

Open Lung Biopsy in Diffuse Infiltrative Lung Disease with Progressive Dyspnoea : Is It Useful?

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ABSTRACT

Eleven subjects, aged between 15 and 60 years, presenting with diffuse infiltrative lung disease (DILD) and progressive dyspnoea, underwent an open lung biopsy (OLE). The authors feel that OLD does give a confidence to the treating physician to begin with a specific therapy in the form of steroids. But, as a matter of fact, at most health care delivery centres in the country, facilities for OLB are not available. Hence, the specific therapy should be instituted presumptively following an overall suggestion of disease based upon the clinical, physiological (chiefly comprising the pulmonary function test or PFT) and the radiological criteria, so that progression of disease could be arrested at an early stage.

Key words : Diffuse infiltrative lung disease, Open lung biopsy.

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INTRODUCTION

In patients presenting with diffuse interstitial lung disease (DILD), a specific therapy is not only crucial to relieve symptoms but also necessary to improve the quality of life. A reasonably sure diagnosis may be made by history, clinico-radiologic examination, laboratory investigations and PFT. An open lung biopsy (OLB) has consistently given high percentage of diagnosis in all age groups with results ranging from 92-100 percent¹⁻⁴. Experience with open lung biopsy in Indian patients is limited. We carried out OLB in Indian patients with DILD having progressive dyspnoea.

MATERIAL AND METHODS

Eleven patients aged between 15 and 60 years

attending the Department of Tuberculosis and Chest Diseases at JIPMER, Pondicherry, between the years 1989 to 1992 and presenting with diffuse interstitial lung disease were categorized into early (symptoms < 5 years) and late (symptoms \geq 5 years) stages. Provisional diagnosis was based upon the clinico-radiologic evidences and the pulmonary functions. Tests for rheumatoid arthritis factor (RA factor), lupus erythematosus cell (LE cell) and antinuclear factor (ANF) were performed in all cases. An OLB carried out by the thoracic surgeon was done under local anesthesia by a mini-thoracotomy through the fifth intercostal space. Multiple pieces taken from lingula were subjected to histopathological examination and also sent for cultures for fungi, tubercle bacilli and pyogenic organisms.

RESULTS

A progressive dyspnoea (varying in duration from 1 to 30 years), history of anti-tubercular treatment (ATT) / antibiotics, radiologic evidence of bilateral reticulonodular shadows and a restrictive PET were observed in all subjects. Two of them also had an obstructive pattern. RA factor was positive in two subjects, whereas LE cell and ANF were negative in all. Characteristics of these patients including the clinico-radiologic findings, PFTs and histopathological features are presented in the table.

DISCUSSION

Diffuse interstitial lung disease (DILD) is characterized by progressive dyspnoea, clubbing, crepitations, reticulonodular shadows/interstitial infiltrates (on chest skiagram) and restrictive ventilatory impairment with diminished gas transfer. High resolution CT-scan (HRCT) and fiberoptic bronchoscopy (FOB) have revolutionised the diagnostic evaluation in recent times (with the cytological proof provided by a bronchoalveolar lavage and the histological confirmation by a transbronchial biopsy). FOB is a safe procedure, but may not provide a representative sample many times. An OLB remains the gold standard for diagnosis. Steroids constitute the mainstay of treatment during the early stages. With progression, fibrosis sets in and honeycombing becomes radiologically visible. Giving steroids to these late stage patients generally elicits a poor response⁵.

Most of the subjects in the present series had taken anti-tubercular treatment earlier. RA factor was positive in two of them (which may suggest the possibility of a connective tissue disorder), whereas LE cell and ANF were negative in all. One subject had an exposure to silica. DILD was provisionally diagnosed in all subjects on the basis of symptoms, occupational factors, clinical features (like clubbing, crepitations, etc.), laboratory investigations (suggesting association with some collagen disorder) besides the radiological abnormalities

and the pulmonary function defects. The histopathological findings on OLB did not suggest tuberculosis (TB), malignancy or any other specific disease process and corroborated with the clinical diagnoses. Thereafter, he were put on prednisolone (1 mg/kg/day) for six weeks followed by the tapering to a maintenance dose. All, except one patient (who expired) showed symptomatic improvement during the follow up of one to three years.

Although a diagnosis of DILD can be made on clinical, radiological and physiological grounds, there may be reluctance to begin with a specific therapy (i.e. steroids) for various reasons. First, persistence of other symptoms, such as the low grade fever favouring a tubercular process and the joint pains/stiff ness pointing to a collagen disorder, make a histopathological proof desirable for a conclusive diagnosis. Secondly, a physician needs to know the presenting stage of disease and the prognosis. Thirdly, an unwarranted long-term usage (in case of a mistaken diagnosis) is not free from side effects and can aggravate the patient's agony. Therefore, an OLB is usually advised in the developed countries.

Our clinical diagnosis in each subject was corroborated with the histopathology following an OLB. More recently, HRCT and FOB are being increasingly used to diagnose DILD. During the time of actual study performance more than a decade ago, these practices had not evolved, especially in India. Therefore, these were not carried out. Most health centres (majority of the peripheral and a few tertiary ones) are not equipped with such facilities and expertise even today. Hence we feel that even though histopathology gives a confidence to the clinicians in the institution of a specific steroid therapy, in the centres lacking these facilities a presumptive diagnosis of DILD may be made on the basis of clinical, radiological and physiological (chiefly comprising the PFT) criteria and the specific therapy may be instituted promptly to arrest the progression of disease at an early stage. Cases, in which a doubt persists, should be referred by these centres to the tertiary institutes for a histological confirmation by a bronchial biopsy or an OLB.

Table 1. Characteristics of patients having DILD

Case No.	Age & Sex	Clinical profile	Roentgenogram findings	Histopathology	PFT	Remarks
1.	31 F	Breathlessness - 6 years Joint pain and stiffness-1 year	Bilateral reticular shadows over lower zones	Emphysematous changes, mild interstitial non-specific inflammation and fibrosis	Restrictive pattern	• ATT taken for 1 year
2.	42 M	Breathlessness, cough with expectoration, low grade fever-2 years	Bilateral reticular and alveolar shadowing both mid and lower zones	Focal interstitial fibrosis with pneumonitic and bronchiectatic changes	Restrictive pattern	• RA factor positive
3.	50 M	Breathlessness -10 years; Clubbing present	Bilateral diffuse & dense honey-combing	Marked pleural fibrosis, dilated bronchioles with chronic non-specific inflammation, interstitial fibrosis and pulmonary hypertension	Mixed pattern	• Taken ATT for 18 months • Expired
4.	42 M	Breathlessness, cough, low grade fever-1 year; Clubbing present	Bilateral diffuse reticulonodular pattern	Mild emphysematous with focal interstitial fibrotic changes	Restrictive pattern	• ATT taken for 1 year
5.	45 F	Breathlessness and wheezing-5 years	Bilateral reticulonodular pattern	Diffuse fibrosis with focal chronic inflammation	Restrictive pattern	• RA factor positive
6.	40 F	Cough with expectoration, breathlessness-1 year	Bilateral reticular pattern both mid and lower zones	Marked interstitial pulmonary fibrosis in focal areas along with infiltration by chronic inflammatory cells in interstitium and alveolar lumen	Restrictive pattern	• ATT taken for 8 months
7.	56 M	Breathlessness, dry cough-4 years Clubbing present	Bilateral diffuse honey-combing, cardiomegaly present	Diffuse interstitial fibrosis with honeycombing and scattered foreign body giant cell reaction	Restrictive pattern	• ATT taken for 2 years • Electrical fitter in railways & exposure to silica present
8.	43 M	Cough, breathlessness - 30 years; Clubbing present	Bilateral coarse reticulonodular pattern	Diffuse interstitial fibrosis infiltrated by lymphocytes and plasma cells, sclerosed vessel and reactive changes in lymphnode	Mixed pattern	• Off & on ATT taken for 2 years
9.	18 M	Cough, breathlessness-1 year; Clubbing and polycythemia present	Bilateral reticulonodular pattern. No cardiac valvular lesion	Mild fibrosis with pneumonitis	Restrictive pattern	• DLC-Eosinophils 27% • Expired
10.	23 M	Breathlessness - 1 year	Bilateral honeycombing	Focal emphysematous changes with fibrosis No evidence of sarcoidosis	Restrictive pattern	• Improved on steroids • Lost on follow-up
11.	50 F	Breathlessness, cough-15 years; Stiffness of joints-2 years	Bilateral reticulonodular pattern over lower zones	Bronchiectasis with fibrosis and surrounding changes of pneumonitis	Restrictive pattern	• ATT & antibiotics taken off and on

There has been some controversy regarding the yield from lingular segmental biopsy as representative of pulmonary disease ranging from being unreliable and non-specific as reported by Newman *et al*⁶ to a 100% significant yield as found by Wetstein². We feel that a lingular biopsy can safely and quickly be performed by the thoracic surgeon under local anesthesia.

To conclude, although the OLD provides a confident diagnosis in patients of DILD, a specific treatment should not be withheld in absence of this facility. A diagnosis should be attempted on the clinical, physiological and radiological criteria and a specific therapy should be started as soon as possible.

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