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Interstitial Lung Disease: A Tertiary Care Center Experience

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Abstract:
Objective: *To evaluate the clinical profile of Diffuse Parenchymal Lung Diseases with reference to the presenting symptoms and clinical course in a tertiary care teaching institution. The diagnosis of specific form of interstitial lung disease and management of the same to achieve stabilization of the disease is quite challenging to the clinicians. The present study was undertaken in view of the paucity of clinical data on Interstitial lung disease (ILD).*
Materials and Methods: *A prospective observational study of patients with the diagnosis of interstitial lung disease over a period of two years in a tertiary care center. Patients with complaints of cough and breathlessness showing radiological features suggestive of interstitial lung disease were included in the study*
Results: *26 patients were diagnosed as having interstitial lung disease during the study period. Idiopathic pulmonary fibrosis (IPF) was the most common finding.*
Conclusion: *Lack of recognition at an early stage lead to delayed diagnosis in most of the patients.*
Keywords: *disease,fibrosis,interstitial,idiopathic,lung, pulmonary*

I. Introduction

Interstitial lung disease is a heterogeneous group of lung diseases which are challenging to the clinicians. Interstitial lung diseases are also referred to as diffuse parenchymal lung diseases. [1] The data available on the epidemiology of interstitial lung disease is relatively limited. This was a prospective study undertaken to analyse the demographic profile, clinical and radiological characteristics of the patients with interstitial lung disease in a tertiary care setting. The understanding of the distinct appearance of the diffuse parenchymal lung diseases in high resolution computed tomography (HRCT) has greatly reduced the need for biopsy of the condition.

II. Materialsand Methods

Twenty six patients with chief complaints of dyspnea and cough with chest radiograph and HRCT thorax suggestive of interstitial lung disease were included in the study. Patients with ILD secondary to malignancy were excluded from the study. The relevant data included age, sex, occupation, presenting complaints, smoking status, environmental exposures, history of anti-tuberculous treatment, connective tissue diseases, family history and physical examination findings. All the patients with suspected ILD were subjected to HRCT thorax, spirometry and diffusion capacity for carbon monoxide. Contrast study was done in patients suspected to have mediastinal lymphadenopathy. Six minute walk test was performed whenever patient's condition permitted for the same. Baseline 2D echocardiogram was done in all the patients. The Antinuclear antibody (ANA) profile, Rheumatoid factor (RF), Anti –cyclic citrullinated peptide (Anti-CCP), Antinuclear cytoplasmic antibody (ANCA) and serum Angiotensin converting enzyme were estimated wherever it was indicated.

Diagnosis of IPF was done after excluding the known causes and the HRCT findings suggestive of UIP pattern based on the current guidelines of the American Thoracic Society/European Respiratory Society. Bronchoscopy, Broncho alveolar lavage and transbronchial lung biopsy was done in only one patient.

STATISTICS: Data were analysed using SPSS version 18. Descriptive statistics of death and all relevant data were collected. Chi square test was used to compare the differences of death rate between male and female patients.

Institutional Ethical committee clearance was taken for the study.

III. Results

Total number of 26 patients with diffuse parenchymal lung disease were enrolled in the study. The mean age of the patients was 66 years. Less than 50 years age group was 3 (11.5%), 50 and above age group was 23 (88.5).The male: female ratio was 0.85:1(STD 11.983). Six patients died during the study period, male were 5 (41.7%) and female was 1 (7.1%).The difference was found to be statistically significant ($P < 0.05$). The

chief complaint of all the patients was dry cough (100%) and progressive dyspnea (100%).The duration of the symptoms varied from 20 days to 10 years before the diagnosis of interstitial lung disease.

Table 1 shows the clinical symptoms of the patients

Clinical signs included clubbing in 2 patients (7.7%) and 25 patients (96%) had bibasilar "Velcro" crackles. 2D echocardiogram showed pulmonary artery hypertension in 7 (26.92%) patients.

Based on the clinical parameters and the x-ray features suggestive of interstitial lung disease, patients were subjected to high resolution CT scanning of thorax with or without contrast. Table 2 shows the findings on HRCT thorax.

In our analysis, Idiopathic pulmonary fibrosis was the most common variety (80.76%) as shown in Table 3.

Table 3 shows the diagnosis.

Majority of the IPF patients were diagnosed with an end stage disease. Fig.1 shows extensive honeycombing.

Significant number of patients had associated co-morbidities as shown in Table 4. Three patients had rheumatoid arthritis prior to the development of ILD. One patient was diagnosed to have rheumatoid arthritis while undergoing evaluation for ILD. Rheumatoid arthritis patients were given disease modifying drugs.

Twenty-four patients performed spirometry in which twenty showed restrictive pattern and four were normal. Thirteen patients performed DLCO and eleven patients had reduced value. Five patients were able to perform the 6 minute walk test .Three patients could complete the test and had desaturation during the test. Remaining 2 could not perform in view of breathlessness. Disease severity was assessed using the GAP index, a clinical prediction model based on sex, age and lung physiology which was validated previously in IPF patients.

Table 5 shows the GAP Index.

IV. Figures and Tables

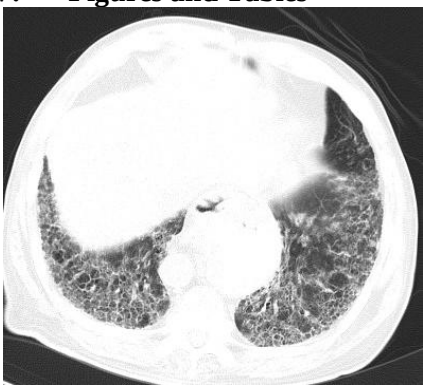

Figure 1. High-Resolution Computed Tomography Thorax Showing Bilateral Honeycomb Pattern.

Table 1. Incidence Of Symptoms In ILD

SYMPTOMS	No of patients presented	Percentage(%)
Breathlessness	**26**	**100**
Cough	**26**	**100**
Fever	**1**	**3.85**
Chest pain	**1**	**3.85**
Joint pain	**3**	**11.53**
Weight loss	**8**	**30.77**

Table 2: Radiological Patterns

Honey comb pattern	19	73%
Ground glassing	14	53.84%
Septal thickening	5	19.23%
Traction bronchiectasis	4	15.38%
Bronchiolectasis	2	7.7%

Table 3. Diagnosis

Interstitial pulmonary fibrosis	21	8 0.76%
NSIP	2	7 .7%
HP	1	3 .8%
COP	1	3 .8%
Sarcoidosis	1	3 .8%

Table 4. Comorbidities

Hypertension (HTN)	2	7.7%
Diabetes (DM)	2	7.7%
HTN+DM+ Coronary artery disease(CAD)	1	3.84%
HTN+CAD	1	3.84%
HTN+DM	3	11.53%
Thyroid disease	6	23.07%

Table 5. Gap Index

Total no of Patients – 26

GAP Index	Patients (n)
0-3	18
4-5	8
6-8	0

V. Discussion

Interstitial lung disease (ILD) represents a group of about 200 distinct disorders involving lung parenchyma. ILD is also termed as Diffuse parenchymal lung disease (DPLD) and classified broadly into idiopathic interstitial pneumonia (IIP) and other than IIP. [2] In 2002, the American Thoracic Society/European Respiratory Society (ATS/ERS) guidelines classified idiopathic interstitial pneumonias (IIPs) into seven specific entities and offered standardized terminology and diagnostic criteria. In the revision of the IIP classification by ATS/ERS in 2013 major IIPs are distinguished from rare IIPs and unclassifiable IIPs. [2] According to ATS guidelines, any patient with IIP a multidisciplinary diagnosis involving close communication between clinician, radiologist and when appropriate pathologist is required. Clinical data and radiological findings are essential for multidisciplinary diagnosis. [2] The need for histological diagnosis was changed to a multidisciplinary approach. Patients with ILD must undergo complete evaluation to establish specific form of ILD. Treatment and management decisions may vary according to the specific form of ILD. An accurate diagnosis of ILD needs a thorough history elicitation including the past medical, social, family and occupational histories.

In the present study most of the patients were above 50 years of age which was observed in other studies as well. [3] Female patients outnumbered male patients. Similar observation was done in the study by Jindal et al. [4] The commonest presenting symptoms were dry cough and breathlessness in the present study and the same has been observed in other Indian studies. [5] HRCT is considered as a standard procedure during the initial evaluation of all patients with ILD.[6] It is a useful diagnostic tool for IPF without biopsy.[7]

Idiopathic pulmonary fibrosis was the most common diagnosis with 21 patients (male 10, female 11) and the same has been reported in other studies.[8] Honeycombing was observed in 43% of the patients by Sen &Udwadia[5] and 73% in our study. IPF is a chronic fibrosing interstitial pneumonia of unknown cause. It occurs mainly in older adults. [10] Christopher et all [11] have used the ILD-GAP model for predicting survival across chronic ILD. We could not apply the GAP model to predict the disease specific survival estimation because of small sample size in our study.

IPF is a disease with poor prognosis with limited therapeutic options. [12]

Treatment with anti-inflammatory /immunosuppressive drugs for any form of ILD has not been validated in placebo controlled clinical trials. However, there is reasonable evidence that administration of agents such as corticosteroids is associated with improvements for cryptogenic organizing pneumonia and cellular nonspecific interstitial pneumonia. [13]

In this study we have given treatment in the form of corticosteroid, immunosuppressant therapy and home oxygen. All the patients received N-acetylcysteine. Aim was to formulate an appropriate management plan that can relieve symptoms and improve quality of life. [13] There is no standard treatment for ILD with extensive fibrosis. Pirfenidone,a novel antifibrotic and anti-inflammatory drug has shown favorable benefit risk profile in some studies.[14] Some clinical trials have shown that pirfenidone may have a significant impact on

progression of disease versus placebo.[14][15] In our study 3 patients were given pirfenidone and one each survived for 12 and 18 months after starting treatment. The third patient is doing well and is on regular follow – up. Patients with IPF and /or non-IPF ILD can benefit from detection and treatment of various associated co-morbid conditions. Patients with secondary pulmonary hypertension were treated with sildenafil. A recent analysis of the data shows that sildenafil therapy may benefit a subset of patients with right heart dysfunction. [16] Gas exchange evaluation at rest and with ambulation 6 minute walk test using pulse oximetry helps in early oxygen therapy. A decrease of 4 or 5 absolute percentage points or greater with exercise is generally considered as significant. [6] Supportive care in the form of oxygen therapy and pulmonary rehabilitation can have an impact on quality of life.[13] Interstitial lung disease associated with connective tissue disease has been proven to have a better long term survival rate than IPF.[17] In our study 3 patients with rheumatoid arthritis and ILD are on disease modifying drugs .

VI. Conclusion

The majority of the patients in the present study were diagnosed as ILD when they had extensive fibrosis. Interstitial lung disease is under-diagnosed because of lack of awareness among physicians. A limitation of this study is the lack of Broncho alveolar lavage and transbronchial lung biopsy data. This was due to the late presentation of the patients. Lack of recognition at an early stage lead to delayed diagnosis in most of the patients.This stresses on the importance of taking detailed history and clinical evaluation with appropriate imaging modalities to make an early specific type of diagnosis of ILD.

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References

[1]. American Thoracic Society, European Respiratory Society. American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias. Am J RespirCrit Care Med.2002;165:277-304.
[2]. Travis WD, Costabel U, Hansell DM et al. An official American Thoracic Society/European Respiratory Society statement: Update of the international multidisciplinary classification of the idiopathic interstitial pneumonias. Am J RespirCrit Care Med.2013;188:733-48.
[3]. Palat P, Parmar HD. Recent trends in interstitial lung disease: Study of 25 cases. Int J Med Sci Public Health.2013; 2(1): 52-55.
[4]. SK Jindal et al. Fibrosingalveolitis- a report of 61 cases seen over past 5 years. Indian journal of chest disease and allied science.1979;21: 174.
[5]. Sen T,UdwadiaZF.Retrospective study of interstitial lung disease in a tertiary care centre in India.Indian J Chest Dis Allied Sci.2010 Oct-Dec;52(4):207-11
[6]. Raghu G, Brown KK. Interstitial lung disease: Clinical evaluation and keys to an accurate diagnosis. Clin Chest Med 2004;25:409419.
[7]. ElliotTL, Lnch DA, Newell JD Jr et al. High resolution computed tomography features of nonspecific interstitial pneumonia and usual interstitial pneumonia. J Comput Assist Tomogr.2005;29:339-345.
[8]. Tiyas Sen and Zarir F. Udwadia.Retrospective Study of Interstitial Lung Disease in a Tertiary Care Centre in India.The Indian Journal of Chest Diseases & Allied Sciences. 2010;52.
[9]. SomenathKundu, SubhraMitra, JoydeepGanguly et al.Spectrum of diffuse parenchymal lung diseases with special reference to idiopathic pulmonary fibrosis and connective tissue disease: An eastern India.2014; 31: 354-360
[10]. Raghu G, Collard HR, Egan JJ et al. An official ATS/ERS/JRS/ALAT statement: idiopathic pulmonary fibrosis: evidence-based guidelines for diagnosis and management**.** Am J RespirCrit Care Med.2011;183**:**788-824.
[11]. Ryerson CJ, Vittinghoff E, Ley B, et al. Predicting Survival Across Chronic Interstitial Lung Disease Chest 2014;145(4):723-728.
[12]. Nathan SD, Shlobin OA, Weir N et al.Long-term course and prognosis of idiopathic pulmonary fibrosis in the new millennium.Chest. 2011;140:221–229.
[13]. Keith C Meyer. Diagnosis and management of interstitial lung disease. Translational Respiratory Medicine 2014, **2**:4
[14]. Noble PW, Albera C, Bradford W.Z et al. Pirfenidone in patients with idiopathic pulmonary fibrosis (CAPACITY): Two randomisedtrials.TheLancet. 2011;377(9779):1760-1769.
[15]. Taniguchi H, Ebina M, Kondoh Y et al. Pirfenidone in idiopathic pulmonary fibrosis. EurRespir J.2010;35:821-829.
[16]. Han MK, Bach DS, Hagan PG et al. Sildenafil preserves exercise capacity in patients with idiopathic pulmonary fibrosis and right-sided ventricular dysfunction. Chest. 2013; 143:1699-1708.
[17]. Vincent Cottin. Interstitial lung disease.EurRespir Rev.2013;22: 127 -32