ORIGINAL ARTICLE

FREQUENCY OF POSITIVE FINDINGS ON CHEST X-RAY OF PATIENTS PROVED TO HAVE IDIOPATHIC PULMONARY FIBROSIS ON HIGH RESOLUTION COMPUTED TOMOGRAPHY (HRCT)

Ramish Riaz

Department of Radiology, Rawalpindi Medical College, Rawalpindi, Pakistan.

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ABSTRACT

BACKGROUND: Idiopathic Pulmonary Fibrosis is a chronic pulmonary disorder characterized by thickening of inner lining of lungs, fibrosis of interstitium. Follow up scans are needed during long treatment procedure which create excessive radiation burden on patient. X-rays can be used for this purpose. Disease carry poor prognosis. Early detection could help in early treatment of disease. M**ATERIALS &METHODS:** X-rays of 30 diagnosed cases of idiopathic pulmonary fibrosis on HRCT were investigated. **RESULTS:** Out of 30 patients 27/30 (90%) had positive finding on chest X ray. **CONCLUSION:** 90% of the patients had positive x-ray finding. Thus X-Ray being widely available plays an important role in diagnosing idiopathic pulmonary fibrosis and for follow up scans.

Introduction

Idiopathic Pulmonary Fibrosis also known as cryptogenic fibrosing alveolitis is a chronic pulmonary disorder characterized by thickening of inner lining of lungs, fibrosis of interstitium. By definition, the cause of pulmonary fibrosis remains unknown.[1] According to American Thoracic Society / European Respiratory Society IPF/UIP (usual interstitial pneu-monia) is the most common of all interstitial pneumo-nias with prevalence of 13 in 100000 of general population.[2] Disease shows very vague onset symptoms which make it challenging for physician to diagnose at early stage. Earliest sign of disease is exertional dyspnea and non productive cough. Many other cardiac and pulmonary disorders also presents with same symptoms.[3] Some patients present with other symptoms i.e. weight loss, low grade fever,fatigue, arthralgias or myalagias. 5% of the patients remain asymptomatic at the time of diagnosis.[4]

Correspondence : Mr. Ramish Riaz
Department of Radiology,
Rawalpindi Medical College,
Rawalpindi, Pakistan.
Email: ramish_exclusive@hotmail.com

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The etiology of pulmonary fibrosis still remained unidentified, however many environmental and occupational factors are proposed to play role in pathogenesis. Exposure to contaminants like asbestos, coal dust,silica,smoke may lead to initiate alveolar damage.[5] Different diseases thought to be involved in pathogenesis of idiopathic pulmonary fibrosis include gastro esophageal reflux disease (GERD), pulmonary infections and chronic aspiration.[6]

Despite extensive investigations, etiopathogenesis of IPF remains enigmatic. However current hypothesis regarding its development propose that it is an epithelial fibroblastic disease in which several, minute, focal and isolated episodes of epithelial injury are followed by pathologic fibrotic repair mechanism.[7] Disease starts with exposure to unknown stimulants which results in nonspecific insults to both epithelial barrier and pulmonary parenchyma resulting in diffuse epithelial cell proliferation and aberrant epithelial cell repair.[8]

Idiopathic pulmonary fibrosis tends to have a poor

prognosis. Up till now no proven effective drug therapy is available for the cure of disease beyond lung transplantation. Recently by advances in understanding the pathogenesis of disease, instead of eliminating disease paradigm is shifted towards slowing or preventing progression of this chronic fibrotic illness. Pirfenidone (5-methyl-1-phenylpyridin-2[1H]-one) is a novel antifibrotic, anti inflammatory drug that has been demonstrated to slow down the disease progression.[9] Other drugs used for controlling Idiopathic Pulmonary Fibrosis are acetyl cystiene, azathioprine,[10] thalidomide[11] and intedanib.[12]

Investigations

Diagnosing Idiopathic pulmonary fibrosis at early stage remains a challenge. The most accurate and sensitive imaging technique to investigate IPF is High Resolution Computed Tomography (HRCT).[13] Other tests include Chest X rays, pulmonary function test, bronchoalveolar lavage etc.

HRCT:

High Resolution Computed Tomography is a protocol performed via Computed Tomography Scanner to detect interstitial lung disease. It is the best available diagnostic test till now for diagnosing idiopathic pulmonary Fibrosis.

According to UCSF ILD program Protocol, a narrow slice width of 1-2 mm is used, with field of view minimized and exposure factors set to enhance spatial resolution. Patient is usually in supine position but prone position is preferred for screening purposes as it clearly shows the bases of lung from where the disease starts.

Idiopathic pulmonary fibrosis on HRCT is characterized by presence of patchy, predominantly peripheral, sub pleural, bibasilar reticular opacities with or without associated traction bronchiectesis.[14] Sub pleural honeycombing less than 5 mm is also an important finding. It appears as clustered cystic air spaces with density equal to that of air.[15] Ground glass opacities are also common finding. But it must be differentiated from nonspecific interstitial pneumonia (NSIP), chronic hypersensitivity pneumonitis,or desquamative interstitial pneumonia (DIP) as it is more common in them.[16]

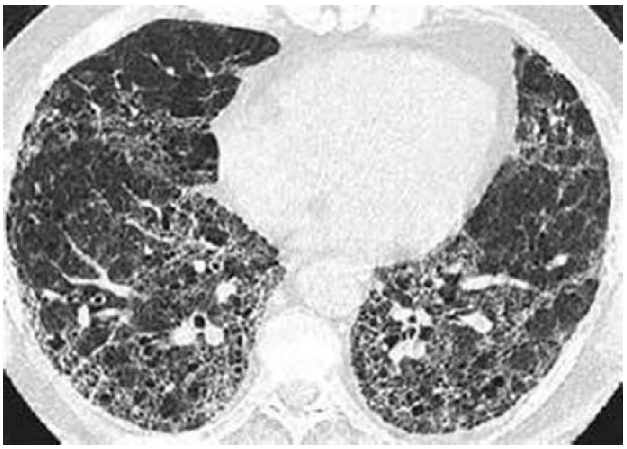


Figure 1: HRCT of an IPF patient

Chest X ray:

Chest radiograph employs ionizing radiation to create the image of chest. Typical radiation doses for an adult scan is around 0.02 mSv (2 mrem) for anterior (PA) projection and 0.04 mSv (4 mrem) for lateral projection.[17]

Although the Chest X ray is not diagnostic for Idiopathic pulmonary fibrosis, however almost all the patients have abnormal radiographic finding at the time of presentation.[5] Presence of bibasilar peripheral reticular opacities (curvilinear or net like linear densities), honey combing are suggestive of Idiopathic pulmonary fibrosis. Lower lobe volume loss could also be seen. Reticular abnormality must be distinguished from coarse linear scarring which appear as a result of previous pneumonia or infarction.[18] (Fig. 2) represents typical findings observed on CXR.[18]

OTHER TESTS:

Pulmonary Function Testing: The main finding on pulmonary function testing of IPF patients is reduced diffusion capacity for carbon monoxide and restrictive ventilatory defect. Pulmonary function testing aids in assessing disease severity and prognosis, However it lacks diagnostic specificity thus it must be used in conjunction with clinical, radiological and pathological information to ensure accurate diagnosis of IPF.[19]

6-Minute Walk Test: 6MWT is used for the clinical assessment of patients of idiopathic pulmonary fibrosis. It shows the functional exercise capacity and is independent prognostic factor. Desaturation below

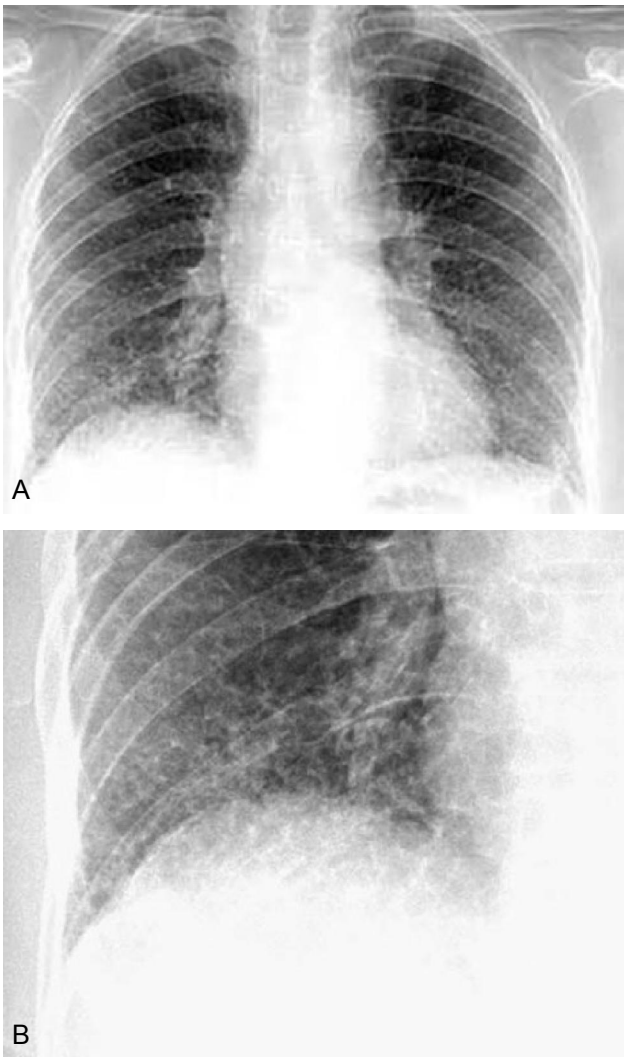


Figure 2A & B: CXR (PA & Magnified view) of an IPF patient

88% during the test is associated with increased mortality.[20]

Bronchoalveolar Lavage: Bronchoalveolar lavage (BAL) cellular analysis is immensely useful adjuvant test in those lacking confidant diagnosis on HRCT. Increased amount of neutrophils and eosinophils are found in IPF patients. A cut level of 30% lymphocytosis in BAL fluid helps in distinguishing IPF from non specific interstitial pneumonias.[21]

Laboratory Studies: Results from routine laboratory studies are nonspecific for the diagnosis. Some patients have shown positive results for antinuclear anti bodies or rheumatic factor however, titers are not generally high.

Surgical Lung Biopsy: Surgical lung Biopsy helps in establishingthe firm clinicopathologic diagnosis especially with those having inconsistent UIP pattern on HRCT. It helps in classification of HRCT.[22]

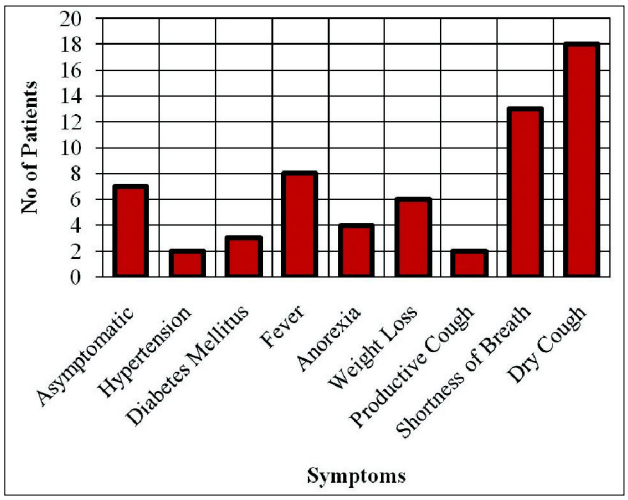
Histological Findings: IPF shows distinctive morphological features which are distinctive for the disease. Disease is characterized by appearance of honey comb change, micro anatomic distribution of inflammation, fibroblast proliferation, collagen deposition, and architectural remodeling.[23]

Materials and Methods

The study was a diagnostic validity study conducted at Islamabad Diagnostic Centre, Islamabad during September 2012 to February 2013. X ray reports of 30 diagnosed cases of Idiopathic Pulmonary Fibrosis on High Resolution Computed Tomography were evaluated. Consecutive sampling technique was adopted. All cases regardless of age and gender having positive disease features on HRCT were included in the study. Only those patients record were considered who have undergone HRCT and chest X-rays at same time. HRCT was performed by multislice, multidetector Aquilion Toshiba machine. 2 mm slice thickness; non contrast axial images through chest on HRCT protocol were taken. PA and lateral view of X-rays were also taken of same patient. Radiologist approved reports were taken. Data was analyzed by using SPSS v 10. Frequencies of positive finding on chest X-rays were calculated.

Results

Total 30 cases who met the inclusion criterion were included in the study during a period of 6 months. Majority of the patients were above age 55 years (80%) and were male (63.33%). Dry cough was found to be the most common presentation. Second most common symptomwas shortness of breath. Out of 30, 27 patients had positive X-Ray finding which shows its important role in diagnosis of disease. (Fig. 3) shows the frequency of symptoms presented by IPF patients.



Symptoms	Frequency	Percentage
Asymptomatic	7	23.33
Hypertension	2	6.66
Diabetes mellitus	3	10.00
Fever	8	26.66
Anorexia	4	13.33
Weight Loss	6	20.00
Productive Cough	2	6.66
Shortness of Breath	13	43.33
Dry Cough	18	60.00

Figure 3: Bar diagram of symptoms of IPF

Positive finding on Chest X-Rays:

Chest X-rays of patients, having positive IPF feature on HRCT were evaluated.

Value Label	Value	Frequency	Percent	Valid Percent	Cum Percent
Positive	1	27	90	90	90
Negative	2	3	10	10	100
	Total	30	100	100	

Table 1: Shows the frequencyof positive findings on Chest X Rays of patients diagnosed to have IPF on HRCT.

X-RAY		
N	Valid	30
	Missing	0
Mode	1	

Table 2: Chest X-Ray Findings

90% of the patients had positive X ray finding. This shows the importance of X-rays in diagnosing IPF.

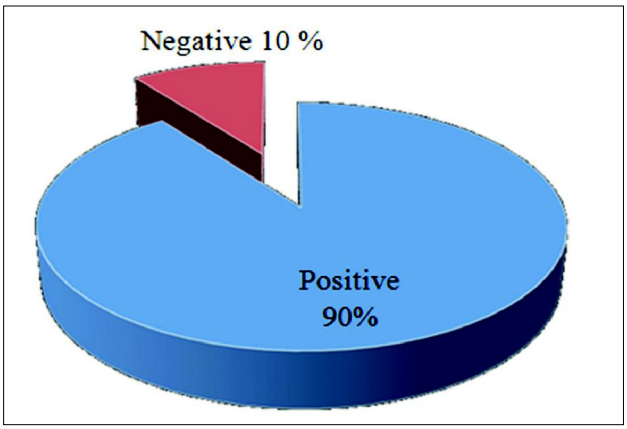


Figure 4: Pie chart for chest X-ray findings

Discussion

Idiopathic Pulmonary fibrosis is rare disease involving lung parenchyma having bad prognosis. Severe disease is not treated medically and leads to lung transplantation. Early detection of disease could help in medical treatment of disease.

Test used for diagnosing IPF include Chest X-ray, HRCT, lung function tests, exercise test, arterial blood gases and lung biopsy and other laboratory investigations. HRCT is the most sensitive and accurate method for diagnosing IPF. Average HRCT dose is 5-7 mSv.[24,25] HRCT shows a patchy predominantly peripheral sub pleural and basilar reticular pattern with sub pleural cysts (honey combing) and/or traction bronchiectesis. While on Chest X-ray in most of the cases disease appears as lower zone bi basilar reticular and reticunodular opacities.[26]

With arrival of HRCT many thought of obsolescence of X-rays. However, that's not the case. X-rays is still useful for the patients of IPF. X-rays because of their widespread availability and low cost is still among the First line investigations of IPF. It helps in early management of patients on one side and low radiation doses on other side. Also in countries like Pakistan, where HRCT is not affordable for every person, X-Ray becomes the prime investigation. Normal PA view on Chest X ray gives 0.02 mSv[17] and can be used for follow-up scans instead of HRCT. The radiation dose for HRCT is 100 times that of Chest X rays.[27] As disease require prolong follow-up around 2-3 years with serial HRCT at every 6 month causes extreme radiation burden to the patient with inconclusive significance.[28] In such cases X-ray could serve as good option.

Conclusion

Role of Chest X-ray in diagnosing IPF is still important and cannot be neglected especially in areas where HRCT is not available or patient could not afford HRCT, X-rays are good alternative. Also for the purpose of follow up and screening among coal worker, HRCT causes extreme radiation burden. Serial chest X-rays could serve the purpose.
The study was on short scale due to limited time and resources. I suggest that it should be conducted on large scale to fully understand the potential of conventional X-ray imaging. Also there is a need to conduct studies on using X-rays for screening purposes in Pakistan.

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