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Spectrum of interstitial lung disease at a tertiary care centre in India

Spectrum śródmiąższowych chorób płuc w trzeciorzędowym ośrodku opieki w Indiach

The authors declare no financial disclosure

Abstract

Introduction: The available data on the epidemiology of interstitial lung disease (ILD) from India is sparse. Hence, the present study was undertaken with the aim to analyse the demographic profile and clinical, radiological and pathological characteristics along with physiological parameters of various subgroups of ILD patients.

Material and methods: We retrospectively studied 289 patients diagnosed with ILD during the years 2001–2013 at one of the respiratory units of Vallabhbhai Patel Chest Institute.

Results: Mean age at presentation was 44.24 years; females comprised 54.68% of the patients. Prior to presentation at our centre, 14.84% patients had been treated with antituberculous therapy due to misdiagnosis of tuberculosis. In the pool of ILDs analysed, sarcoidosis (37.3%) was found to be the most common subgroup, followed by IPF (27.6%) and NSIP (25.6%). Cough (92.97%) was the most common presenting symptom; exertional dyspnoea was found in 79.2% of patients. Digital clubbing was commonest in IPF, found in 30% of patients. Significant desaturation on six-minute walk test was most frequenty seen (50%) in NSIP patients. The most common pattern on chest roentgenogram was reticular/reticulo-nodular pattern (80.2%) and on HRCT — interstitial fibrosis (49.9%). Mean of predicted total lung capacity (TLC) was 64.3%, the lowest being in the IPF group (58.88%). Mean of predicted DLCO was 50.56%, the lowest being in the IPF group (42.75%). The overall diagnostic yield of bronchoscopic biopsy was 83.04%, the highest yield being among sarcoidosis patients (96.29%).

Conclusions: We found sarcoidosis, IPF and NSIP to be the most common ILDs in northern India. ILDs are still frequently misdiagnosed as TB, and increased awareness, education and diagnostic facilities are required to diagnose ILDs at an early stage.

Key words: interstitial lung diseases, diffuse parenchymal lung diseases, idiopathic pulmonary fibrosis, high resolution computed tomography, sarcoidosis

Pneumonol. Alergol. Pol. 2014; 82: 218–226

Streszczenie

Wstęp: Istnieje stosunkowo mało informacji dotyczących epidemiologii śródmiąższowych chorób płuc (ILD) w Indiach. Aktualna praca została podjęta w celu oceny danych demograficznych, kliniczno-patologicznych i fizjologicznych różnych typów chorób śródmiąższowych.

Materiał i metody: Badaniem retrospektywnym objęto 289 pacjentów, u których rozpoznano ILD w latach 2001–2013 w jednym z oddziałów Vallabhbhai Patel Chest Institute (Indie, Delhi).

Wyniki: Średni wiek chorych w chwili rozpoznania wynosił 44,4 roku, kobiety stanowiły 54,68% ogółu chorych. Z powodu mylnego rozpoznania gruźlicy 14,84% chorych było uprzednio leczonych przeciwprątkowo. Sarkoidozę rozpoznano u 37% chorych,

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DOI: 10.5603/PiAP.2014.0029
Praca wpłynęła do Redakcji: 23.12.2013 r.

ISSN 0867–7077

a IPF i NSIP odpowiednio w przypadku 27,6 i 25,6% chorych. Najczęściej zgłaszanymi objawami były kaszel (92,97%) i duszność wysiłkowa (79,2%) a palce pałeczkowate stwierdzono u 30% chorych na IPF. Znamienny spadek utlenowania krwi w 6-minutowym teście chodu najczęściej obserwowano u chorych na NSIP (50%).
Zmiany guzkowo-siateczkowe w obrazie radiologicznym uwidoczniono u 80,2% chorych, a śródmiąższowe włóknienie w badaniu HRCT wykryto u 49,9% chorych.
Średnia wartość zdolności dyfuzyjnej płuc (DLCO) wynosiła 50,56% wartości należnej i była najbardziej upośledzona w grupie chorych na IPF (42,75%). Wartość diagnostyczna biopsji wykonanej podczas bronchoskopii wynosiła 83,04% i dotyczyła szczególnie chorych na sarkoidozę (96,29%)
Wnioski: Autorzy pracy stwierdzili, że w północnych Indiach najczęstszymi chorobami śródmiąższowymi są: sarkoidoza, IPF i NSIP. Śródmiąższowe choroby płuc są w Indiach często rozpoznawane błędnie jako gruźlica, dlatego konieczne jest szkolenie personelu i stworzenie ułatwień diagnostycznych, aby były one trafnie rozpoznane we wczesnym stadium.

Słowa kluczowe: śródmiąższowe choroby płuc, rozlane miąższowe choroby płuc, samoistne włóknienie płuc, tomografia komputerowa o wysokiej rozdzielczości (HRCT), sarkoidoza

Pneumonol. Alergol. Pol. 2014; 82: 218–226

Introduction

The term interstitial lung disease (ILD), or diffuse parenchymal lung disease (DPLD), encompasses a number of clinical disorders that involve the alveolar structures, pulmonary interstitium and small airways [1, 2]. Some of these diseases are benign and self-limiting; others are chronic, progressive, irreversible and even fatal. All ILDs, however, have certain common clinical, radiological, pathological and physiological features that should be recognised.

The available data on the frequency of occurrence of ILDs is sparse [3]. The incidence of ILDs is variable around the world. Literature shows the incidence of ILDs varying from 3.62 per 100,000 person-years in southern Spain [4] to 31.5 per 100,000 person-years in males and 26.1 per 100,000 person -years in females in New Mexico, USA [5], a huge eightfold deviation in incidence across the globe.

In a developing country like India, with a high prevalence of tuberculosis (TB), ILDs are often initially misdiagnosed as TB. Data on ILDs has been limited to just a few dispersed studies [6–14]. The largest ILD series published from India comprised just 274 patients [13]. Also, most of the previous studies on ILD from India lacked any computed tomography (CT) evaluation. This retrospective study was, therefore, undertaken with the aim to study the spectrum of ILDs presenting to a tertiary care centre. The demographic profile and clinical, radiological and pathological characteristics along with physiological parameters of these ILD patients were retrospectively analysed.

Material and methods

This study includes 289 patients diagnosed to have ILDs during the years 2001–2013 at one of the respiratory units at Vallabhbhai Patel Chest Institute, Delhi. The records of the patients were retrospectively reviewed for clinical presentation, and radiological and pathological findings along with their pulmonary function test at presentation. The final diagnosis of ILD was based on histopathology wherever available, other cases were labelled as ILD on the basis of clinical and radiological parameters. The American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of Idiopathic Interstitial Pneumonias 2001 guidelines were used in the diagnosis and classification of ILD [15]. The diagnosis of sarcoidosis was based on compatible clinical, radiological, laboratory and/or histopathological features as per the joint statement of the American Thoracic Society, the European Respiratory Society and the World Association of Sarcoidosis and Other Granulomatous Disorders (ATS/ERS/WASOG) and also exclusion of any other causes of the same [16].

A detailed record of the medical history and examination at the time of initial presentation was analysed. Laboratory investigations such as haemogram, chest radiograph, electrocardiogram and sputum smear examination for acid-fast bacilli (AFB), Mantoux test and pulmonary function test (PFT) were recorded. All serological investigations such as serum anti-nuclear antibody (ANA), serum calcium, serum angiotensin converting enzyme (ACE) levels, cytoplasmic anti-neutrophil cytoplasmic antibodies (c-ANCA), perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA), anti topoisomerase I antibody (Scl-70) rheumatoid factor (RA), anti-cyclic citrullinated peptide antibodies (anti-CCP), anti double-stranded DNA (anti-dsDNA), along with other relevant investigations such as 24-hour urinary calcium records, were obtained.

Chest radiograph and high-resolution computed tomography (HRCT) findings were analysed. Fibre optic bronchoscopy (FOB), trans-bronchial lung biopsy (TBLB), endobronchial biopsy (EBB) and trans-bronchial needle aspiration (TBNA) had been performed in stable patients willing to undergo the procedure.

A six-minute walk test (6MWT) was performed in patients wherever indicated and desaturation of > 4% from baseline was considered as significant. In patients who were either not fit to undergo FOB or refused, the diagnosis was made on the basis of clinical, laboratory and radiological features.

Results

The study included 289 patients diagnosed with interstitial lung disease on the basis of clinical radiological and pathological characteristics. The overall mean age at presentation was 44.24 years, 43.78 years in sarcoidosis and 53.83 years in IPF. Of the 289 ILD cases there were 158 (54.68%) females, and the majority (89.4%) were housewives. The overall mean duration of symptoms at diagnosis was 3.07 years: 2.35 years in sarcoidosis and 2.23 years in IPF. The most frequent presenting symptom in the ILDs was cough, present in 268 (92.7%), followed by exertional dyspnoea in 229 (79.2%), arthralgia/ /arthritis in 32 (11.0%), fever in 30 (10.3%) and skin involvement in 11 (3.8%). Also, haemoptysis was present in 6 (2.07%) subjects. Dysphagia was observed in 5 (1.7%) cases, all belonging to the systemic sclerosis subgroup. The clinical profile disease-wise has been summarised in Table 1. Additionally, a disease-related summary of the radiological and physiological findings of different ILDs in the study has been depicted in Table 2.

On examination, digital clubbing was noted in a total of 41 (14.1%) patients, with 30% in cases of IPF and 17.56% in cases of NSIP. Chest crepitations were present in 273 (94.4%) of all patients. Prior history of anti-tuberculosis treatment due to misdiagnosis as tuberculosis was present in 43 (14.8%) cases of ILD, with it being most common in sarcoidosis (22.22%). Significant desaturation on 6MWT was observed in 97 (33.56%) cases of ILD at presentation, with a frequency of 32.5% in IPF and 22.22% in sarcoidosis. All patients were sputum smear-negative for acid fast bacilli.

Chest roentgenogram revealed reticular/reticulo-nodular pattern in 232 (80.2%) and hilar-adenopathy in 55 (19.03%) patients of ILD. The overall patterns documented on HRCT (n = 289) were interstitial fibrosis (49.9%), honeycombing (37.71%), ground glass opacities (34.25%), intrathoracic lymphadenopathy (20.76%), traction bronchiectasis (17.64%) and pleural fibrosis (5.8%). Honeycombing was present in 4.62% cases of sarcoidosis, 31.08% cases of NSIP and in all IPF cases. Pleural involvement was seen in 12.96% (14 subjects) of sarcoidosis subjects, in 2 subjects of rheumatoid arthritis-associated ILD and in 1 subject of SLE. Nodular pattern was noted in 12.03% (13 subjects) of sarcoidosis subjects and in 57.14% (4 subjects) of hypersensitivity pneumonitis (HSP) subjects. Serological evaluation in cases of sarcoidosis found that the mean level of serum ACE was 82.84 IU/L, serum calcium was 8.8 mg/dL and 24-hour urinary calcium levels were 308.23 mg/day. In cases of rheumatoid disease-associated ILDs, rheumatoid factor was positive in all 7 patients. Systemic sclerosis was diagnosed on the basis of positive Anti Scl-70 antibody in all 5 cases. SLE was diagnosed in one patient with positive anti ds-DNA antibody. Skin biopsy showing granulomatous lesions consistent with sarcoidosis was reported in 3 patients (out of 108 sarcoidosis subjects), while the skin biopsies of 3 other patients had histological features suggestive of systemic sclerosis (out of 6 systemic sclerosis subjects).

Spirometry was done in all cases, followed by static lung volume and diffusion capacity wherever possible. The mean TLC was 64.3% of predicted and DLCO was 50.56% of predicted with the DLCO/VA ratio being 92.9% of predicted. FOB was performed in all 289 patients, and biopsy reports (TBLB and/or EBB) were diagnostic in 240 (83.04%) cases.

Sarcoidosis

Sarcoidosis was diagnosed in 108 (37.37%) subjects in the present cohort. These comprised 45 males and 63 females. The mean age at presentation was 43.78 years, with mean duration of symptoms being 2.35 years. The details of clinical symptoms, radiology, bronchoscopy findings and physiological parameters have been described in Table 1 and 2.

Idiopathic pulmonary fibrosis

Idiopathic pulmonary fibrosis was the second largest subgroup in the ILD subset. This subgroup included 80 (27.68%) patients and had a male-to-female ratio of 1:1. The average age at presentation was 53.83 years and mean duration of symptoms was 2.23 years. The detailed clinical, radiological, pathological and physiological parameters of this subgroup are depicted in Table 1 and 2.

Table 1. Clinical profile of 289 interstitial lung disease patients

	Total N (%)	Sarcoidosis, N (%)	IPF N (%)	NSIP N (%)	HSP N	RA-ILD N	SS N	SLE N	LIP N	DIP N	LCH N	PAP N
Number of subjects	289	108 (37.37)	80 (27.68)	74 (25.6)	7	6	6	1	2	3	1	1
Age (Years)	44.24	43.78	53.83	49.83	45.71	51.66	45.56	40	42	49.3	35	30
Male/Female	131 (45.32)/ /158 (54.68)	45 (41.66)/ /63 (58.34)	40 (50)/40 (50)	34 (45.94)/52 (44.06)	5/2	5/1	1/5	0/1	0/2	1/2	0/1	0/1
Smoking	38 (13.14)	14 (12.96)	11 (13.75)	10 (13.51)	0	1	0	0	0	2	0	0
Duration of symptoms (Yrs)	3.07	2.35	2.23	2.64	2.85	4.5	3.5	3	4.2	3.5	3	2
Cough	268 (92.73)	103 (95.31)	71 (88.75)	72 (97.29)	7	5	3	1	2	3	1	1
Dyspnoea	229 (79.23)	85 (78.70)	55 (68.75)	70 (94.95)	5	4	3	1	2	3	1	1
Haemoptysis	6 (2.07)	2 (1.8)	1 (1.2)	3 (4.0)	0	0	0	0	0	0	0	0
Fever	30 (10.31)	12 (11.11)	10 (12.5)	6 (8.1)	0	0	0	1	1	0	0	0
Joint Symptoms	32 (11.07)	12 (11.11)	7 (8.75)	5 (6.7)	0	7	0	1	0	0	0	0
ATT intake	43 (14.87)	24 (22.22)	9 (11.25)	6 (8.1)	1	1	0	0	1	1	1	0
Clubbing	41 (14.18)	4 (3.7)	24 (30.0)	13 (17.56)	0	0	0	0	0	0	0	0
Desaturation on 6MWT	97 (33.56)	24 (22.22)	26 (32.5)	37 (50.0)	4	3	1	1	0	0	1	0

ATT — anti tubercular therapy, 6MWT — six minute walk test, IPF — idiopathic pulmonary fibrosis, NSIP — non-specific interstitial pneumonitis, HSP — hypersensitivity pneumonitis, SLE — systemic lupus erythematosus, LIP — lymphoid interstitial pneumonitis, DIP — desquamative interstitial pneumonitis, LCH — Langerhan`s cell histiocytosis, PAP — pulmonary alveolar proteinosis, RA-ILD — rheumatoid arthritis associated interstitial lung disease; SS — systemic sclerosis

Nonspecific interstitial pneumonitis

This was the third largest subgroup in the studied cohort, comprising 74 (25.6%) patients; 34 males and 52 females. The average age at presentation was 49.83 years, with mean duration of symptoms being 2.64 years. The detailed clinical history, radiological features, bronchoscopic evaluation and physiological parameters have been described in Table 1 and 2.

The diagnosis of the remaining 27 patients included hypersensitivity pneumonitis (7), rheumatoid lung disease (6), systemic sclerosis (6), desquamative interstitial pneumonitis (3), lymphoid interstitial pneumonitis (2) and one case each of systemic lupus erythematosus, Langerhan's cell histiocytosis (LCH) and pulmonary alveolar proteinosis. All of the findings of these patients have been described in detail in Table 1 and 2.

Discussion

The true burden of ILD in India is not clearly known due to under recognition, attributed to lack of awareness, paucity of diagnostic facilities as well as to the huge spectrum that this entity encompasses. Reports from western literature show an increase in the prevalence and incidence of ILD in recent decades [18]. However, data on clinical presentation and diagnosis of the spectrum of ILDs from India is limited. The results of previous Indian studies on ILDs are summarised in Table 3. The previous studies, depicted in Table 3, did not have uniform diagnostic case definition and classification criteria of ILDs and did not have computed tomography evaluation in all studies. To the best of our knowledge, the present study is the first study of ILD epidemiology on an Indian population, with defined criteria [15, 16] of diagnosis and classification.

Table 2. Radiological findings and pulmonary function test results of 289 Interstitial Lung Disease Patients

	Total N (%)	Sarcoidosis N (%)	IPF N (%)	NSIP N (%)	HSP N	Rheumatoid N	Systemic Sclerosis, N	SLE N	LIP N	DIP N	LCH N	PAP N
CxR findings												
Reticular/reticulo-nodular	232 (80.27)	76 (70.37)	71 (88.75)	67 (90.54)	5	3	5	1	2	2	Cysts	0
Hilar Lymphadenopathy	55 (19.03)	46 (42.59)	0	8 (10.81)	0	1	0	0	0	0	0	0
Honey Combing	23 (7.9)	2 (1.85)	20 (25.0)	1 (1.3)	0	0	0	0	0	0	0	0
HRCT findings												
Fibrosis	143 (49.48)	13 (12.03)	80 (100)	37 (50.0)	0	7	5	1	0	0	0	0
Honeycombing	109 (37.71)	5 (4.62)	80 (100)	23 (31.08)	0	0	0	0	0	1	0	0
Ground glass opacity	99 (34.25)	36 (33.33)	2 (2.5)	49 (66.26)	7	0	1	0	2	1	0	0
Interstitial Infiltrates	113 (39.10)	43 (39.81)	0	56 (75.67)	5	5	0	1	2	1	0	1
Subpleural Opacity	110 (38.06)	11 (10.18)	80 (100)	19 (25.67)	0	0	0	0	0	0	0	0
Traction Bronchiectasis	51 (17.64)	3 (2.7)	34 (42.5)	9 (12.16)	0	1	4	0	0	0	0	0
Lymphadenopathy (H/M)	60 (20.76)	46 (42.59)	2 (2.5)	12 (16.21)	0	0	0	0	0	0	0	0
Pleural Opacity	17 (5.80)	14 (12.96)	0	0	0	2	0	1	0	0	0	0
Nodules	17 (5.80)	13 (12.03)	0	0	4	0	0	0	0	0	0	0
Others	–	–	–	–	–	–	–	–	Cysts	–	Cysts	Crazy paving
FOB Biopsy positive	240 (83.04)	108 (96.29)	60 (75)	72 (97.29)	7	5	4	1	2	3	1	1
FEV_1% (mean % predicted)	61.02	66.14	59.1	61.64	75.53	57.83	53.5	43	53.5	68	68	65
FVC% (mean % predicted)	64.41	72.71	61.12	67.24	79.53	67.82	51.66	52	52.5	69	71	64
FEV_1/FVC% (mean)	90.32	85.37	90.52	89.65	90.57	80.17	95.5	71	87.5	95.33	108	100
TLC% (mean % predicted)	64.3	70.77	58.88	61.98	71.6	63.33	63.16	59.2	60	80.33	ND	56
DLCO% (mean % predicted)	50.56	64.15	42.75	50.5	53	42.56	37.56	48	54.5	66.62	ND	45
DLCO/VA% (mean % predicted)	92.9	99.46	85.3	90.26	99.5	74.33	85.4	94	88.5	122.23	ND	90

CXR — chest x-ray, HRCT — high resolution computed tomography,H/M–Hilar/Mediastinal, FOB — fibre optic bronchoscopy, FEV_1 — forced expiratory volume in 1^{st} sec, FVC — forced vital capacity, TLC — total lung capacity, DLCO — diffusion capacity of carbon monoxide. VA — alveolar volume, IPF — idiopathic pulmonary fibrosis, NSIP — non-specific Interstitial pneumonitis, HSP — hypersensitivity pneumonitis, SLE — systemic lupus erythematosus, LIP — lymphoid interstitial pneumonitis, DIP — desquamative interstitial pneumonitis, LCH — Langerhan`s cell histiocytosis, PAP — pulmonary alveolar proteinosis

Table 3. Comparison of Different Interstitial Lung Disease Studies from India

Reference	Age range	M/F	Smoking history N (%)	Duration of symptoms (%)	Cough (%)	Dyspnoea (%)	Joint Symptom (%)	Skin involvement (%)	Clubbing (%)	CXR					HRCT			Serology (%)	IPF/CFA N (%)	Sarcoidosis N (%)	Connective tissue disease – ILD (CTD–ILD) N (%)	Others N (%)
										Reticular (%)	Reticulonodular (%)	Nodular (%)	Honeycombing (%)	Ground glass opacity (%)	Ground glass opacity (%)	Honeycombing (%)	Fibrosis (%)					
[7] n = 61	30 yrs–60 yrs	26/35	NA	< 6 m – 24.6 6–12 m – 20.4 1–3 yrs – 30.4 3–5 yrs – 24.6	65.60	100	61.90	42.30	73.20													
[8] n = 161	< 25 yrs – 10% ≤ 45 yrs – 46% > 45 yrs – 44%	86/75	NA	< 1 yr – 30: 1–2 yr – 18: 3–5 yr – 24: > 5 yr – 28	82.00	100	2.40	NA	53.00	62.3	16.4	6.6	14.7	–	The percentage of different radiological patterns were not mentioned			RA factor – 33.3 LE cell phenomenon – 11.1 ANA – 7.4	28 (46)	–	31 (50.8)	2 (3.2)
[12] n = 73	IPF – 53 ± 10 yrs Secondary DPLD – 45 ± 11yrs	22/51	11 (15.06)	18 – 27 m	56.16	72.60	NA	NA	41.03	8	–	–	38	–	The %age of different radiological patterns were not mentioned			RA factor – 1.2	34 (53.9)	2 (3.1)	6 (9.5)	21 (33.3)
[13] n = 273	Mean = 48 yrs	91/182	NA	2 m – 5 yrs	100	100	NA	NA	NA	The percentage of different radiological patterns were not mentioned					50.68	38.35	10.95	RA factor – 24.65 ANA – 10.95	33 (45.20)	7 (9.58)	33 (45.20)	–
[14] n = 30	Mean = 43.57 yrs	20/10	NA	< 6 m – 23.31 6–12 m – 23.31 1–3 yr – 33.33 3–5yr – 21.05	43.29	100	16.65	6.66	50	–	–	–	–	82	90	43	–	NA	117 (43)	61 (22)	51 (18)	44 (16.11)
Present study (2013) n = 289	Mean = 44.24yrs	31/158	38 (13.14)	3.07 yr	92.73	79.23	11.07	3.80	14.18	16.67	60	–	13.32	10	The %age of different radiological patterns were not mentioned			NA	The subgroups have not been mentioned			
										80.27		–	7.9	–	49.98	37.71	67.62	RA factor – 2.4 ANA – 7.60	80 (27.68)	108 (37.3)	13 (4.4)	88 (30.4)

Four Indian studies [Reference 6, 9, 10, 11] have not been included in this table as No Information was available for most of the parameters mentioned in the table.
Number in superscript indicates reference ^ The study did not mention the percentage for different parameters; NA — not available in the study; *Different radiological terms/definitions used in different studies; HRCT — high resolution computed tomography; RA — rheumatoid factor; ANA — antinuclear antibody; IPF/CFA — idiopathic pulmonary fibrosis/cryptogenic fibrosing alveolitis

In our study, the mean age at presentation was greater than 40 years, with the exception of ILDs presenting at a younger age, such as LCH and PAM. This finding is similar to previous studies from India [6–14] as well as western literature [17, 18]. The present study observed increased prevalence in females (54.6%) as compared to male patients (45.3%). Similar observations have been reported in other Indian studies [7, 9, 12, 13] and also in a study from Greece. However, an increased prevalence in males has been documented in other studies [8, 10, 11, 14, 19]. This can be explained by the fact that the majority of our subject population consisted of patients with sarcoidosis and non-specific interstitial pneumonia (NSIP), which are female preponderant diseases. In the pool of ILDs analysed, sarcoidosis (37.3%) was found to be the most common subgroup, followed by IPF (27.6%) and NSIP (25.6%). The results were similar to another Indian study [10] and studies from western literature [20, 21]. However, in a study on the occurrence of ILD in Poland based on patients hospitalised in the Regional Pulmonary Unit in Radom, IPF (27.5%) was the most common, followed by sarcoidosis (25%). The incidence of ILDs calculated for the adult population of this region was 5/100,000 [22]. Similarly, studies by Subhash et al. [12] and Udwadia et al. [13], from India, reported a higher prevalence of IPF in the study population.

Another important observation is that almost 15% of cases of ILDs had a history of anti-tubercular treatment, and in the sarcoidosis subgroup this figure was 22%. This might be due to radiological similarities between ILD and pulmonary tuberculosis and a lack of awareness and paucity of diagnostic facilities in remote areas.

The current study included 108 (37.3%) cases of pulmonary sarcoidosis, 63 (58.3%) being females. The higher prevalence in females is coherent with findings in western literature [21]. The mean age at presentation was 43.78 years with the average duration of symptoms being 2.35 years; the majority were non-smokers (78.1%). This data is similar to other Indian studies [23, 24]. In accordance with the literature [21], clubbing (3.7%) was a rare finding in our study. Miliary pattern is an uncommon finding [25], although in the current study it was found in 12.03% of cases. 6MWT showed significant desaturation in 24 (22.22%) cases. The plausible explanation of this could be the advanced stage of the disease at presentation. In bronchoscopic evaluation, only 5 cases of sarcoidosis had visible mucosal nodules. The diagnostic yield of bronchoscopic biopsy was 96.29%, similar to that observed in literature [26].

Idiopathic pulmonary fibrosis is a specific type of ILD, with characteristic radiological features and histopathology. In the present study we had 80 (27.68%) cases of IPF, and it was the second most common subgroup in the pool of ILDs. In contrast, IPF was observed as the most common ILD in other Indian [11–13] and western studies [27, 28]. In the current study, mean age at presentation was 53.83 years, male to female ratio was 1:1 and only 13.75% of cases were smokers. The current study agrees with data from western literature [29–32] in terms of age at presentation with disease typically occurring in 6^{th}–7^{th} decade of life. The literature [31–34] shows more men being diagnosed with IPF than women and the majority being smokers. However, our study reported equal prevalence in male and female subjects and a lesser prevalence of smoking. In another Indian study by Subhash et al. [12], out of 33 cases of IPF, 16 were females and smoking was present in only 18% of all IPF cases. Another point that merits mention is that diagnostic criteria vary across studies, leading to differences in epidemiological parameters of IPF.

In our study, on 6MWT, 32.5% of cases showed significant desaturation (> 4%) at presentation. This finding has clinical implications as studies have advocated that desaturation (i.e. a decline in oxygen saturation to below 88%) during 6MWT is a marker for increased risk of mortality [35]. In the current study, among the ILD subgroups, subjects of IPF showed worse pulmonary function test results with mean TLC of 58.88% of predicted and DLCO 42.75% of predicted. A similar finding was observed in a study by Jindal et al. [7]; the cryptogenic fibrosing alveolitis (CFA) group (synonymous with idiopathic pulmonary fibrosis) [36] showing DLCO of 49.8 ± 15.9% of predicted. Studies [37, 38] have shown that DLCO is a reliable predictor of survival at baseline, and a threshold of approximately 40% of predicted value has been correlated with an increased risk of mortality.

In the present study, we had 74 (25.6%) cases with a diagnosis of NSIP based on clinical radiological and pathological features, but we were unable to elucidate the cause. The mean age at presentation was 49.83 years, 52 were females and only 13.5% were smokers. The review of literature shows NSIP has a mean age of 52 years and is more common in females and never smokers [39].

Patients diagnosed with connective tissue disease (CTD)-ILD may have a classifiable CTD

at the time of diagnosis of ILD; however, in up to 25% of cases clinical and serological findings suggest, but are not entirely diagnostic of, a classifiable CTD [40]. Of 13 cases diagnosed as CTD -associated interstitial lung disease (CTD-ILD), lung involvement at presentation was observed in 6 cases each of rheumatoid arthritis (RA) and scleroderma; only 1 case of SLE was noted. The prevalence of RA-ILD varies from 5–58% [40] and ILD in systemic sclerosis is observed in 40–80% of cases [41, 42]. The prevalence of CTD–ILD in India has been reported as ranging from 5.6% to 50.8% in various studies [7, 10].

Hypersensitivity pneumonitis (HSP) was diagnosed in 7 (2.03%) cases; all were associated with pigeon exposure, with duration of exposure ranging from 3–10 years. All 7 cases had histopathological confirmation of features consistent with diagnosis of HSP. In a previous study from India, Udwadia et al. [13] reported HSP in 15 (6%) from a total of 273 cases. Other ILDs diagnosed as per clinico-radio-pathological criteria in the current study were desquamative interstitial pneumonia (DIP) 3 cases, lymphocytic interstitial pneumonia (LIP) 2 cases and one case each of pulmonary Langerhans cell histiocytosis and pulmonary alveolar proteinosis.

Conclusions

The current study describes the spectrum of ILDs prevalent in northern India. Diagnosis of ILDs at an early stage is paramount to prevent/ /delay progression to irreversible damage to the lungs, especially in treatment-responsive ILDs like sarcoidosis. Hence, in a developing country like India, with high prevalence of pulmonary tuberculosis, education and awareness of general practitioners and physicians about ILDs deserves special attention.

Conflict of interest

The authors declare no conflicts of interest.

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