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Clinical and Diagnostic Profile of Interstitial Lung Disease

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Abstract

Background: Interstitial lung disease are clinically challenging and diverse group of over 300 disorders. These heterogeneous group of disorders share few common clinical radiological and pathological profiles.

Objective: To analyse these Clinical, spirometric, laboratory, radiological and bronchoscopic profile of various ILDs.

Methods: It is an Institutional based prospective study of 50 cases of Interstitial Lung Disease presenting to pulmonology department. After Obtaining Informed Consent patients were subjected to detailed Clinical, spirometric, laboratory, radiological and bronchoscopic examination. Initially a total of 60 patients were enrolled in the study .Out of which 4 patients were excluded based on exclusion criteria and 6 patients were lost to follow up.

Results: After Analysing 50 ILD patients, it showed a female preponderance, presenting mostly in 36-55 year age group with shortness of breath and cough as the predominant complaints. Majority of the patients showed exercise oxygen desaturation and a restrictive abnormality in spirometric evaluation. Radiologically, reticulonodular pattern was the most common presentation on CXR and reticular opacities on HRCT. Most common HRCT pattern was UIP pattern with basal predominance. The most common cause of ILD was IPF (30%) followed by LDCTD which comprised 28% of the cases.

Conclusion: Patients, in their third to fifth decade, especially females, presenting with complaints of breathlessness and cough, with exercise oxygen desaturation, should be evaluated for ILD with complete profile to identify the disease at an early stage.

Keywords: Prospective study, Interstitial lung disease profile, female preponderance, exercise oxygen desaturation, UIP pattern, IPF.

Introduction

Interstitial lung disease represent a large and heterogeneous group of over 300 disorders, many of them belonging to the category of orphan diseases and are characterized by varying degrees of fibrosis and inflammation of the lung parenchyma or interstitium. The interstitial lung disease represent many features in common such as similarities of symptoms, comparable appearance of chest imaging studies, consistent alterations in pulmonary physiology and typical histological features.[1]

Current estimates of incidence and prevalence of ILD are higher than historical estimates. The prevalence of all ILD is estimated at 1 in 3000 to 4000 globally.[2] Among those, Idiopathic Pulmonary Fibrosis is the most common disease representing at least 30% of the incident cases. The disease process extends into alveolar spaces, acini, bronchiolar lumen and bronchioles. The inflammation usually referred to as alveolitis is associatedwith spread to adjacent portions of interstitium and vasculature, resulting in derangement of alveolar capillary architecture leading to alveolo-capillary membrane damage and loss of gas exchange units eventually ending in interstitial fibrosis.[3]The scarring and distortion of lung tissue leads to deranged gas exchange & ventilator function.

Clinical evaluation is the foremost and central pillar for suspecting the presence of ILD. A wealth of diagnostic information can be obtained from initial evaluation. History must include a review of environment factors, occupational exposures, medication and drug usage and family medical history.[4] In addition to respiratory symptoms, extra-pulmonary features of associated disease may provide important hints to the correct diagnosis.

Materials and Methods

Study Design: Random prospective study.

Study Duration: January 2021 to January 2022

Study Setting: Patients presenting to Pulmonology Out-Patient Department (OPD) and In-Patient Department (IPD), Care Hospital.

Sample Size: 50 patients

Inclusion Criteria

Patients with clinical history suggestive of interstitial lung disease like cough , SOB on exertion, with or without extra-thoracic manifestations like arthralgias, skin rashes or pigmentation, dry mouth, dry eyes and features suggestive of Raynaud's phenomenon(digital pain and bluish discoloration on exposure to cold) etc.

1. Patients with Velcro rales on respiratory examination suggestive of ILD.
2. Radiological appearance suggestive of ILD.
3. Known cases of Connective Tissue Disease (CTDs) presenting with features suggestive of ILD.

Exclusion Criteria

1. ILD like infections eg. Miliary tuberculosis and Pneumocystis jirovecii pneumonia(PJP).
2. ILD like malignancies eg. Lymphangitis carcinomatosis or Miliary carcinomatosis.
3. Pulmonary Kochs co-existing with ILD.
4. Occupational lung disease.

After obtaining a written informed consent in all these patients, a detailed history was taken and a thorough clinical examination was done as well and then subjected to investigations. A total of 60 patients were enrolled in the study.Out of which 4 patients were excluded based on exclusion criteria and 6 patients were lost to follow up.

All the patients were thoroughly evaluated with respect to history, clinical examination, Spirometry, exercise oxygen desaturation testing, 2D Echo,Laboratory investigations including comprehensive connective tissue disease profile in selected cases, radiological examination and FOB with BAL.

A thorough clinical examination was done in all cases. Any positive findings like clubbing, peripheral lymphadenopathy, abnormal breath sounds, Velcro rales were noted. Examination of the cardiovascular system was done for any loud P2 and features of cor pulmonale. A 2D-ECHO was done in all cases suspected to be having cardiovascular co-morbidity apart from conducting a detailed examination of the respiratory system. Examination of the musculoskeletal system was done for detecting any joint manifestations. Examination of the nervous system, eye and GIT were done to look for any co-existent abnormalities.

Routine lab Investigations were done in all cases including Haemoglobin, Total and Differential counts, blood urea, serum creatinine, AEC, RBS, and a complete urine examination.Serum ACE, calcium and other investigations were done wherever felt appropriate.

Statistical Analysis: Data was analysed after entering into Microsoft excel sheet and frequency tables were calculated and test of significance was applied wherever necessary as the present study is a descriptive study.

Observation and Results

Table 1: Distribution based on Demographics, symptoms and spirometry

Gender	Frequency (n=50)	Percentage
Male	18	36%
Female	32	64%
Age Group (in yrs)		
15-25	2	4%
26-35	6	12%
36-45	13	26%
46-55	16	32%

Conti..Table 1: Distribution based on Demographics,symptoms and spirometry

Gender	Frequency (n=50)	Percentage
56-65	12	24%
66-75	1	2%
Symptoms		
Cough	46	92%
Dyspnoea on exertion	50	100%
Chest pain	4	8%
Arthritis	7	14%
Skin lesions	6	12%
Dysphagia	4	8%
GERD	20	40%
Haemoptysis	2	4%
Spirometry		
Could perform	42	84%
Could not perform or not done	8	16%

Out of the 50 patients in this study, 18 are males and 32 are females. Female predominance was seen with 64% of the study population and 36% are males. In females, ILD occurred most commonly in 36-55 year age group (42%) in males ILD occurred most commonly in the 46-65 year age group (28%) age range is 18 to 70 years.

Amongst the study population, dyspnoea on exertion was the predominant symptom (100%), followed by cough (92%) and GERD (40%).

In some cases where the general condition of the patient is poor and those in acute exacerbations, spirometry was not done.84% of the study population could perform a pulmonary function test and 16% could not perform the same.

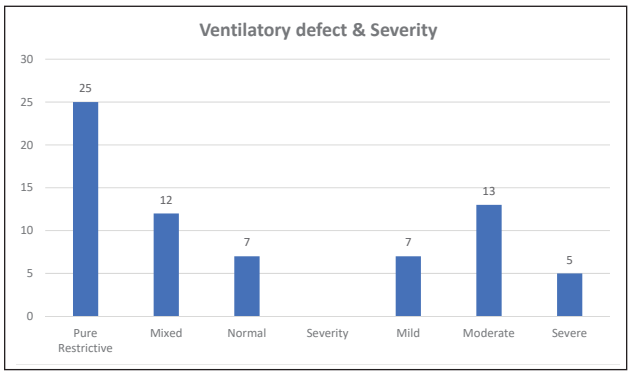

Figure 1: Profile of the ventilatory defect, severity

Restrictive defect was the predominant defect among the patients of the study group (57%) with mixed defect seen in 27%.

Based on the results of the pulmonary function tests, moderate restrictive defect was observed in 52% of the study population. 28% showed a mild restrictive defect.

Table 2: Profile of exercise test performance

Exercise test Result	Frequency (n=50)	Percentage
Complete Test	30	60%
Incomplete Test	10	20%
Did not perform	10	10%
Result		
Positive	22	74%
Negative	8	26%
TIME	SPO 2 (mean)	
PRE-EXERCISE	**95.15%**	
POST-EXERCISE	**88.45%**	

60% of the study population successfully completed the exercise test, while it was incomplete in 20% of the study cases. 10% did not perform the exercise testing.74% of the cases in our study showed a positive exercise test result (desaturation of >4%) and 26% showed a negative test result.

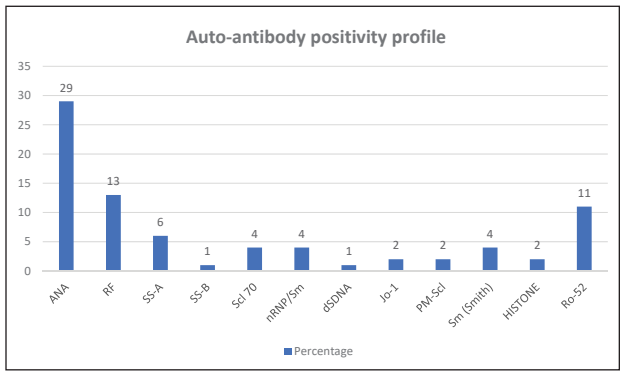

Figure 2: Profile of auto-antibody positivity

Amongst the study population with positive auto-antibodies, ANA was the most frequently encountered auto-antibody (58%), followed by RF (26%) and Ro-52 (22%).

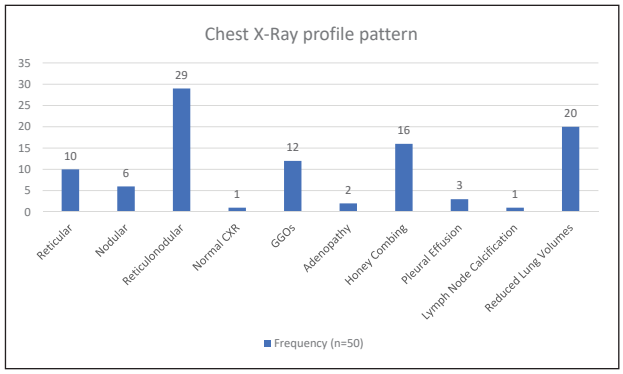

Figure 3: Chest X-ray Profile

Amongst the study population, reticulo nodular opacities were the predominant findings on the chest x-ray (58%), followed by reduced lung volumes (40%), honey combing (32%) and ground glass opacities (28%).

Table 3: HRCT Profile and pattern

HRCT Abnormality	Frequency (n=50)	Percentage
Linear Opacities	30	60%
Nodules	4	8%
Reticular Opacities	32	64%
Ggos	28	56%
Consolidation	4	8%
Honeycombing	26	52%
Cysts	1	2%
Traction Bronchiectasis	20	40%
Mosaic Attenuation	8	16%
Mediastinal Adenopathy	13	26%
Pleural Effusion	3	6%
Pleural Thickening	2	4%
Progressive Massive Fibrosis (Pmf)	2	4%
Emphysema	4	8%
HRCT PATTERN		
UIP	28	56%
NSIP	11	22%
COP	3	6%
AIP	1	2%
LIP	1	2%
OTHERS	6	12%

In the study population, reticular opacities were the most common findings noted on HRCT (64%), followed by linear opacities (60%), ground glass opacities (56%), honey combing (52%) and traction bronchiectasis (40%). UIP pattern was the most frequent noted pattern on HRCT (56%). NSIP and COP constituted 22% and 6% respectively.

Table 4: Aetiology profile

Aetiology	Frequency (n=50)	Percentage
IPF	15	30%
Non-IPF IIPs	4	8%
LDCTD	14	28%
CTD-ILD	8	16%
Pneumoconiosis	2	4%
Pulmonary Alveolar Proteinosis	2	4%
Sarcoidosis	2	4%
Hypersensitivity Pneumonitis	3	6%

IPF was the most common diagnosed interstitial lung disease in our study population (30%), followed by LDCTD (28%) and CTD ILD (16%). MCTD and RA were equally predominant (50% each) amongst those cases in our study who were diagnosed with CTD – ILD.

Discussion

Interstitial lung disease are a heterogeneous group of diseases involving lung interstitium. They have features in common like similarities of symptoms, comparable radiographic appearances, consistent alterations in the pulmonary physiology and typical histological features.In literature, most of the studies on ILD have included miliary tuberculosis, lymphangitis carcinomatosis and tropical pulmonary eosinophilia.[5,6] In the present study infectious and malignant causes of ILD were excluded. In all studies on ILD in the literature, all IIPs were considered as IPF and some studies have further divided IIPs into IPF and non-IPF IIPs. In the present study, IIPs were divided into IPF, non-IPF IIPs and Lung Dominant Connective Tissue Disease(LDCTD).The rationale behind this classification is that in some ILDs, an underlying autoimmune disease is suspected but the patients do not meet the criteria for a final diagnosis of CTD.[7]

Furthermore, in the Sen et al study, the third most common cause of ILD was CTD-ILD (18%),[8] and in our study the CTD-ILD was the second most common cause constituting 16% of the cases. Sarcoidosis was the second most common ILD in Sen et al study (22%) whereas in our present study there were 4% cases of sarcoidosis. In Muhammed Shafeeq K et al study Spirometry showed restrictive abnormality in 64.3%, mixed in 31.4% and normal reports in 4.3%.[9] In our study restrictive abnormality was seen in 50% of the patients, mixed in 24% and normal in 14% of the cases.

In a study by Lima et al, clinical profile of patients, who had lung biopsy were analyzed most common cause was IPF (51.5%) Collagen Vascular Disease – pulmonary fibrosis (15.2%) and Hypersensitivity pneumonitis (9.%).[10]In our study too the most common ILD was IPF and CTD-ILD constituted 16% of the cases. Furthermore, UIP was the most common pattern in IPF and CTD-ILD which concurred with the findings in our study.

In a study done by Vijet al, a group of ILD patients having features of auto immune disease but not meeting the criteria for connective tissue disease were identified and their prevalence and characteristics were determined.Any extra thoracic symptom or sign suggestive of connective tissue disease and a serologic test reflective of autoimmune process, if present was considered as a case of AIF-ILD in this study. AIF-ILD was identified in 32% , IPF in 29% and CTD-ILD in19%.[11] In our study too LDCTD which was analogous to AIF –ILD, constituted 28%, IPF -30% and CTD-ILD-16%, the results very close to those in the study done by Rekha Vij. et al.

In aOlga Tourin et alstudy, Out of 31 patients . A total of 18 cases of IPF were identified out of which 4 cases were LDCTD and a total of 8 cases of NSIP were identified out of which 2 were LDCTD. So, a total of 6 cases of LDCTD were identified in 31 cases which accounted for nearly 20% of the cases.[12] This study bears resemblance to ours. In thepresent study, 14 cases of LDCTD were identified accounting for 28% of the cases because, the study population went through similar investigations like HRCT imaging and screening for auto antibodies like our study group and no surgical biopsy was done.

In a study by Mittoo S et al, unrecognised collagen vascular diseases were identified in a group of ILD patients. Around 15% of collagen vascular diseases were found among this study group.[13] In our study too the CTD-ILD comprised 16% of the cases.

Limitations

- Our study is small and done in a tertiary care hospital of a specific region and so it may not represent the whole population.
- DLCO testing was not done in functional evaluation of ILD patients in our study due to technical issues.
- Biopsy which is a definitive mode of diagnosis in few ILDs, was not done as it was cumbersome, invasive and was not acceptable to most of the patients.
- Occupational lung diseases were not included.

Conclusion

Patients, in their third to fifth decade, especially females, presenting with complaints of breathlessness and cough, with exercise oxygen desaturation, should be evaluated for ILD with complete profile to identify the disease at an early stage.

Ethical Clearance: Ethical Clearance was obtained from the institutional ethics committee of Care Hospital prior to the commencement of study.

Source of funding: Self

Conflict of interest: Nil

References

1. Mapel DW, Hunt WC,Utton R, Baumgartner KB, Samet JM, Coultas DB: Idiopathic pulmonary fibrosis: survival in population based and hospital based cohorts. Thorax2000,53:469-476
2. Coultis DB, Zumwalt RE, Black WC, Sobonya RE: The epidemiology of interstitial lung disease. Am J Respir Crit Care Med 1994; 150: 967-972.
3. Popper MS, Bogdonoff ML, Hughes RL. Interstitial rheumatoid lung disease. A reassessment and review of the literature. Chest 1972;62(3):243–50.
4. Frank ST, Weg JG, Harkleroad LE, et al. Pulmonary dysfunction in rheumatoid disease. Chest 1973; 63(1):27–34.
5. Keane MP, Lynch JP 3rd: Pleuropulmonary manifestations of systemic lupus erythemato-sus. Thorax 2000;55:159–166.

6. Costabel U, Guzman J, Bonella F, et al. Bronchoalveolar lavage in other interstitial lung diseases. Semin Respir Crit Care Med 2007;28(5):514–24.

7. Mittoo S , Gelber AC , Christopher-Stine L , Horton MR , Lechtzin N , Danoff SK . Ascertainment of collagen vascular disease in patients presenting with interstitial lung disease . *Respir Med* . 2009 ; 103 (8): 1152 – 1158

8. Swensen SJ, Viggiano RW, Midthun DE, et al. Lung nodule enhancement at CT: multicenter study. Radiology 2000; 214:73.

9. Shafeeq MK, Nair S, Mathew M, et al. Aetiology and clinicradiological profile of interstitial lung disease in a tertiary care centre. Pulmon 2011;13(1):12-5

10. Lima TRL, Guimarães FS, Silva LA, et al. Relationship between functional capacity, joint mobility and pulmonary function in patients with systemic sclerosis. J Bodyw Mov Ther 2015;19(1):17-24.

11. Vij R, Strek ME. Diagnosis and treatment of connective tissue disease-associated interstitial lung disease. Chest 2013;143(3):814-24

12. Olga Tourin , Charlene D. Fell , Tracy Elliot , Sharon Le Clercq , Marvin Friztler__, Lung-Dominant Connective Tissue Disease" In Patients Evaluated For Interstitial Lung Disease B43. DRUG INDUCED AND CONNECTIVE TISSUE RELATED LUNG DISORDERS. May 1, 2011, A3034-A3034

13. Mittoo S , Gelber AC , Christopher-Stine L , Horton MR , Lechtzin N , Danoff SK . Ascertainment of collagen vascular disease in patients presenting with interstitial lung disease . *Respir Med* . 2009 ; 103 (8): 1152 – 1158