Article published online: 2022-06-14

Original Article

Demographic Characteristics and Clinical Manifestations of Interstitial Lung Disease with Systemic Sclerosis in Eastern Part of Libya

Fathi M. Elbraky, Khaled D. Alsaeiti[1], Fathiyah S M Aboulqasim, Saleh M. Alawgali

Department of Internal Medicine, Faculty of Medicine, Benghazi University, [1]Department of Internal Medicine, Aljamhorya Hospital, Benghazi, Libya

Abstract

Introduction: Pulmonary involvement, such as Interstitial Lung Disease (ILD) and Pulmonary Hypertension (PH), accounts for significant morbidity and is the leading cause of Systemic sclerosis (SSc)-related morbidity and mortality. The aim of the current study was to study the frequency of occurrence of ILD in SSc and to describe the clinical and radiological picture of pulmonary involvement in SSc. **Patients and Methods:** Forty patients attending different rheumatology clinics in eastern part of Libya between January 2018 and September 2020 were included. Basic details including age, gender, disease duration, detailed history, and clinical examination were noted. Autoimmune profiling included rheumatoid factor, antinuclear antibodies, and anti-Scl-70 antibodies. Pulmonary function test, chest X-ray (CXR), and high-resolution computed tomography of the chest (HRCT) in all patients were recorded. Data presented either as frequencies and percentages or as means ± standard deviation. **Results:** The male: female ratio was 1:9 with a mean age of 37.5 ± 9.6 years and duration of illness 6 ± 4 years. diffuse cutaneous SSc was seen in 62.5% of the patients. 77.5% of the participants had bilateral crepitation and 57.5% had loud P2. Presenting complaints included gastrointestinal reflux in 72.5%, digitalis ulcerations in 40%, and synovitis/arthritis of all patients. Other comorbidities included congestive heart failure in 12.5%, PH in 15%, and renal impairments in 7.5% of all patients. Anti-Scl-70 antibody was the most common in all patients (45%), followed by anti-centromere Ab (25%), anti-U3 RNP (10%), and anti-U1 RNP (5%). 72.5% of the participants had reticulonodular shadows on CXR. HRCT showed honeycombing as the predominant finding (37.5%). Echocardiograms showed that 15% of all patients have signs of PH. Duration of disease, dyspnea, cough, bilateral crepitations, and CXR were found to be significantly associated with extensive ILD ($P < 0.05$). **Conclusion:** ILD is a serious complication of SSc, it is more common among patients with dcSSc. Chest HRCT is very sensitive to detect ILD. A significant association was found in Libyan patients between the severity of ILD and the duration of disease, dyspnea, cough, bilateral crepts, and CXR.

Keywords: Interstitial lung disease, Libya, systemic sclerosis

Introduction

Systemic sclerosis (SSc) is a multisystem connective tissue disease with unknown etiology, characterized by aberrant immune activation, endothelial dysfunction, vascular injury, and fibroblast dysfunction with resultant excessive collagen production and fibrosis of the skin and various internal organs.[1,2]

Pulmonary involvement, such as interstitial lung disease (ILD) and pulmonary hypertension (PH), accounts for significant morbidity and is the leading cause of SSc-related morbidity and mortality.[3,4] The exact prevalence of ILD in SSc is difficult to estimate because the patient is clinically asymptomatic early in the course.[5] It has been detected in 50%–60% of SSc patients by high-resolution computed tomography (HRCT).[6] The risk factors for the development of SSc-ILD include diffuse cutaneous systemic sclerosis (dcSSc),[7] shorter disease duration,[8] older age at disease onset, and the presence of anti-topoisomerase

Access this article online

Quick Response Code:

Website: journal.limu.edu.ly

DOI: 10.4103/liuj.liuj_51_20

Address for correspondence: Dr. Khaled D. Alsaeiti, Department of Internal Medicine, Aljamhorya Hospital, Benghazi, Libya. E-mail: kads2930@yahoo.com

How to cite this article: Elbraky FM, Alsaeiti KD, Aboulqasim FS. Alawgali SM. Demographic characteristics and clinical manifestations of interstitial lung disease with systemic sclerosis in eastern part of Libya. Libyan Int Med Univ J 2021;6:27-32.

Received: 29-12-2020 **Revised:** 01-03-2021
Accepted: 28-03-2021 **Published:** 21-06-2021

I antibody and/or the absence of anticentromer antibody (ACA).[7] ILD typically occurs early in the course of dcSSc, especially within 3 years after the onset of disease,[7,8] whereas ILD occurs at any time of disease course in limited cutaneous systemic sclerosis (lcSSc) patients.[9] The clinical course of SSc-ILD is variable; some patients show stability in forced vital capacity (FVC), while others show a progressive decline in lung function.[10] ILD mostly progresses within 4 years after the onset of SSc, and afterward, the progression becomes slow or stops completely, even without any treatment.[11] It is reported that severe ILD, showing a decline in FVC below 50%, constitutes around 15% of total SSc.[12]

The aim of the current study was to investigate the characteristics and clinical manifestations of ILDs with SSc in eastern part of Libya.

Patients and Methods

Of all patients attending different rheumatology clinics in eastern part of Libya between January 2018 and September 2020, forty patients (36 females and 4 males) were included in this study. The selected patients were diagnosed as having SSc and fulfilled the American College of Rheumatology/European League Against Rheumatism 2013 revised classification criteria of having either a dcSSc or lcSSc disease subset for SSc.[13] Patients with other collagen vascular diseases/mixed connective tissue disorders and overlap syndromes were excluded.

After obtaining informed consent, basic demographic details were collected, including age, gender, disease duration at the time of presentation, detailed history, and clinical examination. Autoimmune profiling included rheumatoid factor, antinuclear antibodies (ANAs), and autoantibodies against topoisomerase I (anti-Scl-70 antibodies) immunofluorescence in all the patients and its various patterns were noted.

Pulmonary function test (PFT) was performed according to the American Thoracic Society guidelines.[14] Restrictive lung disease was diagnosed if the percentage of predicted FVC was <80%.[15] Obstructive lung disease was diagnosed if the forced expiratory volume at 1 s/FVC was <70%. All patients underwent a chest radiograph, and HRCT was performed when indicated.

Pulmonary involvement was defined as either pulmonary fibrosis (bilateral reticular nodular on chest X-ray [CXR], interstitial pneumonitis/ground-glass opacities/fibrosis on HRCT) or FVC <70% of predicted.[16]

Data were analyzed using the Statistical Package for the Social Sciences (SPSS) 17.0 (SPSS Inc., Chicago, IL, USA). Descriptive statistics of the different variables were presented as frequencies and percentages or as means ± standard deviation. For statistical comparisons, independent samples Chisquare test was employed for testing statistical significance of association between two discrete variables. Significant value is set up at $P<0.05$.

Results

Patients data

The baseline characteristics of the study participants are shown in Table 1. The male: female ratio was 1:9 with a mean age at diagnosis 37.5 ± 9.6 years and duration of illness 6 ± 4 years. Diffuse SSc was seen in 62.5% of the patients with the rest diagnosed with limited SSc. Using the New York Heart Association function test, 75% of patients suffered from dyspnea of different intensity and 37% suffered from dry cough. The respiratory symptoms leading to diagnosis of ILD were apparent in 45% of patients [Table 1].

Common presenting complaints and comorbidities

The most common presenting complaints included gastrointestinal reflux (GIR) in 72.5%, digitalis ulcerations in 40%, and synovitis/arthritis (diffuse 40% and limited 25%) of all patients. Other comorbidities included congestive heart failure in 12.5%, PH in 15%, and renal impairments in 7.5% of all patients [Table 2].

Table 1: Demographic characteristics of the studied participants

Variables	Mean±SD, *n* (%) (total 40)
Male: female	1:9 (4 males, 36 females)
Age at diagnosis (years)	25-65 (37.5±9.6)
Disease duration (years)	2-10 (6±4)[#]
Type of SSc (scleroderma)	
Diffuse SSc	25 (62.5)*
Limited SSc	15 (37.5)
NYHA functional class	
No dyspnea	10 (25)
I-II	20 (50)[#]
III-IV	10 (25)
Dry cough	15 (37.5)[#]
Respiratory symptoms leading to diagnosis of ILD	18 (45)

*Significantly different from limited SSC ($P<0.05$), [#]Significantly associated with extensive ILD with FVC <70 ($P<0.05$). SD: Standard deviation, SSc: Systemic sclerosis, NYHA: New York Heart Association, FVC: Forced vital capacity, ILD: Interstitial lung disease

Table 2: Common presenting complaints of the studied participants

Cinical symptom/sign	*n*(%)
Gastrointestinal reflux, *n* (%)	29 (72.5)[#]
Digital ulceration, *n* (%)	16 (40)
Synovitis/arthritis, *n* (%)	
Diffuse	16 (40)
Limited	10 (25)
Comorbidities, *n* (%)	
Congestive heart failure	5 (12.5)
Pulmonary hypertension	6 (15)
Renal impairment	(7.5)
Myositis	3 (7.5)

[#]Significantly associated with extensive ILD with FVC <70 ($P<0.05$). FVC: Forced vital capacity, ILD: Interstitial lung disease

Systemic findings

Among systemic findings, 31 participants (77.5%) had bilateral crepitation and 23 (57.5%) participants had loud P2. Among the forty participants with ILD, all of them had skin thickening and Raynaud's phenomenon [results are not shown in a table].

Blood investigations

Blood investigations are given in Table 3. Thirty-seven and a half percent of the participants had hemoglobin <10 g/dL. Erythrocyte sedimentation rate of more than 30 mm/h was seen in 52.5%, and 10% of participants had creatinine blood levels of more than 1.2 mg/dL. ANAs were present in 15% of the participants. On analysis of the immunoblotting for ANAs, anti-Scl-70 was the most common and seen in 45%, followed by anti-centromere Ab (25%), anti-U1 RNP (5%), and anti-U3 RNP in 10% of the participants [Table 3].

Diagnostic tests

Among radiological features, 72.5% of the participants had reticulonodular shadows on CXR [Table 4]. PFT showed that 45% of all participants had FVC <70%. Among forty participants, HRCT showed ground-glass appearance in 20% of the patients; honeycombing was the predominant finding in 37.5%, followed by traction bronchiectasis seen in 20% of the participants. regarding echocardiographic findings, 12.5% of the patients found to have cardiomyopathy, 15% have signs of PH which was confirmed in 10% of them by right-sided cardiac catheterization.

Statistically, the duration of the disease, dyspnea, dry cough, GIR, bilateral crepitation, and reticulonodular shadow in CXR were all significantly associated with extensive ILD with FVC <70 (P<0.05). Other factors, such as anti-Scl-70 positivity and abnormal ECHO, were not significantly associated with pulmonary involvement ($P > 0.05$).

Table 3: Blood investigations of the studied participants

Characteristics	*n* (%)
Hemoglobin (<10 g/dL)	15 (37.5)
Erythrocyte sedimentation rate (>30 mm/h)	21 (52.5)
Creatinine blood levels (>1.2 mg/dL)	4 (10)
Antinuclear antibodies	6 (15)
Anti-Scl-70	18 (45)
Anti-centromere Ab	10 (25)
Anti-U1 RNP	2 (5)
Anti-U3 RNP	4 (10)
Ribonucleoprotein	

Table 4: Diagnostic tests done for the studied participants

Test	*n*(%)	
CXR (reticulonodular shadow), *n* (%)	29 (72.5)#	
Pulmonary function test	**Baseline**	**6 months later**
FVC <70 (extensive disease), *n* (%)	18 (45)	14 (35)
FVC ≥70 (limited disease), *n* (%)	22 (55)	26 (62.5)
HRCT, *n* (%)		
Ground-glass	6 (20)	
Reticulonodular	1 (2.5)	
Honeycomb	15 (37.5)	
Traction bronchiectasis	7 (20)	
ECHO, *n* (%)		
Abnormal	12 (30)	
Normal	38 (95)	

Significant *P*=0.001 CXR: Chest X-ray, FVC: Forced vital capacity, ECHO: Echocardiogram, HRCT: High-resolution computed tomography

Treatments given to patients

The results of treatments given to the patients are not shown in a table. The treatments started in all patients with low doses of oral steroids (10 mg or less). Two patients (5%) received mycophenolate 2 g daily. Sixteen patients (40%) received cyclophosphamide 1 g monthly for 6 months. Two patients (5%) received methotrexate up to 12 mg weekly. Comparing response to treatment among our patients 6 months later, we found a partial response with improvement of PFT and stationary disease progression on serial HRCT scan among those who received cyclophosphamide. No deterioration of PFT was reported among the one patient who received methotrexate, or the two patients who received mycophenolate.

Unfortunately, five patients (12.5%) died during the study period: two of them (5%) were complicated by PH and renal crises, two (5%) have severe cardiomyopathy, and one (2.5%) has severe lung disease with renal impairment.

Discussion

Few publications reported findings specific to the SSc-ILD population.[17] ILD was estimated to affect ~35% of the SSc patients in Europe and ~52% in North America; however, the method of ILD assessment may potentially contribute to differences in the observed frequencies. For example, when diagnosed through HRCT, ILD was estimated to affect 32.3%–47.0% of the SSc patients in Europe, whereas only 18.8% of the patients were reported to be affected when diagnosed based on reduced lung function.[18] According to the European Scleroderma Trials and Research group, ILD was reported to affect 53% of the cases with dcSSc and 35% of the cases with lcSSc.[19]

The mean age at diagnosis of SSc-ILD patients in the present study (37.5 ± 9.6 years) was comparatively similar to patients reported from Egypt (40.6 ± 12.5)[20] and slightly lower than the mean age of patients reported in the UK (46 ± 11 years) over the period of 1985–2001[21] and (61.8 ± 11.1) from 2000 to 2009.[22] Another study from the USA in the period from 1997 to 2013 reported a mean age at diagnosis of SSc-ILD of (54.5 ± 13.2 years).[23] Similarly, the Pulmonary Hypertension Assessment and Recognition of Outcomes in Scleroderma (PHAROS) registry in the United States and Canada estimated the median age of SSc-ILD patients to be 52.5 years.[24]

As for many autoimmune diseases,[25] most patients with SSc-ILD are female. Our current study revealed that SSc-ILD

is predominantly more frequent among females, with a male: female ratio of 1:9. Same results were reported by an Egyptian study with male to female ratio of 1:14[20] another UK cohort study which was conducted over the period of 2000-2009 reported a female predominance of pulmonary involvement among SSc patients.[22] PHAROS registry also found predominant female gender of 78%[24] and accounting for 89% of SSc-ILD patients who had undergone HRCT in a cross-sectional study conducted in Canada.[22]

In the current study, 32 patients (80%) have evidence of ILD by HRCT of the chest; this was consistent with Hafez *et al*.[20] and Solomon *et al*.[26] The most common patterns in the HRCT in the current study were honeycombing followed by ground-glass opacity, traction bronchiectasis, and reticulonodular shadowing. Compared with other studies done,[19,20] ground-glass opacity was the most frequent finding on HRCT, followed by septal thickening, honeycombing, bronchiectasis, and consolidations.

In the current study, PH was reported among 15% of patients; a higher prevalence was reported by other studies,[20,21,26,27] who reported PH in 13%–35% of patients. Furthermore, Hachulla *et al*.[28] showed that 55% of patients had PH. This difference could be due to dependence on transthoracic ECHO assessment, with no further evaluation by right-sided heart catheterization.

As expected, ILD was more prevalent among patients with dcSSc compared with those with lcSSc (62.5% and 37.5%, respectively). This is consistent with other reported studies.[19,20,29]

The most frequently used therapy was cyclophosphamide 1 g monthly for 6 months; we found a partial response with improvement of PFT and stationary disease progression on serial HRCT. Becker *et al*. described a high SSc-ILD response rate assessed by FVC and diffusing capacity for carbon monoxide (DLCO) in patients with low FVC values before cyclophosphamide therapy.[30] On the other hand, two meta-analyses failed to show a significant benefit of cyclophosphamide on SSc-ILD lung functions.[31] The same results were reported by Adler *et al*.,[32] in which analysis of using cyclophosphamide alone or in combination with steroids did not result in differences in the slope of DLCO or FVC values compared to all other patients.

In this study, a significant association was found between severity of ILD and patients with gastroesophageal reflux disease (GERD), longer duration of disease, dyspnea, cough, bilateral crepts, and CXR. The same results were reported by Hafez *et al*.[20] who reported that longer disease duration is associated with increased risk of both ILD and PH, dry cough, crepitations, and dyspnea. The association between ILD and GERD is well documented in a recent study.[33]

Conclusion

ILD is a serious complication of SS; it is more common among patients with dcSS. Chest HRCT is very sensitive to detect ILD. In this study, a significant association was found between severity of ILD and patients with GERD, longer duration of the disease, dyspnea, cough, bilateral crepts, and CXR.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Asano Y. Systemic sclerosis. J Dermatol 2018;45:128-38.
2. Hochberg MC, Smolen JO, Weinblatt ME, Weisman MH. Rheumatology. 3rd ed., Vol. 2., Ch. 132-135., Sec. 10. USA: Mosby; 2003. p. 1455-506.
3. Kane GC, Varga J, Conant EF, Spirn PW, Jimenez S, Fish JE. Lung involvement in systemic sclerosis (scleroderma): Relation to classification based on extent of skin involvement or autoantibody status. Respir Med 1996;90:223-30.
4. Elhai M, Meune C, Boubaya M, Avouac J, Hachulla E, Balbir-Gurman A, *et al*. Mapping and predicting mortality from systemic sclerosis. Ann Rheum Dis 2017;76:1897-905.
5. Witt C, Borges AC, John M, Fietze I, Baumann G, Krause A. Pulmonary involvement in diffuse cutaneous systemic sclerosis: Broncheoalveolar fluid granulocytosis predicts progression of fibrosing alveolitis. Ann Rheum Dis 1999;58:635-40.
6. Steele R, Hudson M, Lo E, Baron M; Canadian Scleroderma Research Group. Clinical decision rule to predict the presence of interstitial lung disease in systemic sclerosis. Arthritis Care Res (Hoboken) 2012;64:519-24.
7. Nihtyanova SI, Schreiber BE, Ong VH, Rosenberg D, Moinzadeh P, Coghlan JG, *et al*. Prediction of pulmonary complications and long-term survival in systemic sclerosis. Arthritis Rheumatol 2014;66:1625-35.
8. Jaeger VK, Wirz EG, Allanore Y, Rossbach P, Riemekasten G, Hachulla E, *et al*. Incidences and risk factors of organ manifestations in the early course of systemic sclerosis: A longitudinal EUSTAR study. PLoS One 2016;11:e0163894.
9. Asano Y, Ihn H, Yamane K, Kubo M, Tamaki K. The prevalence and clinical significance of anti-U1 RNA antibodies in patients with systemic sclerosis. J Invest Dermatol 2003;120:204-10.
10. Man A, Davidyock T, Ferguson LT, Ieong M, Zhang Y, Simms RW. Changes in forced vital capacity over time in systemic sclerosis: Application of group-based trajectory modelling. Rheumatology (Oxford) 2015;54:1464-71.
11. Steen VD, Conte C, Owens GR, Medsger TA Jr. Severe restrictive lung disease in systemic sclerosis. Arthritis Rheum 1994;37:1283-9.
12. Morgan C, Knight C, Lunt M, Black CM, Silman AJ. Predictors of end stage lung disease in a cohort of patients with scleroderma. Ann Rheum Dis 2003;62:146-50.
13. van den Hoogen F, Khanna D, Fransen J, Johnson SR, Baron M, Tyndall A, *et al*. 2013 classification criteria for systemic sclerosis: An American College of Rheumatology/European League against Rheumatism collaborative initiative. Arthritis Rheum 2013;65:2737-47.
14. Villalba WO, Sampaio-Barros PD, Pereira MC, Cerqueira EM, Leme CA Jr., Marques-Neto JF, *et al*. Six-minute walk test for the evaluation of pulmonary disease severity in scleroderma patients. Chest 2007;131:217-22.
15. Gilson M, Zerkak D, Wipff J, Dusser D, Dinh-Xuan AT, Abitbol V, *et al*. Prognostic factors for lung function in systemic sclerosis: Prospective study of 105 cases. Eur Respir J 2010;35:112-7.
16. Assassi S, Sharif R, Lasky RE, McNearney TA, Estrada-Y-Martin RM, Draeger H, *et al*. Predictors of interstitial lung disease in early systemic sclerosis: A prospective longitudinal study of the GENISOS cohort. Arthritis Res Ther 2010;12:R166.
17. Bergamasco A, Hartmann N, Wallace L, Verpillat P. Epidemiology of systemic sclerosis and systemic sclerosis-associated interstitial lung disease. Clin Epidemiol 2019;11:257-73.
18. Vonk MC, Broers B, Heijdra YF, Ton E, Snijder R, van Dijk AP, *et al*. Systemic sclerosis and its pulmonary complications in The Netherlands: An epidemiological study. Ann Rheum Dis 2009;68:961-5.

19. Cappelli S, Bellando Randone S, Camiciottoli G, de Paulis A, Guiducci S, Matucci-Cerinic M. Interstitial lung disease in systemic sclerosis: Where do we stand? Eur Respir Rev 2015;24:411-9.
20. Hafez EA, Hamza SH, Morad CS, Abd Alkader AA. Pulmonary manifestations in Egyptian patients with systemic sclerosis. The Egyptian Journal of Internal Medicine 2018;40:39-44.
21. Davie N, Haleen SJ, Upton PD, Polak JM, Yacoub MH, Morrell NW, *et al*. ET(A) and ET(B) receptors modulate the proliferation of human pulmonary artery smooth muscle cells. Am J Respir Crit Care Med 2002;165:398-405.
22. Wan ES, DeMeo DL, Hersh CP, Shapiro SD, Rosiello RA, Sama SR, *et al*. Clinical predictors of frequent exacerbations in subjects with severe chronic obstructive pulmonary disease (COPD). Respir Med 2011;105:588-94.
23. Ryerson CJ, O'Connor D, Dunne JV, Schooley F, Hague CJ, Murphy D, *et al*. Predicting mortality in systemic sclerosis-associated interstitial lung disease using risk prediction models derived from idiopathic pulmonary fibrosis. Chest 2015;148:1268-75.
24. Hao Y, Hudson M, Carreira P, Stevens W, Rabusa C, Tatibouet S, *et al.* Early mortality in Australian, Canadian and Spanish scleroderma patients: rationale for establishing a multi-national inception cohort of patients with systemic sclerosis [abstract]. Arthritis Rheum. 2014;66(Suppl 10):S316.
25. Whitacre CC. Sex differences in autoimmune disease. Nat Immunol 2001;2:777-80.
26. Solomon J, Olson A, Fischer A, Bull T, Brown K, Raghu G. Scleroderma lung disease. Eur Respir Rev 2013;22:6-19.
27. de Azevedo AB, Sampaio-Barros PD, Torres RM, Moreira C. Prevalence of pulmonary hypertension in systemic sclerosis. Clin Exp Rheumatol 2005;23:447-54.
28. Hachulla E, Launay D, Mouthon L, Sitbon O, Berezne A, Guillevin L, *et al.* Is pulmonary arterial hypertension really a late complication of systemic sclerosis? Chest 2009;136:1211-9.
29. Steen VD, Medsger TA Jr. Severe organ involvement in systemic sclerosis with diffuse scleroderma. Arthritis Rheum 2000;43:2437-44.
30. Becker MO, Schohe A, Weinert K, Huscher D, Schneider U, Burmester GR, *et al*. Responders to cyclophosphamide: Results of a single-centre analysis among systemic sclerosis patients. Ann Rheum Dis 2012;71:2061-2.
31. Nannini C, West CP, Erwin PJ, Matteson EL. Effects of cyclophosphamide on pulmonary function in patients with scleroderma and interstitial lung disease: A systematic review and meta-analysis of randomized controlled trials and observational prospective cohort studies. Arthritis Res Ther 2008;10:R124.
32. Adler S, Huscher D, Siegert E, Allanore Y, Czirják L, DelGaldo F, *et al*. Systemic sclerosis associated interstitial lung disease – Individualized immunosuppressive therapy and course of lung function: Results of the EUSTAR group. Arthritis Res Ther 2018;20:17.
33. Bédard Méthot D, Leblanc É, Lacasse Y. Meta-analysis of gastroesophageal reflux disease and idiopathic pulmonary fibrosis. Chest 2019;155:33-43.

ملخص المقال باللغة العربية

الخصائص الديموغرافية والمظاهر السريرية لمرض الرئة الخلالي مع التصلب الجهازي في الجزء الشرقي من ليبيا

المؤلفون:

فتحي البركي، خالد السعيطي، فتحية بلقاسم [1]، صالح الاوجلي [1]

قسم الأمراض الباطنة، كلية الطب، جامعة بنغازي، [1] قسم الأمراض الباطنة، مستشفى الجمهورية، بنغازي، ليبيا.

المؤلف المسؤول: خالد السعيطي. **البريد الإلكتروني**: kads2930@yahoo.com

مقدمة: مرض الرئة الخلالي وارتفاع ضغط الدم الرئوي، يمثلان مراضة كبيرة، وهو السبب الرئيسي للتصلب الجهازي والوفيات المرتبطة به. كان الهدف من الدراسة الحالية هو دراسة تواتر حدوث مرض الرئة الخلالي في التصلب الجهازي ووصف الصورة السريرية والإشعاعية للتورط الرئوي في التصلب الجهازي.

المرضى والطرق: تم تضمين أربعين مريضاً في عيادات أمراض الروماتيزم المختلفة في الجزء الشرقي من ليبيا بين يناير 2018 وسبتمبر 2020م. تم تسجيل التفاصيل الأساسية بما في ذلك العمر والجنس ومدة المرض والتاريخ التفصيلي والفحص السريري. اشتمل تحديد خصائص المناعة الذاتية على عامل الروماتويد، والأجسام المضادة للنواة، والأجسام المضادة ضد توبويزوميراز 1. تم تسجيل اختبار وظائف الرئة، والأشعة السينية للصدر، والتصوير المقطعي المحوسب عالي الدقة للصدر في جميع المرضى. تم تقديم البيانات إما على شكل تكرارات ونسب مئوية أو المتوسط الحسابي ± الانحراف المعياري.

النتائج: كانت نسبة الذكور إلى الإناث 1 إلي 9 بمتوسط عمر 37.5 ± 9.6 سنة، ومدة المرض 6 ± 4 سنوات. لوحظ انتشار تصلب الجلد الجهازي في 62.5٪ من المرضى. 77.5٪ من المشاركين لديهم كراكر (خرخرة) ثنائية الجانب للرئتين و 57.5٪ لديهم P2 بصوت عالٍ في الرئتين. تضمنت شكاوى المرضي ارتجاع معدي معوي بنسبة 72.5٪، تقرحات الأطراف بنسبة 40٪، والتهاب الغشاء المفصلي والتهاب المفاصل لدى جميع المرضى. شملت الأمراض المصاحبة الأخرى فشل القلب الاحتقاني في 12.5٪، ارتفاع ضغط الدم الرئوي في 15٪، والاعتلال الكلوي في 7.5٪ من جميع المرضى. كانت الأجسام المضادة ضد توبويزوميراز 1 الأكثر شيوعًا في جميع المرضى (45٪) ، يليه الأجسام المضادة للنواة (25٪) ، الأجسام المضادة الذاتية المضادة للفيبريلارين (10٪)، والأجسام المضادة الذاتية البروتين النووي الصغيرة (5٪). كان لدى 72.5٪ من المشاركين ظلال شبكية للأشعة السينية للصدر، كما أظهر التصوير المقطعي المحوسب عالي الدقة للصدر مساحات هوائية كيسيه عنقودية في نسبة عالية من المرضي (37.5٪). أظهرت مخططات صدى القلب أن 15٪ من جميع المرضى لديهم علامات ارتفاع ضغط الدم الرئوي. وجدت علاقة ذات دلالة إحصائية ما بين مدة المرض، وضيق التنفس، والسعال، والتشققات الثنائية، ونتائج أشعة الصدر السينية مع مرض الرئة الخلالي (P <0.05).

الخلاصة: مرض الرئة الخلالي هو أحد المضاعفات الخطيرة للتصلب الجهازي، والأكثر شيوعاً كان تصلب الجلد الجهازي. إن التصوير المقطعي المحوسب عالي الدقة للصدر حساس جدًا للكشف عن مرض الرئة الخلالي. وجد أنه هناك ارتباط كبير في المرضى الليبيين بين شدة مرض ومدته، وضيق التنفس، والسعال، ونتائج أشعة الصدر السينية مع مرض الرئة الخلالي.

الكلمات المفتاحية: مرض الرئة الخلالي، ليبيا، تصلب جهازي.