56.4%	0.0%	0.0%	50.0%	0.0%	0.0%	0.0%			
Subpleural sparing	4	2	1	0	1	0	0			
	10.3%	25.0%	20.0%	0.0%	33.3%	0.0%	0.0%			
Subpleural/Peripheral	10	0	0	4	1	0	1			
	25.6%	0.0%	0.0%	50.0%	33.3%	0.0%	25.0%			
Subpleural/PB	2	0	0	1	0	1	0			
	5.1%	0.0%	0.0%	12.5%	0.0%	33.3%	0.0%			
	Table-7. Distribution of abnormalities in lung in various CTDs									

Connective tissue disorder associated	М	F	Total			
RA(39)	5	18	23			
SLE(8)	0	2	2			
Sjogren(5)	1	2	3			
SYS sclerosis(8)	0	6	6			
Polymyositis(3)	1	2	3			
MCTD(3)	0	2	2			
Others(4)	0	3	3			
Total(70)	7	35	42			
Table-8: Number of patients having CTD related Interstitial lung disease (ILD)						

Connective tissue disorder associated	Usual interstitial pneumonia (UIP)	Nonspecific interstitial pneumonia (NSIP)	Cryptogenic organizing pneumonia (COP)	Diffuse alveolar damage (DAD)	Lymphoid interstitial pneumonia (LIP)				
RA	11	11	1	0	0				
	28.2%	28.2%	2.6%	0.0%	0.0%				
SLE	0	1	0	1	0				
	0.0%	12.5%	0.0%	12.5%	0.0%				
Sjogren	0	1	0	0	1				
	0.0%	20.0%	0.0%	0.0%	20.0%				
SYS Sclerosis	4	0	1	0	0				
	50.0%	0.0%	12.5%	0.0%	0.0%				
Polymyositis	0	2	0	1	0				
	0.0%	66.7%	0.0%	33.3%	0.0%				
MCTD	0	0	1	0	0				
	0.0%	0.0%	33.3%	0.0%	0.0%				
Others	1	0	1	0	1				
	25.0%	0.0%	25.0%	0.0%	25.0%				
Total	16	15	4	2	2				
	22.9%	21.4%	5.7%	2.9%	2.9%				
Table-9: Type of ILD diagnosed in various CTDs									



Figure-1: Parenchymal abnormalities on HRCT in various CTDs

and fever was the second most common symptom. Few patients of RA and SLE also presented with hemoptysis. (table3)

Connective tissue disorders affect the different compartments of lung in characteristic pattern. Various radiological patterns were noted affecting the lung parenchyma, airway and the pleura (Table-4, Figure-1).

Parenchymal abnormalities

Rheumatoid arthritis

Reticulations were the most common parenchymal abnormality in RA accounting for 68 percent of cases followed by GGO which was present in over 44 percent of cases. Mosaic attenuation was present in 36 percent of cases. Honeycombing was present in 31 percent of cases. Nodules, consolidation, cysts were present in 20.5, 17.9, 15.4 percent of cases respectively.

SLE

GGO was the most common parenchymal abnormality present in SLE accounting for 50 percent of cases. Reticulations, mosaic attenuation, nodules and consolidation

were present in 25 percent of cases each. Honeycombing and cyst formation were not seen.

Sjogren's syndrome

Mosaic attenuation was most common finding in sjogrens presenting in 60 percent of cases. Reticulations were present in 40 percent of cases. Nodules and cysts were present in 20 percent of cases each.

Systemic sclerosis

Reticulations (87.5%) were most common finding in systemic sclerosis. Honeycombing was present in 50 percent of cases. Mosaic attenuation and nodules were found in 25 percent of cases each. GGO and cysts were found in 12.5 percent of cases each.

Polymyositis

Reticulations and GGO were present in all the cases with mosaic attenuation seen in one of the case.

MCTD

Out of the three cases reticulations and mosaic attenuation was found in one of the case. One of the case had consolidation



Figure-1: Axial and coronal HRCT sections showing evidence of reticulations, Honeycombing and few areas of ground glass opacification involving posterior segments of Bilateral lower lobes suggestive of UIP pattern



Figure 2: Axial HRCT sections showing Extensive reticulations and ground glass opacification of bilateral lungs is seen with evidence of subpleural sparing suggestive of NSIP pattern.



Figure-3: Axial HRCT sections (mediastinal window) showing evidence of bilateral pleural effusion (R>L)



Figure-4: Axial HRCT sections

and the other had nodules.

Airway abnormality

RA

Bronchiectasis was most common abnormality and was found in almost 67 percent of cases. Emphysema was found in 18 percent and GGN in 5 percent of cases.

SLE

Emphysema was present in one of the case. No other airway abnormality was detected in patients of SLE.

Sjogrens

Bronchiectasis was seen in two cases. No other airway abnormality was detected.



Figure-5: Axial and coronal HRCT sections showing thin walled cysts (blue arrow), reticulations in the form of interlobular septal thickening(red arrow and bronchiectasis(green arrow)



Figure-6: Axial HRCT sections showing Reticulations and microcystic honeycombingis seen in bilateral lower lobes suggestive of UIP pattern



Figure-7: Axial HRCT sections showing reticulations(black arrow) and honeycombing(red arrow)

Systemic sclerosis

Bronchiectasis was found in three cases. No other airway abnormality was detected.

Polymyositis

No airway abnormality was detected.

MCTD

Bronchiectasis was present in two cases.

Others

One case was found to have bronchiectasis and one was found to have emphysema

Other abnormalities in various CTDs

Pulmonary hypertension detected in two cases of sjogrens, one case each of SLE, systemic sclerosis, polymyositis and others and in Four cases of RA

Pericardial effusion was seen in two cases of SLE and one case each of sjogrens, systemic sclerosis and MCTD.

Pleural effusion was present mostly in patients with SLE (50%). One patient of sjogren syndrome (SSJ) also presented with pleural effusion.

LAP was found in nine cases RA, two cases of systemic sclerosis and one case of sjogrens

Table 6 shows the affection of different zones of lung in

CTDs. It is evident that lower lobe predilection was seen in RA and SLE.

Apico-basal gradient was seen in 22 cases (56.4%) of RA and four cases (50%) of systemic sclerosis. Subpleural sparing was seen in four cases (10.3%) of RA, two cases of SLE, and twocase each of sjogrens and polymyositis. Peripheral/subpleural distribution was seen in 10 cases (25.6%) of RA, four cases (50%) of systemic sclerosis, one case of polymyositis and one case in others category. Peribronchovascular and subpleural distribution was found in two cases of RA, one case of systemic sclerosis and one case of MCTD (table-7).

ILD was diagnosed in 23 cases of RA out of a total of 39, two cases of SLE (8 three cases of sjogren (5), six cases of systemic sclerosis (8), all cases of polymyositis, two cases of MCTD (3) and three in other autoantibody positive cases polymyositis, two cases of MCTD (3) and three in other autoantibody positive cases of polymyositis, two cases of MCTD (3) and three in other autoantibody positive cases. (table-8)

In rheumatoid arthritis UIP pattern was found in 11(28.2%) cases same as that NSIP. One case was diagnosed as COP. In SLE one case was diagnosed as NSIP and one case as DAD. In sjogren's syndrome one case was diagnosed as NSIP and one case as LIPIn systemic sclerosis four cases were diagnosed as UIP and one case as COP. In polymyositis two cases were of NSIP and one case of DAD. One case of MCTD was diagnosed as COP. Out of the four autoantibody positive cases one case was diagnosed as UIP, one as COP and one as LIP (Table 9).

DISCUSSION

Involvement of the respiratory system is common in the collagen vascular diseases and results in significant morbidity and mortality. Lung injury from collagen vascular disease can affect each portion of the lung, commonly; more than one compartment. Although there is some overlap, each collagen vascular disease is associated with a characteristic pattern of pulmonary involvement. The lung disease in these cases may precede the clinical presentation of the collagen disease, sometimes by more than five years.

High-resolution computed tomography (HRCT) is the method of choice for assessment of pulmonary abnormalities in collagen vascular diseases, offering the best correlation with histological findings, disease severity, prognosis, evaluation of disease progression, and differential diagnosis. It plays an important role in early detection and characterization of interstitial lung disease. However, it has some limitations. In many cases, HRCT appearance is nonspecific and may or may not be related to an underlying CTD. Thus, radiologic findings should never be interpreted without knowledge of the clinical picture.

In this study, .Out of the 70 patients, 67(95%) showed a variety of parenchymal, pleural, and mediastinal abnormalities on HRCT. Three patients had a normal study. This is consistent with Webb (2001)⁹ who reported sensitivity level of 94% with MDCT.

Majority of the patients in our study (55%) had RA while SLE and systemic sclerosis contributed to 11% of the cases.

consistent with Gaude et al $(2009)^{10}$ having RA(61%) and SLE(16.4%) and Vermaet al $(2013)^{11}$ having RA(48.6%) and SLE(21.2%).

Majority of the patients in our study were females(82%) similar to Vermaet al (2013)¹¹ with females(71.9%)and Gaude et al (2009)¹⁰ with females (64%)

Mean age of presentation in RA was 60.6, in SLE was 49.7, in systemic sclerosis was 47, in sjogrens was 57.6, in polymyositis was 55.3, and in MCTD was 52 years. Similar results were noted by Vermaet al $(2013)^{11}$ who found majority of patients to be in fifth decade of life. Gaude et al $(2009)^{10}$ reported 85% of their patients with connective tissue disorders having age more than 40 years.The mean age of presentation as noted by Bilgici et al $(2004)^{12}$ was 53.6 years and by Perez et al $(1997)^{13}$ was 57.8 years.

Cough and dyspnea were the commonestrespiratory symptoms with Similar observation was made by both Gaude et al (2009)¹⁰ and Vermaet al (2013).¹¹

Various abnormalities on HRCT affecting the parenchyma (reticulations, GGO, honeycombing, nodules, cysts), airway (bronchiectasis, GGN, emphysema), pleura (effusion) and mediastinum (PAH, pericardial effusion, lymphadenopathy (LAP)) were evaluated.

Reticulations in the form of inter and intralobular septal thickening were found in almost 67% of cases of RAsame seen by J.Biederer et al (2003)¹⁴ (75%) and Bilgici et al¹² (65%). GGO was noted in 43% of cases consistent with J.Biedere et al (2003)¹⁴ (37.5%). Honeycombing which is defined as thin walled cysts sharing walls arranged in layers in subpleural location was found in 30.8% of cases of RA similar to verma et al 2013 (31%).¹¹ Lynch et al (2009)² found honeycombing in 10% of cases.

Mosaic attenuation with areas of air trapping on expiratory film was found in 35.9% of cases. Similar to Perez et al 1998¹³ (32%) and Cortet et al 1997¹⁵ (25%). Nodules were seen 20.5% of cases in RA.consistent with B.Crestani 2005¹⁶ (20%). Similarly Cortet et al 1997¹⁵ reported 28% of parenchymal nodules in RA. Consolidation was noted in 18% cases of RA.

Bronchiectasis was seen in 66.7% of cases in RA which was much higher than the study done by Rockall et al., (2001)¹⁷ and Cortet et al 1997¹⁵ who reported bronchiectasis in 30% of cases. The colonization of these bronchiectases by different microorganisms is the cause of repeated respiratory infections, which is very important to take into account in these patients who receive immunosuppressive treatment for their underlying disease.

Emphysema in RA patients was seen in the current study in 17.9% of cases. Similar to Zrour et al 2005¹⁸ (13.3%) and Tanaka et al., (2004).¹⁹ The distribution of abnormalities was found predominantly in peripheral location with lower lobes predominance .

The prevalence of PAH in RA can only be grossly estimated by echocardiographic data, and according to several studies, it ranges from 0.8% to 21-27.5.²⁰⁻²¹ In our study PAH was found in 10.3% cases of RA. Mediastinal lymphadenopathy was noted in 23% cases.

Out of total 39 cases of RA, 23 (59%) cases were found to have CTD related ILD. Both UIP (Fig 1) and NSIP (Fig 2)

pattern was observed in 28.2% of cases with COP in 2.6% of cases. Similar to verma et al 2013¹¹ and Ysamatmarfa et al 2013.²²

The most common pulmonary manifestation of SLE is unilateral or bilateral pleural effusion (Fig 3) frequently associated with pericardial effusion.²³⁻²⁴

In this study pleural effusion associated with pleural thickening was found in 50% of cases same as Fekih et al. $(2011)^{25}$ with pleural abnormalities seen in 50% cases.

Sepal thickening in the form of reticulations was seen in 25% cases. Kakati et al., $(2007)^{26}$ reported a higher value of 39.4% and Gaude et al $(2009)^{10}$ reported a lower value of 12.5%. No evidence of bronchiectasis in our SLE patient, the same results as Gaude et al., $(2009)^{.10}$

Ground-glass opacification in SLE patients was seen in the current study in 50% of cases which was much higher than that of Kakati et al., (2007)²⁶ who reported GGO in 26.3% of their cases. Diffuse alveolar hemorrhage is a rare and severe manifestation, with a prevalence of 2.0%–5.4% among patients with systemic lupus erythematosus.²⁷ The most common radiologic imaging features are bilateral areas of consolidation, ground-glass opacities, and septal thickening. One patient in our study had the feature favouring DAD (Fig 4). Areas of air space consolidation was noted in 25% cases same as Hend M Maghrapyet al (2013).²⁸

Pulmonary artery hypertension (PAH) is uncommon in SLE; it was seen in this study in 12.5%, relatively nearer to Fekih et al., (2011).²⁵ The causal relationship between SLE and PAH may be due to multiple small vessel inflammation and/or vasculitis as well as sustained vasoconstriction, in situ thrombosis, and/or thromboembolism and interstitial pulmonary fibrosis (Arnaud 2011).²⁹

ILD and PAH are the most common cardiopulmonary findings in systeic sclerosis.³⁰⁻³¹ ILD in systemic sclerosis typically manifests on as predominantly GGO with an admixture of pulmonary fibrosis consistent with the NSIP pattern. Honeycomb cystic changes are reported in 11% to 37% of patients . This is unlike other patients with NSIP, who have little or no cystic change,^{32, 33, 34} Honeycomb cystic change is typically a marker for usual interstitial pneumonia (UIP) and pulmonary fibrosis.³⁵ In our study reticulations, honeycombing, and GGO was present in 87.5%, 50% and 12.5% of cases respectively. Bronchiectasis was seen in 37.5% of cases highlighting the hallmark features of parenchymal fibrosis and ILD (fig-6).

Sjogren's syndrome is characterized by T lymphocyte infiltration of various organs, most often the lacrimal and salivary glands and respiratory tract.³⁶ NSIP is the most common subtype of ILD in SS patients, with a prevalence ranging between 28% and 61%.³⁷ Historically, the predominant form of ILD in primary SSj was deemed to be lymphocytic interstitial pneumonitis (LIP). In a case series by Parambilet al³⁸ 17% of primary SSj patients with ILD had a diagnosis of LIP. In our study ILD identified was NSIP and LIP pattern (20%) (fig-5). Common findings at high-resolution chest CT include ground-glass opacities, nodules, thickening of interlobular septa, and cysts. Consolidation and honeycomb cysts are less frequently encountered. In our study reticulations was seen in 40%, mosaic attenuation in

60%, nodules and cysts in 20% of cases.

Interstitial lung disease associated with polymyositis and dermatomyositis has a wide spectrum of histopathologic features, on the basis of which four major disease patterns can be identified: cryptogenic organizing pneumonia, UIP, NSIP, and diffuse alveolar damage. The most common pattern is NSIP³⁹ and cryptogenic organizing pneumonia.⁴⁰ In our study 66.7% cases had NSIP pattern on HRCT. Mixed connective tissue disease, a condition in which manifestations of systemic lupus erythematosus, rheumatoid arthritis, progressive systemic sclerosis, and polymyositis or dermatomyositis are combined, was described by Sharp et al in 1972.41 Thoracic involvement in mixed connective tissue disease occurs with a frequency ranging from 20% to 85%.42-43 In our study almost (fig7) 33% of cases presented with parenchymal abnormalities like reticulations, mosaic attenuation, nodules, and consolidation. Pattern of COP was identified in one of the patient.

CONCLUSION

From this study it is evident that thoracic organs are frequent targets of immune- mediated injury in CTDs. Any thoracic compartment (parenchyma, airway, pleura and mediastinum) can be affected; however, the most important manifestation is interstitial lung disease. HRCT is the diagnostic modality for early detection which can help in prompt treatment for better prognosis.

REFERENCES

- Turesson C, Jacobsson L, Crowson CS, Gabriel SE, Matteson EL. Extra-articular disease manifestations in rheumatoid arthritis: incidence trends and risk factor over 46 year. Ann Rheum Dis. 2003;62(1):722–7.
- Lynch DA. Lung disease related to collagen vascular disease. J Thorac Imaging. 2009;24 (3):299–309.
- Grutters JC, Wells AU, Wuyts W, Schenk P, Leroy S, Dawson JK, et al. Evaluation and treatment of interstitial lung involvement in connective tissue diseases: a clinical up date. EurRespir Mon. 2006;34 (5):27–49.
- Turesson C, O'Fallon WM, Crowson CS, Gabriel SE, Matteson EL. Extra-articular disease manifestation in rheumatoid arthritis: incident trends and risk factors over 46 years. Ann Rheum Dis. 2003;62 (2):722–7.
- Ancochea J, Gómez J, Vilar J, Xaubet A. Consenso para el diagnóstico de las neumoníasintersticialesidiopáticas. Arch Bronconeumol. 2010;46(4):1–21.
- Devaraj A, Wells AU, Hansell DM. Computed tomographic imagingin connective tissue diseases. SeminRespirCrit Care Med.2007;28 (6):389-97.
- Leung AN, Miller RR, Müller NL. Parenchymal opacification inchronic infiltrative lung diseases.CTpathological correlation.Radiology. 1993;188 (3):209-14.
- Churg A, Müller NL. Cellular versus fibrosinginterstitialpneumonias and prognosis: a practical classifi-cation of IIP and pathologically/radiologically similar conditions. Chest. 2006;130 (6):1566-70.
- Webb WR. Normal lung anatomy, technical aspects, In Webb WR., Muller NL., Naidich DP. (Eds): HRCT of the lung, 3rd Ed. Philadelphia: Lippincott Williams and Wilkins, ch. 2001;2(3):1-70.
- 10. Gaude GS, Mahishale V, Srivastva A. Pulmonary manifestations in connective tissue disorders: Hospital-

based study at a tertiary care hospital. Indian J Chest Dis Allied Sci. 2009;51(5):145-51

- 11. Verma SK, Saheer S, Kumar P, Kumar M, Das SK, Prasad R et al. Respiratory manifestations in patients with connective tissue disorders. JIACM. 2013;14(1):28-32.
- Bilgici A, Ulusoy H, Kuru O, et al. Pulmonary involvement in rheumatoid arthritis. Rheumatol Int. 2005;25(1):429– 435.
- Thierry Perez, Martine Remy-Jardin, And Bernard Cortet. Airways Involvement in Rheumatoid Arthritis: Am J RespirCrit Care Med 1998;157 (2):1658–1665.
- Biederer J, Schnabel A, Muhle C, Gross WL, Heller M, Reuter M. Correlation between HRCT findings, pulmonary function tests and bronchoalveolar lavage cytology in interstitial lung disease associated with rheumatoid arthritis. EurRadiol. 2004;14(2):272–280.
- Cortet B, Perez T, Roux N, et al. Pulmonary function tests and high resolution computed tomography of the lungs in patients with rheumatoid arthritis. Ann Rheum Dis. 1997;56(10):596–600.
- Crestani B. The respiratory system in connective tissue disorders. Allergy 2005;60(6):715–734.
- Rockall AG, Rickards D and Shaw PJ. Imaging of the pulmonary manifestations of systemic disease. Postgrad Med J 2001; 77 (5): 621-38.
- Zrour SH, Touzi M, Bejia I, Golli M, Rouatbi N, Sakly N, Younes M, Tabka Z and Bergaoui N. Correlations between high-resolution computed tomography of the chest and clinical function in patients with rheumatoid arthritis: prospective study in 75 patients. Joint Bone Spine 2005;72(3):41-47.
- Tanaka N, Kim JS, Newell JD, Brown KK, Cool CD, Meehan R,et al. Rheumatoid arthritis-related lung diseases: CT findings.Radiology. 2004;232 (3):81-91.
- J.K. Dawson, N.G. Goodson, D.R. Graham, M.P. Lynch, Raised pulmonary arterypressures measured with Doppler echocardiography in rheumatoid arthritispatients, Rheumatology 2000;39(12):1320-1325.
- X. Yang, J. Mardekian, K.N. Sanders, M.A. Mychaskiw, J. Thomas, Prevalence of pulmonary arterial hypertension in patients with connective tissue diseases: a systematic review of the literature, ClinRheumatol 2013;32 (3):1519.
- YsamatMarfá R, Benito Ysamat A, Espejo Pérez S, Blanco Negredo M, Roldán Molina R. Lung disease associated with connective tissue disease. Radiologia 2013;55(5):107-17.
- Bankier AA, Kiener HP, Wiesmayr MN, et al. Discrete lunginvolvement in systemic lupus erythematosus: CT assessment.Radiology. 1995;196(1):835–840.
- Fenlon HM, Doran M, Sant SM, et al. High-resolution chestCT in systemic lupus erythematosus. AJR Am J Roentgenol.1996;166(2):301–307.
- Fekih L, Boussoffara L, Chaouachi S, Fenniche S, et al; Thoracic manifestations of systemic lupus erythematosus .Tunis Med. 2011; 89(3):269-73.
- Kakati S, Doley B, Pal S, Deka UJ. Pulmonary Manifestations in Systemic Lupus Erythematosus (SIE) with Special Reference to HR CT Original Article J API 2007;55(3):839-841.
- Nellessen CM, Pöge U, Brensing KA, Sauerbruch T, Klehr H, Rabe C. Diffuse alveolar hemorrhage in a systemic lupus erythematosus patient successfully treated with rituximab. A case report.Nephrol Dial Transplant. 2008;23(1):385–6.
- Maghrapy HM. Assessment of pulmonary manifestations of collagen diseases: High resolution computed tomography findings. 2013;10(4):16-33.

- Arnaud L, Agard C, Haroche J, Cacoub P, Piette JC, Amoura Z. Pulmonary arterial hypertension in systemic lupus erythematosus. Revue de Medecine Interne.; 32(11):689– 697
- Silver RM, Miller KS: Lung involvement in systemic sclerosis.Rheum Dis Clin North Am 1990, 16(2):199–216.
- Ramirez A, Varga J: Pulmonary arterial hypertension in systemicsclerosis: clinical manifestations, pathophysiology, evaluation, andmanagement. Treat Respir Med 2004, 3(4):339–352.
- Bouros D, Wells AU, Nicholson AG, Colby TV, PolychronopoulosV, Pantelidis P, et al. Histopathologic subsets of fibrosingalveolitis in patients with systemic sclerosis and their relationshipto outcome. Am J RespirCrit Care Med. 2002;165(5):1581-6.
- Kim DS, Yoo B, Lee JS, et al.: The major histopathologic pattern of pulmonary fibrosis in scleroderma is nonspecific interstitialpneumonia. Sarcoidosis Vasc Diffuse Lung Dis 2002;19(5):121–127.
- Minai OA, Dweik RA, Arroliga AC: Manifestations of scleroderma pulmonary disease. Clin Chest Med 1998; 19(2):713–731.
- Hunninghake GW, Fauci AS. Pulmonary involvement in thecollagen vascular diseases. Am Rev Respir Dis. 1979;119(3):471–503.
- Fox RI. Sjögren's syndrome. Lancet. 2005; 366(9482):321– 31.
- Ito I, Nagai S, Kitaichi M, Nicholson AG, Johkoh T, Noma S, et al. Pulmonary manifestations of primary Sjogren's syndrome: a clinical, radiologic, and pathologic study. Am. J. Respir. Crit. Care Med. 2005; 171(6):632–8.
- Parambil JG, Myers JL, Lindell RM, Metteson EL, Ryu JH.Interstitial lung disease in primary Sjögren syndrome. Chest. 2006;130:1489-95.
- Tansey D, Wells AU, Colby TV, Ip S, Nikolakoupolou A, du Bois RM, et al. Variations in histological patterns of interstitial pneumoniabetween connective tissue disorders and their relationshipto prognosis. Histopathology. 2004;44(2):585-96.
- Akira M, Hara H, Sakatani M. Interstitial lung disease in associationwith polymyositis-dermatomyositis: long-term follow-upCT evaluation in seven patients. Radiology. 1999;210(3):333-8.
- Sharp GC, Irvin WS, Tan EM, Gould RG, Holman HR. Mixed connective tissue disease: an apparently distinct rheumatic disease syndrome associated with a specific antibody to an extractable nuclear antigen (ENA). Am J Med 1972; 52(3): 148–159.
- Kim EA, Lee KS, Johkoh T, et al. Interstitial lung diseases associated with collagen vascular diseases: radiologic and histopathologic findings. RadioGraphics 2002;22(spec issue):S151–S165.
- Bodolay E, Szekanecz Z, Dévényi K, et al. Evaluation of interstitial lung disease in mixed connective tissue disease (MCTD). Rheumatology (Oxford) 2005;44(5):656–661.

Source of Support: Nil; Conflict of Interest: None Submitted: 12-02-2018; Published online: 15-03-2018