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The Role of HRCT in Evaluation of Thoracic Manifestations of Collagen Vascular Diseases

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Abstract

Background: High-resolution computed tomography has proven to play a basic role in the detection and characterization of various pulmonary lesions (interstitial, small air way disease etc.) in patients with collagen vascular diseases. Accurate HRCT detection of pulmonary involvement in collagen vascular disease has important therapeutic and prognostic implications.

Aim of Study: The aim of this study is to evaluate role of HRCT in diagnosis of thoracic manifestation of collagen vascular diseases.

Patients and Methods: 100 patients diagnosed as collagen vascular disease were referred to Radiology Department in Faculty of Medicine, Cairo University for HRCT of the chest.

Results: Interstial lung disease was seen in 48 patients (48%), while 31 patients (31%) showed signs of small airway disease.

Conclusion: HRCT is fundamental for evaluation of all thoracic manifestation of collagen vascular diseases.

Key Words: *HRCT – Collagen vascular diseases (CVD).*

Introduction

COLLAGEN vascular diseases are a heterogeneous group of immunologically mediated inflammatory disorders that may affect various organs **[1]**.

Pulmonary alterations (physiologic or anatomic) may be the first manifestations of collagen vascular disease **[2]**.

Accurate detection of pulmonary involvement in collagen vascular disease has important therapeutic and prognostic implications because prompt treatment may lead to improved outcomes **[2]**.

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The collagen vascular diseases that most commonly involve the lung are rheumatoid arthritis, scleroderma, systemic lupus erythematosus, polymyositis and dermatomyositis, mixed connective tissue disease, and Sjögren syndrome **[3,4]**.

The pulmonary manifestations detected by HRCT in case of CVD include diffuse lung involvement (Interstial lung disease, small airway disease, Pulmonary macronodules), oesophageal abnormalities, pleural and pericardial effusions and thickening, pulmonary arterial enlargement, mediastinal lymphadenopathy **[5]**.

The two thoracic manifestations with the greatest clinical importance in patients with collagen vascular diseases are interstitial lung disease and pulmonary arterial hypertension, which are responsible for a large part of the mortality and morbidity in this patient group. Nonspecific interstitial pneumonia has been found to be the most common histological pattern of ILD in patients with collagen vascular diseases **[6]**.

Aim of the work:

The aim of the work is to evaluate the role of High resolution computed tomography of lungs in

Abbreviations:

HRCT	: High resolution computed tomography.
CVD	: Collagen vascular disease.
RA	: Rheumatoid arthritis.
SSC	: Systemic Sclerosis.
SLE	: Systemic Lupus Erythematosus.
DM	: Dermatomyositis.
MCTD	: Mixed Connective Tissue Disease.
SJ	: Sjogren disease.
NSIP	: Nonspecific Interstial pneumonia.
LIP	: Lymphocyte Interstial pneumonia.
UIP	: Usual Interstial pneumonia.
IP	: Interstitial pneumonia.
IPF	: Interstial pulmonary fibrosis.

diagnosis of thoracic manifestations of collagen vascular Diseases.

Patients and Methods

The study was conducted in the period from January 2017 to January 2019 in Radiology Department in Kasr Al Ainy Hospital.

This cross sectional study included 100 patients 12 males and 88 females with age range from 13 to 72 years (mean age of 40.66 years) with collagen vascular disease according to International diagnostic criteria (7) presenting with clinical symptoms suggesting lung affection as progressive dyspnea and chronic cough (Table 1). HRCT of the lungs was done to all patients as requested.

Table (1) Chest main complaints among CVD cases of this study.

They were referred from the outpatient clinic of Rheumatology and Immunology and Chest departments in Kasr Al Ainy Hospital to perform HRCT of lungs in the Radiology Department.

Table (2) Number and percent of cases per collagen disease with gender distribution.

Inclusion criteria:

Patients diagnosed as collagen vascular disease, according to International diagnostic criteria as relayed from records of inpatients in Rheumatology and Immunology department, who presented with symptoms suspicious of pulmonary affection (Table 1).

Exclusion criteria:

- Patient with known associated chest diseases and non-immune mediated pulmonary pathology e.g. tuberculosis.
- Patients with history of smoking.
- Patients with Sarcoidosis.

The study was approved by the ethical committee and informed consent was obtained assuring respect of the confidentiality of the medical records.

Methods:

HRCT chest was done to all patients using 16 channels MSCT (simens) in Kasr Al-Ainy. Reconstructed axial, coronal and sagittal images were done to all patients; also complementary mediastinal images were taken (see Table 3 for HRCT technique used in this study).

Table (3) HRCT techniques used in this study.

Statistics:

The package of statistical analysis used was SPSS version 21. The data was coded, entered on excel sheet and analyzed by SPSS. Numerical data was presented by mean and standard deviation. Qualitative data was presented by frequency and percentage. Non-parametric tests as chi-squared test and ficher exact test were used on cells less than 50%.

Table (1): Chest main complaints among CVD cases of this study.

Disease	Dyspnea	Dry cough	Expectoration
RA	20 (64.5%)	8 (25.9%)	3 (9.6%)
SSc	17 (60.7%)	9 (32.3%)	2 (7%)
SLE	15 (62.5%)	6 (25%)	3 (12.5%)
DM	7 (87.5%)	1 (12.5%)	0 (0%)
MCTD	6 (75%)	1 (12.5%)	1 (12.5%)
SJ	1 (100%)	0 (0%)	0 (%)

Table (2): Number and percent of cases per collagen disease with gender distribution.

CVD	No.of patients and percentage of total cases	Gender distribution	
		Females no.	Males no.
RA	31 (31%)	25	6
SSc	28 (28%)	27	1
SLE	24 (24%)	22	2
DM	8 (8%)	6	2
MCTD	8 (8%)	7	1
SJ	1 (1%)	1	0

Table (3): HRCT techniques used in this study.

Conventional HRCT	
Scout	Kv 130 mA 25 Holding breath
Scan type Helical	
Detector Row 16	
Pitch 1.25mm	
Detector configuration 16x0.6	
Beam collimation 5.0mm	
Gantry tilt 0.0	
FOV Depends on the Patient size	
Reconstructed images	
Axial, sagittal and coronal HRCT images	
WW 1000 WL-700	
Axial MIP images.	
Axial mediastinal window images.	

Results

Interstial lung disease in the form of interstial pneumonia were encountered in 48 patients (48% of cases). They showed bilateral symmetrical predominately sub-pleural basal lesions with involvement of pleural recesses. The types of IP encountered ere UIP, NSIP and LIP. Table (4) shows the distribution of these entities among the patients of collagen vascular diseases.

On HRCT Usual Interstitial pneumonia (UIP) is characterized by the presence of reticular opacities, often associated with traction bronchiectasis. Architectural distortion suggestive of lung fibrosis is also frequently recognized (Fig. 1). Ground-glass attenuation, if present, is less extensive than reticular abnormality. In advanced stages of IPF, honeycombing and lower lung volume loss are usually prominent [8].

The most common features seen at high-resolution computed tomography (CT) in NSIP cases are ground-glass opacities; reticular abnormalities, which usually represent fine fibrosis; and traction bronchiectasis or bronchiolectasis, which is almost universal in patients with fibrotic NSIP (Fig. 2) [9].

Other findings include the presence of consolidation and honeycomb cysts are usually less prominent in NSIP than in UIP [9].

Ground-glass opacities and nodules are almost universal features in LIP (Fig. 3) with cysts seen in about two-thirds of patients [10].

Table (4) Incidence of different types of IP among each CVD entity.

Airway disease was detected in 31 (31%) of patients. Signs of airways diseases include hyperlucency, bronchial wall thickening, mosaic pattern and centrilobular nodules (Fig. 4). HRCT signs of airway affection in different CVD entity is shown in (Table 5).

Table (5) HRCT signs of airway disease among each CVD entity.

Pulmonary hypertension was recorded in 33 patients (33%) (Table 6) illustrating the incidence of pulmonary hypertension in different CVD.

Table (6) Incidence of pulmonary hypertension among different types of CVD in this study.

Cardiomegaly was noted in 17% of CVD cases (Table 7) to see incidence of cardiomegaly in different CVD.

Table (7) Incidence of cardiomegaly among each CVD entity.

Thirty-three of studied cases (33%) had pericardial affection (Fig. 5) in form of effusion or thickening (Table 8).

Pleural affection was recorded in 12 of studied cases (12%) (Table 8).

Mediastinal lymph node enlargement was noted in 34% of cases (Table 8).

Esophageal abnormality was recorded in 28% of cases in form of patulous thoracic esophagus and/or presence of air fluid level within (Table 8).

Table (8) shows the incidence of different HRCT pulmonary lesions in CVD cases.

Table (4): Incidence of different types of IP among each CVD entity.

Disease	UIP	NSIP	LIP
RA	7	4	1
SSc	7	13	2
SLE	1	4	0
DM	1	1	0
MCTD	4	2	0
SJ	0	0	1

Table (5): Distribution of signs of airway affection among each CVD entity.

Disease	Hyperlucency	Bronchial wall thickening	Mosaic pattern	Centrilobular nodules
RA	3	4	1	4
SSc	7	12	3	14
SLE	2	4	2	3
DM	3	3	0	1
MCTD	0	4	2	2
SJ	0	0	0	0

Table (6): Incidence of pulmonary hypertension among different types of CVD in this study.

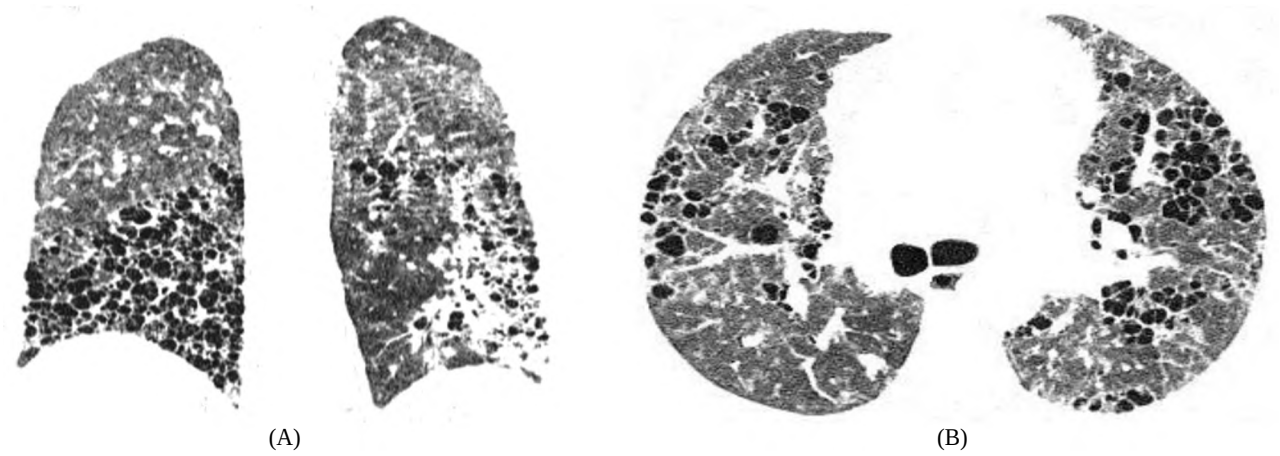
Disease	Pulmonary hypertension
RA	10
SSc	11
SLE	2
DM	5
MCTD	4
SJ	1

Table (7): Incidence of cardiomegaly among each CVD entity.

Disease	Cardiomegaly
RA	1 (3.2%)
SSc	7 (25%)
SLE	6 (25%)
DM	1 (12.5%)
MCTD	2 (25%)
SJ	0

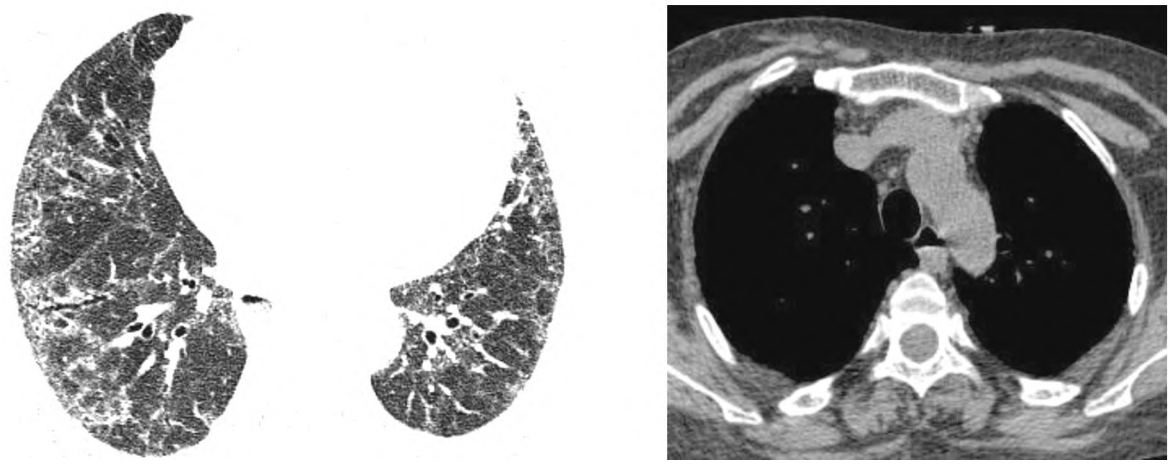
Table (8): Collective incidence of each thoracic manifestation among each CVD group.

Disease	RA	SSc	SLE	DM	MCTD	SJ
IP	39%	78.5%	20%	25%	75%	100%
Airway disease	13%	50%	25%	37.5%	50%	0%
Pulmonary hypertension	33%	32%	8%	62.5%	50%	100%
Pericardial affection	16%	28.5%	62.5%	37.5%	25%	0%
Pleural affection	6.4%	3.5%	33.3%	0%	12.5%	0%
Cardiomegaly	3%	25%	25%	12.5%	25%	0%
Mediastinal lymphadenopathy	35%	28.5%	25%	0%	0%	0%
Esophageal dysmotility	9%	82%	25%	0%	0%	0%



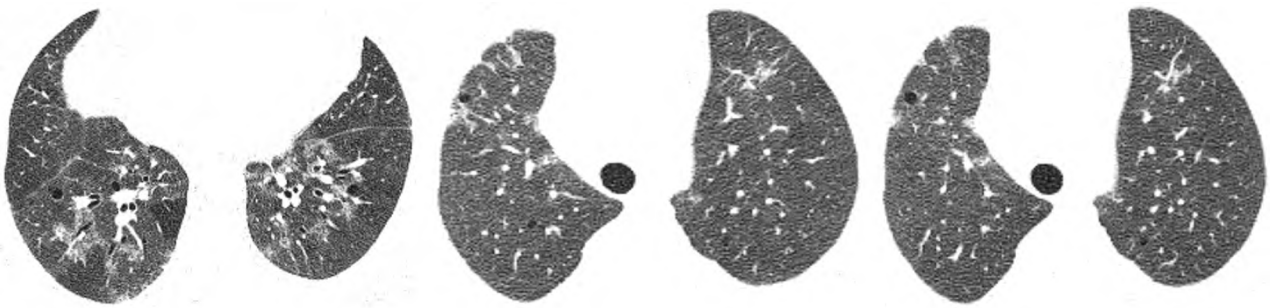
(A) (B)

Fig. (1): 49-year-old female patient known case of Scleroderma presenting with progressive dyspnea on mild exertion and dry cough. (A) Coronal image, (B) Axial image show bilateral fairly symmetrical sub pleural predominantly basal honeycombing, filling pleural recesses. Picture impressive of UIP.



(A) (B)

Fig. (2): 53-year-old female patient known case of Scleroderma presenting with dyspnea on minimal exertion and dry cough. (A) Axial HRCT image shows bilateral fairly symmetrical subpleural ground glass opacification and reticulations with traction bronchiolectasis suggesting fibrotic NSIP. (B) Axial mediastinal image show patulous esophagus with air fluid level within.



(A) (B) (C)

Fig. (3): 42-year-old female patient known case of mixed connective tissue disease presenting with dry cough and dyspnea. (A,B,C) Axial images show few tiny air containing subpleural cysts with bilateral basal ground glass opacifican denoting early interstitial pneumonia with cysts suggesting LIP.

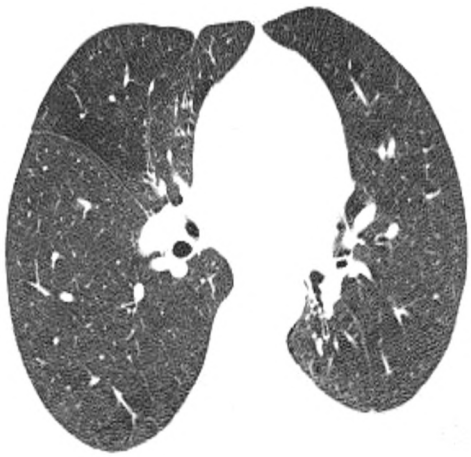
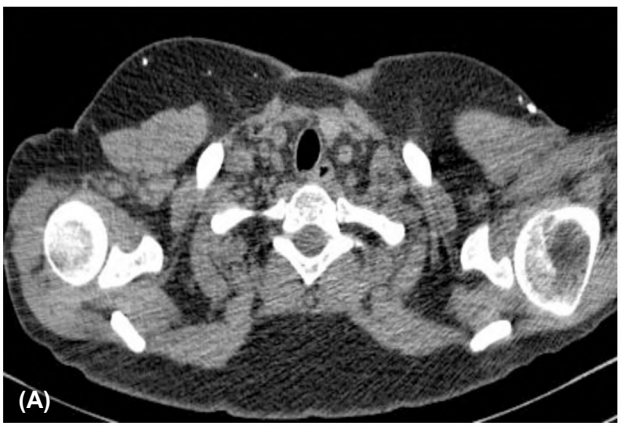
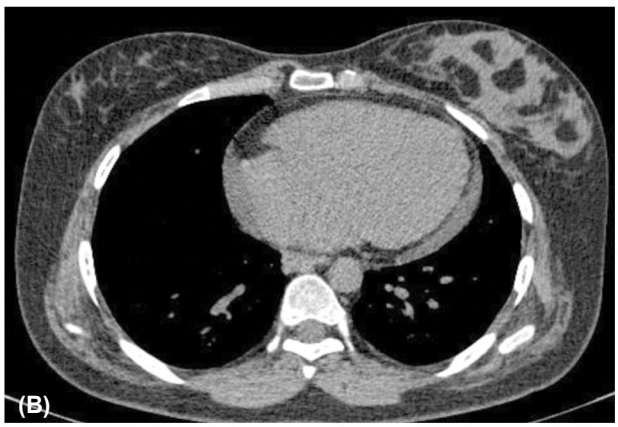
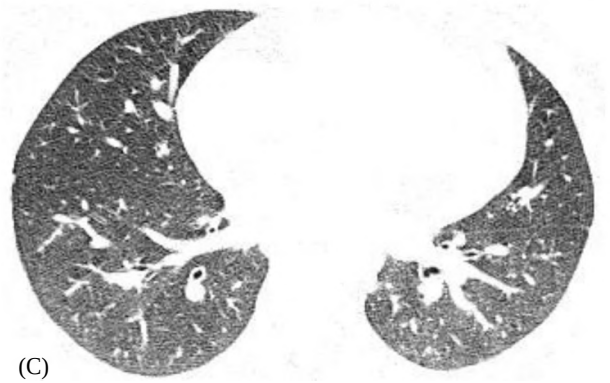


Fig. (4): 39-year-old female patient known case of Rheumatoid arthritis presenting with progressive dyspnea on exertion. (A) Axial HRCT image shows mosaic perfusion with areas of hyperlucency intermingled with areas of normal perfusion. HRCT diagnosis: Bronchiolitis obliterans.







(C)

Fig. (5): 24-year-old female patient known case of Dermatomyositis presenting with dyspnea on mild exertion. (A) Axial mediastinal image shows scattered foci of subcutaneous calcification (B) Axial mediastinal image shows pericardial effusion, (C) Axial HRCT image reveals bilateral bronchial wall thickening denoting air way disease.

Discussion

This study involved 100 cases diagnosed as CVD with clinical manifestations suggestive of pulmonary affection; HRCT of lungs was done to all cases in Kasr Al Ainy Hospital's radiology department in the period from January 2017 to January 2019.

Interstitial pulmonary fibrosis was noted in 48% of this study cases, which agrees with Capobianco [11] who stated that IP is one of the commonest forms of thoracic affection among CVD entities.

NSIP was the most common type of IPF among this study cases, found in approximately 24 patients (24% of cases) followed by UIP found in 20 patients (20% of cases) and LIP was the least common type of ILD (Fig. 3), found in only 4 patients (4% of cases), which agrees with Capobianco [11] and Silva and Müller [12] who noted the same incidence pattern among different CVD entities.

Six of eight (75%) MCTD cases had IPF. The incidence stated by Bodolay [13] was 66.6% of cases and Kinder [14] stated that most of advanced cases of MCTD had ILD.

Airway disease as a single entity was detected in 31 (31%) of patients, which agrees with Lynch [15] who declared the incidence to be 28% and is less than incidence noted by Silva and Müller [12] who declared the incidence to be 40% of cases.

Pulmonary hypertension was recorded in 33% of this study cases, which agrees with Hoeper [16] who reported the incidence to be approximately 30%. Karam [17] reported a higher incidence of 45%.

Thirty-three of studied cases (33%) had pericardial affection (Fig. 5) in form of effusion or thickening, which disagrees Silva and Müller [15] and Capobianco [12] who reported a lower incidence of 14% and 17% respectively for pericardial affection among CVD entities.

Among patients with RA; none of cases had pericardial effusion, while 5 of 31 (16%) cases had pericardial thickening, which partly agrees with Gauhar [18] as regard rare occurrence of pericardial effusion among RA cases (less than 5%) and disagrees with them as regard pericardial thickening incidence noted by him in a higher incidence (50% of cases). Antin-Ozerkis [19] stated that pericardial affection is commonly noted in cases of RA induced pulmonary hypertension.

Pleural affection was recorded in 12 of studied cases (12%), which is much less than incidence noted by Ha et al., [20] and Karam [17] who stated that all CVD groups showed signs of pleural involvement with incidence ranging from 40 and 55% respectively.

Among cases of DM, none had pleural effusion and or thickening, which agrees with Marie [21] and Fathi [22] who stated their DM cases had no signs of pleural involvement.

Cardiomegaly was noted in 17 % of CVD cases, which is close to incidence noted by Karam [17] who reported the incidence to be 14%.

Among patients with SLE, 6 cases (25%) of cases had cardiomegaly with 4 of cases having associated pleural effusion and 4 of cases having associated pericardial effusion, which is lower than incidence reported by Vincze [23] who reported cardiomegaly in 15% of their SLE cases.

Mediastinal lymph node enlargement was noted in 34% of cases, which is lower than incidence reported by Lynch [15] and Silva and Müller [12] who reported the incidence to range from 11 and14% respectively.

Esophageal abnormality was recorded in 28% of cases, which is lower than incidence noted by Karam [17] and Chung [24] reported by them in 10 and 12% of cases respectively.

In cases with SSc; esophageal dilatation occurred in 23 of 28 cases (82%) and was in form of patulous esophagus in 12 cases and air fluid level within esophagus in 11 cases, which agrees with Lynch [15] and Capobianco [11] who stated that esophageal dysmotility was noted in 80 and 90% of SSc cases respectively and disagrees with Karam [17] who reported a lower incidence of 33% among his study cases.

Conclusion:

We concluded that HRCT is fundamental for evaluation of thoracic manifestation of collagen vascular diseases. Early and accurate detection of pulmonary involvement in collagen vascular disease has important therapeutic and prognostic implications because prompt treatment may lead to improved outcomes. So HRCT must be done for any patient with collagen vascular disease even if asymptomatic.

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دور الأشعة المقطعية عالية الجودة فى تقييم المظاهر الصدرية لأمراض الكولاجين الوعائية

شملت هذه الدراسة ١٠٠ حالة (١٢ ذكور و ٨٨ إناث)، تتراوح أعمارهم بين ١٣ إلى ٧٢ سنة، مشخصين بأمراض الكولاجين الوعائية يعانون من أعراض توحى بالتأثرالرئوى. تم إجراء أشعة مقطعية عالية الجودة للرئتين لجميع الحالات فى قسم الأشعة فى مستشفى قصر العينى فى الفترة من يناير ٢٠١٧ إلى يناير ٢٠١٩.

كشف تحليل ٣١ حالة من إلتهاب المفاصل الروماتويدى أن التليف الرئوى الخلالى كان أشيع مظهر صدرى (٣٩٪) وأن الإلتهاب الرئوى الخلالى المعتاد هو النوع الأكثر شيوعاً. كان اعتلال العقد اللمفية المنصفية (٣٥٪) ثانى أكثر مظاهرة شيوعاً يليه ارتفاع ضغط الدم الرئوى (٣٣٪).

فى حالات تصلب الجلد الـ ٢٨ خلل حركة المرئ كان أشيع مظهر صدرى (٨٢٪). كان التليف الرئوى الخلالى ثانى أكثر مظاهرة شيو عاً (٧٨.٥٪). لوحظت أمراض مجارى الهواء فى ٥٠٪ من الحالات، تليها ارتفاع ضغط الدم الرئوى فى ٣٢٪ من الحالات.

شملت الدراسة ٢٤ حالة من الذئبة الحمراء حيث كان تأثر غشاء التامور أشيع مظهر صدرى (٦٢.٥٪) وكان تأثر الغشاء البلورى ثانى أكثر مظاهر الذئبة الحمراء شيوعاً (٣٣.٣٪). لوحظت أمراض مجرى الهواء واعتلال العقد اللمفية المنصفية وتضخم القلب فى نسبة متساوية من الحالات (٢٥٪).

دراسة ٨ حالات إلتهاب الجلد والعضلات كشف ان ارتفاع ضغط الدم الرئوى كان أشيع مظهر صدرى (٦٢.٥٪). لوحظت أمراض مجرى الهواء وتأثر غشاء التامور فى نسبة متساوية من الحالات (٣٧.٥).

\الاستنتاج: الأشعة المقطعية عالية الجودة هى المعيار الذهبى فى تقييم المظاهر الصدرية لأمراض الكولاجين الوعائية. ينصح بدراسة المظاهر الصدرية لكل مرض كولاجين وعائى بشكل منفصل وعلى نطاق اوسع.