

# Thoracic Imaging Findings of Collagen Vascular Diseases: A CT Study

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**Background:** collagen vascular diseases (CVDs) are well known causes of pulmonary involvement, leading to significant morbidity. The purpose of this study was to identify several thoracic computed tomographic findings of CVDs.

**Materials and Methods:** The study included 56 patients (15 males and 41 females) with histopathologically and clinically proven CVDs who were identified retrospectively. The presence, extent and distribution of various CT findings were evaluated by a radiologist.

**Results:** Lung parenchyma (96.4%) was the most common area of involvement. The lower lobes (89.2%) were the most frequent sites of involvement. The predominant CT patterns were reticulation (55.3%), peripheral subpleural interlobular septal thickening (51.7%) and ground glass opacity (50%). The most common histopathological findings according to CT features were obliterative bronchiolitis (OB, 44.6%) and non-specific interstitial pneumonia (NSIP, 33.9%). Usual interstitial pneumonia was seen in 12.5% and organizing pneumonia in 26.7% of patients.

**Conclusion:** A combination of reticular pattern, peripheral subpleural interlobular septal thickening and ground glass opacity is seen in the majority of patients with CVDs. The results indicate that OB is more prevalent than what has been reported in previous studies. The CT patterns of pulmonary fibrosis are similar to those in most other studies.

**Key words:** Imaging, Thorax, Collagen vascular disease

## INTRODUCTION

Collagen vascular diseases refer to a group of autoimmune disorders including rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), scleroderma, polymyositis/dermatomyositis and mixed connective tissue disease (MCTD) (1). These multisystemic diseases affect a number of organs such as kidneys, joints, skin,

serous membrane, the lungs and the vascular and nervous systems.

Pleural and pulmonary involvements are detected in all CVDs to some degrees (2). In addition, pulmonary involvement is known as the leading cause of morbidity and mortality in CVDs (3).

Imaging modalities play a significant role in diagnosis of thoracic diseases. Thoracic manifestations of CVDs contain those of parenchyma (interstitial pneumonia), airways (bronchiectasis and upper respiratory track diseases), pleura (pleural effusion and pneumothorax), vascular system (pulmonary hypertension and pulmonary arthritis), secondary disorders (drug complication, infection and malignancy) and several other conditions (4).

CT scan is the most popular cross sectional method for chest imaging; however, high-resolution computed tomography (HRCT) is the selective imaging technique for diagnosis of pulmonary fibrosis, interstitial lung diseases and bronchiectasis (5).

Knowledge about thoracic imaging findings of CVDs can lead to early diagnosis and consequently decrease the hospital stay and improve treatment efficacy. The purpose of this study was to identify several thoracic computed tomographic findings of CVDs.

**MATERIALS AND METHODS**

This retrospective study was conducted on 56 CVD patients including 15 males and 41 females who were admitted to Masih Daneshvari Hospital. Diagnoses were made based on clinical and histopathological assessments and cases with confirmed diagnosis were enrolled in this study. Patients with probable diagnosis or no available imaging were excluded from the study. All patients underwent spiral CT or HRCT scans with Siemens Somatom CT scanner in the hospital and their images were stored.

A chest radiologist observed the images and a checklist of CT findings including location and characteristics of parenchymal involvement, pleural involvement, pulmonary nodules, bronchial dilation, lymphadenopathies and several other parameters was fulfilled for each patient.

Data analysis was carried out using statistical tests for frequency distribution and comparison analysis was performed between different groups classified according to different types of CVDs and histopathological patterns.

**RESULTS**

We evaluated CT findings of 56 CVD patients including 15 males and 41 females with a mean age of 46 years (range 14-82 years). This group of 56 cases included 18 scleroderma, 17 SLE, 17 RA, 3 MCTD and 1 polymyositis/dermatomyositis patient. The highest incidence belonged to scleroderma and the lowest to polymyositis/dermatomyositis (1). Table 1 demonstrates the incidence of each diagnosis.

Table1. The incidence of each type of collagen vascular disease in under-study subjects.

Diagnosis	No.	Frequency (%)
Systemic lupus erythematosus	17	30.3
Rheumatoid arthritis	17	30.3
Scleroderma	18	32.1
Mixed connective tissue disease	3	5.3
Polymyositis/dermatomyositis	1	1.7

Regarding the site of involvement, parenchyma was the most common among each group whereas airways in SLE (11.7%), vascular system in RA (11.7%), pleura in scleroderma (38.8%) and airways and vascular system in MCDT (33.3%) demonstrated the lowest prevalence rate. Generally, parenchyma and vascular system were reported to be the most and the least common site of involvement, respectively. Table 2 shows these data in details.

Analyzing lobar involvement, in order of prevalence, RLL and LLL (89.2%), RML (82.1%), RUL and lingula (76.7%) and LUL (75%) had the highest frequency of involvement. Parenchymal involvement was mostly seen as mixed (55.3%) and peripheral pattern (41.7%).

Frequencies of variables were determined in each group of CVDs separately. Reticular pattern, multifocal ground glass opacity (GGO) and mosaic attenuation in RA (52.9%, n=9 each), reticular pattern and thickening of peripheral subpleural interlobular septum in scleroderma (88.8%,

n=16 each), air space consolidation (64.7%,n=11) and pleural thickening (44.4%,n=8) in SLE and peribronchial bundle thickening in MCTD (100%,n=3) were detected as the most common findings in each type of CVD. Tables 3 and 4 summarize the frequency of all variables.

Table 2. Site of involvement in each type of collagen vascular disease.

Diagnosis	Parenchymal involvement	Pleural involvement	Airway involvement	Vascular involvement
Systemic lupus erythematosus	88.2	58.8	11.7	35.2
Rheumatoid arthritis	100	47	52.9	11.7
Scleroderma	100	38.8	55.5	50
Mixed connective tissue disease	100	66.6	33.3	33.3
Polymyositis/dermatomyositis	100	-	100	-

Table 3. CT findings of each type of disease.

CT finding	Rheumatoid arthritis	Scleroderma	Systemic lupus erythematosus	Mixed connective tissue disease	Polymyositis/dermatomyositis
Reticular pattern	9	16	4	2	-
Honey combing	6	8	-	1	-
Ground glass opacity	10	12	4	2	-
Architectural distortion	1	6	4	2	-
Air space consolidation	1	2	11	-	1
Nodule	2	2	3	-	-
Peripheral subpleural interlobular Septal thickening	11	16	-	2	-
Peri bronchial bundle thickening	7	9	-	3	-
Cystic air space	3	4	1	1	-
Air trapping	5	8	2	1	-
Mosaic pattern	9	10	3	2	1
Bronchial dilation	1	2	2	1	-
Pulmonary artery dilation	2	9	5	1	-
Lymphadenopathy	4	2	1	1	1
Pleural involvement	9	8	13	3	-
Esophageal dilation	-	5	-	1	1
Cardiomegaly	2	2	3	1	-
Pericardial involvement	-	1	4	-	-

Table 4. Frequency of CT findings based on histopathological patterns.

CT finding	UIP	NSIP	OP	OB
Reticular pattern	5	18	6	6
Honey combing	3	10	-	4
Ground glass opacity	2	19	7	14
Architectural distortion	3	7	6	9
Air space consolidation	-	1	14	5
Nodule	-	1	5	4
Peripheral subpleural interlobular Septal thickening	7	19	3	14
Peribronchial bundle thickening	3	14	1	7
Cystic air space	3	5	-	4
Air trapping	4	7	4	11
Mosaic pattern	5	7	4	25
Bronchial dilation	1	1	1	5
Pulmonary artery dilation	5	7	4	10
Lymphadenopathy	-	6	1	3
Pleural thickening	4	5	10	12
Pleural effusion	-	2	4	1
Pericardial involvement	-	-	3	-
Cardiomegaly	1	1	5	3

UIP: Usual Interstitial Pneumonia; NSIP: Non Specific Interstitial Pneumonia; OB: Obliterative Bronchiolitis

## DISCUSSION

We assessed 56 CVD patients and their CT findings were interpreted. Lung parenchyma showed the highest involvement (96.4%), mostly presented in mixed (55.3%) and peripheral pattern (41.7%).

In a study by Lim and colleagues on 23 CVID patients, sub pleural involvement was the most common finding (90%). This difference with our result may be due to wide range of diseases in our study including those with diffuse parenchymal involvement (6).

Furthermore, the most common pattern in our study was reticular pattern (55.3%) versus ground glass opacity in their study (57%). However, GGO occurred in 50% of our cases, similarly (6).

Tanaka and colleagues (7) assessed HRCT of 63 RA cases and, in order of prevalence, UIP (41.2%), OP (41.2%), NSIP (30.1%) and OB (17.4%) were reported. In our study obliterative bronchiolitis demonstrated the highest prevalence (52.9%), which highlights the importance of further studies on obstructive airway diseases in RA. In

both studies, pulmonary fibrosis showed a high prevalence rate. These findings emphasize the significant role of HRCT in early diagnosis of parenchymal involvement and pulmonary fibrosis in RA (7).

Pleura has been mentioned as the most common involved site in RA in the study by Mayberry and colleagues while we found parenchyma as the most common site. Considering that our cases were all hospitalized, this difference may be resulted from higher morbidity of parenchyma in our cases. Therefore, it is suggested to undertake further evaluations to achieve more accurate results in this regard (8).

Mayberry and colleagues reported 91% pulmonary fibrosis prevalence in their study of 23 scleroderma patients but another study showed 20-65 % prevalence of pulmonary fibrosis (8). We detected pulmonary fibrosis in all scleroderma cases. It manifests notable prevalence of pulmonary fibrosis in scleroderma. Similar to our results (NSIP: 61.1%), Tanaka and colleagues found NSIP in 78%

of 80 scleroderma patients. On this subject, our study reached results similar to those of previous studies.

Concerning SLE, Lalani and colleagues mentioned pleural involvement as the most common finding (9) whereas we found parenchymal involvement as the most common finding (88.2%). Similar to RA group, hospitalization is assumed to be a contributory factor and it is necessary to carry out more studies to assess the morbidity rate of parenchymal involvement in SLE. On the other hand, Tanaka and colleagues found UIP to be a rare finding in SLE, which is similar to our result (n=0) (10).

Among polymyositis/dermatomyositis cases, we observed only one case with obliterative bronchiolitis; whereas in the study by Tanaka NSIP was reported to be the most common finding (10). Mino and colleagues mentioned pulmonary involvement as the most common finding (11). Small number of polymyositis/dermatomyositis cases in our study may be responsible for the low rate of pulmonary involvement.

A combination of reticular pattern, peripheral subpleural interlobular septal thickening and ground glass opacity is seen in the majority of patients with CVDs. The results indicate that obliterative bronchiolitis is more prevalent than what has been reported in previous studies. CT manifestations of pulmonary fibrosis in our study were similar to those of most other studies.

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