Diffuse Parenchymal Lung Diseases, Clinical Features, Radiological Findings and the Diagnostic Yield of Open Lung Biopsy

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ABSTRACT:

BACKGROUND:

Diffuse parenchymal lung diseases are rare , poorly understood and had not been studied in Iraq .

OBJECTIVE:

To evaluate the clinical features and radiological findings of diffuse parenchymal lung diseases in relation to open lung biopsy.

SUBJECTS AND METHODS:

Twenty eight patients who were suspected to have diffuse parenchymal lung diseases(regarding clinical features, pulmonary function testing , chest-x-ray and conventional computed tomography findings) were recruited from Baghdad Teaching Hospital from the 1st. Jan 2006 to 30th.June 2011 and were subjected to open lung biopsies which had been histopathologically studied.

RESULTS:

There were 16 (57%) males and 12 (43%) females, the mean age was 43.1±1.5 years, progressive dyspnea was the common presenting symptoms in 22 patients (78.6%) , dry cough was the presenting symptoms in 6 (21.4%)patients , restrictive lung defect was present in 26 (92.9%) patients, bilateral fine basal crepetations were heared in 24 (85.7%) patients, clubbing of fingers was present in 17 (60.7%) patients ,chest-x- rays findings were: 24(85.7%) patients had mainly lower zone involvement and 18(64.3%) patients had reticular infiltrate .CT findings were : 20(71.4%) patients had basal infiltrate and11(39.3%) patients had subpleural involvement. Open lung biopsy results were : 11 (39.3%) patients had usual interstitial pneumonia (idiopathic pulmonary fibrosis) , 7 (25%) patients had desquamative interstitial pneumonia,and 2(7.1%) patients had nonspecific interstitial pneumonia 2 (7.1%) patients had non caseating granulomas.

CONCLUSION:

Dyspnea on excertion was a common presenting symptom.

Bilateral fine basal crepetations were a common physical finding.

Restrictive lung defect was the major finding in pulmonary function testing.

Lower zone and basal infiltrates were common findings in chest-x-rays and conventional computed tomographies respectively.

Idiopathic pulmonary fibrosis (usual interstitial pneumonia) was the most common type of diffuse parenchymal lung diseases, followed by desquamative interstitial pneumonia.

Open lung biopsy gave a high diagnostic yield.

KEY WORDS: diffuse parenchymal lung diseases, open lung biopsy , idiopathic pulmonary fibrosis .

INTRODUCTION:

Diffuse parenchymal Lung Diseases (DPLDs) are rare heterogeneous group of conditions affecting the pulmonary interstitium and / or alveolar lumen , however they share similar symptoms, signs, radiological features and disturbance of pulmonary functions[1].The descriptive term "interstitial" reflects the pathologic appearance that the abnormality begins in the interstitium, but the term is somewhat misleading,as most of these disorders are also associated with extensive alteration of alveolar and airway architecture [2].

A definite diagnosis is essential to determine the prognosis and the therapeutic intervention for a given patient [3]. Open Lung Biopsy (OLB) is an intervention method but it is the gold standard diagnostic procedure[4] , with the current advances in technology: High Resolution Computed Tomograph (HRCT) and Video assisted Thoracoscopic Surgery(VATS) the need for open lung biopsy in every patient with suspected parenchymal lung diseases is still a question[5],

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however the role of this procedure remains controversial and many clinicians are reluctant to allow this invasive procedure to a high risk group of patients without assurances that results will lead to a change in therapy for a significant number [6].

SUBJECTS AND METHODS:

Twenty eight patients whose history (stress on occupation,smoking , drugs ,birds breeding and family history), clinical features, chest-x-rays and conventional computed tomographies (CT) chest findings were consistant with diffuse parenchymal lung diseases and were fit for open lung biopsy were included in the study,they were recruited from Baghdad Teaching Hospital from 1st. Jan.2006 to 30th.June 2011. patients who had history of takings drugs which can cause pulmonary fibrosis , on steroids, acute respiratory distress syndromes , immunocompromised patients , collagen and vascular diseases, severe co morbid diseases , poor pulmonary reserve and patients who were unable to stand the procedure of open lung biopsy were all excluded from the study.

Open lung biopsies were done at the cardio – thoracic department in the Specialized Surgical Hospital and the results of the biopsy specimens were examined by the same histopathologist at the same hospital.

RESULTS:

15 (53.6%) patients were males and 13 (46.4%) patients were females.Birds breeders were present in 2 (7.1%) patients , 1 patient(3.6%) was a carpenter , 1 patient (3.6%) was a farmer, smokers 6 (21.6%) patients, ex-smokers 2 (7.2%) patients and non smoker 20(71.4%) patients, 2 (7.1%) patients had positive family history of the same disease , progressive dyspnea was the common presenting symptoms in 22 patients (78.5%) , dry cough was the presenting symptoms in 6 patients (21.4%), restrictive lung defect was present in 26 patients(92.9%) , fine bilateral basal crepetations were heared in 24 patients(85,7%), clubbing of fingers was present in 17 (60.7%) patients ,chest-x-rays findings: 24(85.7%) patients had mainly lower zone involvement and 18(64.3%) patients had reticular infiltrate .CT findings were 20(71.4%) patients had basal infiltrate and11(39.3%) patients had subpleural involvement These results and other findings are shown in table (1).

Table 1: Presenting symptoms, signs, Pulmonary function testing, Chest-x-rays and CT findings in relation to patients .

Presenting symptoms	Total numbers and percentage
Dyspnea on excertion	22 (78.6%)
Dry cough	6 (21.4%)
signs	Total numbers and percentage
Bilateral fine basal crepetations	24 (85.7%)
Clubbing of fingers	17 (60.7%)
Central cyanosis	I0 (35.7%)
Rhonchii	2 (7.1%)
Normal auscultation	2 (7.1%)
Pulmonary function testing	Total numbers and percentage
Restrictive	26 (92.9%)
Mixed	2 (7.1%)
Chest-x-rays findings	Total numbers and percentage
Lower zone infiltrate	24 (85.7%)
Reticular infiltrate	18 (64.3%)
Reticulo –nodular infiltrate	7 (25%)
Nodular infiltrate	3 (10.7%)
Diffuse infiltrate	4(14.3%)
Mediastinal lymph node enlargement	3 (10.7%)
CT findings	Total numbers and percentage
Basal infiltrate	20(71.4%)
Sub pleural involvement	11(39.3%)
Ground glass appearance	6 (21.4%)
Honey combing	5 (17.9%)
Diffuse lung involvement	4(14.3%)
Mediastinal lymph adenopathy	3(10.7%)

Open lung biopsy:

All the 28 patients were subjected to open lung biopsies and the histopathological results were : 11 (39.3%) biopsies showed usual interstitial pneumonia (idiopathic pulmonary fibrosis) , 7 (25%) were desquamative interstitial pneumonia , 2(7.1%) were nonspecific interstitial pneumonia 2 (7.1%) non caseating granulomas and 2 biopsies were in conclusive ,these results and other histopathological results are shown in table(2).

Table 2: Open lung biopsy in relation to patients

Histopathological type	Total number and percentage
Usual interstitial pneumonia	11 (39.3%)
Desquamative interstitial pneumonia	7 (25%)
Non specific interstitial pneumonia	2 (7.1%)
Non caseating granulomas	2 (7.1%)
Hypersensivity pneumonitis	1(3.6%)
Alveolar proteinosis	1 (3.6%)
Lymangio lelomyamatosis	1 (3.6%)
Emphysema	1 (3.6%)
Inconclusive	2 (7.1%)

DISCUSSION:

The mean age of patients in this study was (43.1±1.5) years , the mean age in the Liverpool cohort study by Rizwan et al [6] was 49 years and this age difference could be attributed to the fact that elderly patients with comorbid diseases were excluded from our study, the mean age in the Kuwait study done by Ayed and Raghunathan[7] was 36.1 years as this study compared VATS with OLB as it showed a relatively a high percentage of Tuberculosis and vasculitis (both are common in adults) as compared to our study which did not report T.B cases and vasculitis was excluded from our study .

This study showed that dyspnea on excertion was the first presenting symptoms and dry cough was the second presenting symptoms, in a study done by Kursat et al showed that dry cough was the main presenting symptoms and dyspnea came next [8], clubbing of the fingers in this study was present in 60.7% of patients which was slightly higher than Jhonston et al of 25 to 50% of patients [9] as clubbing is a late feature in DPLD the majority of our patients presented late in the course of the disease , this could explain the relative high percentage of clubbing in our study .

Regarding the chest –x- rays and CT findings of lower,basal, reticular and subpleural lesions which were more consistant findings in our study, if we compare them with the two Turkish studies by Hider et al [10] which showed that sarcoidosis was the commonest histopathological diagnosis and Kursat et al [8] which showed idiopathic interstitial fibrosis of(29%) of cases **,** these above findings could be explained by the high percentage of idiopathic interstitial fibrosis in our study which commonly causes lower zone involvement ,reticular infiltrate and sub pleural lesion[1] .

Establishing an accurate diagnosis is essential so that the patient and his/ her family can be provided with reasonable expectations about the prognosis and the effect of therapy [6], a specific diagnosis in our study was reached in (85%) ,which was similar to Flaboris A et al [11] who reached a diagnosis in 90% and the Kuwait study in 93%[7] , while the Liverpool study reached a diagnosis in 42% [6] and (36-46%) in a study done by Walker et al [12] .

Open lung biopsy provides a sufficient material for histopatholgical diagnosis in most cases [13].

In this study (39.3%) of patients had idiopathic pulmonary fibrosis (usual interstitial pneumonia) which was similar to the Liverpool cohort study(42%) [6], but more than the Kuwait study [7] which was 25% .

This study showed that (25%) of patients had desquamative interstitial pneumonia which was higher than the joint international consensus statement by the American Thoracic Society and the European Respiratory Society [14]which was (3%) and this is rather difficult to explain ,as we need more Iraqi studies to confirm this finding in the future.

Thoracotomy for open lung biopsy has been a standard surgical approach for many years, recently the use of VATS for the diagnosis of DPLD has been increased [15] .

Hiatt et al[16] stated that open lung biopsy has only a modest clinical impact and should be used conservatively, though improvement in radiological methods for diagnosis especially using High Resolution CT increases the diagnostic rate up to(88%) as shown by Raghu [17] , but a normal HRCT does not exclude early and clinically significant interstitial lung disease[18]

CONCLUSION:

Dyspnea on excertion, bilateral fine basal crepetations were common clinical features .

Lower zone and basal involvement were common findings in chest-x-rays and CT respectively.

Idiopathic pulmonary fibrosis (usual interstitial pneumonia) was the most common type of diffuse parenchymal lung diseases, followed by desquamative interstitial pneumonia.

Open lung biopsy gave a high diagnostic yield.

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