

## Case Report

### Case Report of Diffuse Parenchymal Lung Disease with Non-Specific Interstitial Pneumonia Pattern and Good Response to Treatment

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#### Abstract:

The diffuse parenchymal lung diseases (DPLDs) are a heterogeneous group of conditions affecting the pulmonary parenchyma (interstitial) and/or alveolar lumen. IPF (Idiopathic Pulmonary Fibrosis) is a chronic interstitial pneumonia of unknown causes. It is commonest form of DPLD but its treatment response is very poor. On the other hand, NSIP (Non-Specific Interstitial Pneumonia) can still be a variant of DPLD with better treatment response and prognosis. Here we discussed a young female with NSIP with good response to steroid.

**Key words:** Diffuse Parenchymal Lung Disease (DPLD), IPF (Idiopathic Pulmonary Fibrosis), UIP (Usual Interstitial Pneumonia), NSIP (Non-Specific Interstitial Pneumonia), and HRCT (High Resolution Computed Tomography).

#### Case History:

Mrs. Tahera 22 years old normotensive, non-diabetic housewife hailing from Ambikapur, Faridpur admitted to Diabetic Association Medical College Hospital on 16<sup>th</sup> May 2017 with complaints of cough with exertional breathlessness for 3 months. Cough was dry and progressively increasing in intensity. Shortness of breath initially experienced only on strenuous exercise, but progressively increasing in intensity, now she feels shortness of breath on mild exertion. There was no history of diurnal or seasonal variation and not aggravated on dust or fumes. She mentioned about chest pain during coughing but no history of haemoptysis, joint pain, swelling or oral ulcer. She gave no history of loss of appetite or weight loss. Her bowel and bladder habits were normal. She had no history of contact with any tuberculosis patient.

On general examination, she was dyspnoeic, cyanosed, mildly anaemic, having clubbing. Her pulse was 68 b/min, BP-120/70mm Hg, and RR-25 breaths/min. On examination of respiratory system, her breath sound was vesicular, bilateral basal end inspiratory fine crepitation was present, which was not altered by coughing, and bilateral chest expansibility symmetrically reduced. Her investigation findings are shown in table I. Besides these we advised for lung biopsy, but patient party refused.

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**Table I:** Investigation Findings

Investigation	Findings
FVC	Reduced to 48%
FEV1	Reduced
FEV1:FVC ratio	High (90%)
TLC	Reduced.
DLCO	Markedly reduced -34% (Figure I)
CXR PA view	Inhomogeneous reticular opacities both in mid and lower zones (Figure II).
HRCT of chest	Ground glass opacity with mild reticular shadow consistent with NSIP pattern (Figure III).
ANA	Negative
RA test	Negative

On the background of clinical findings and HRCT findings, we started prednisolone 0.5 mg/kg/day for one month, tapered every 2 weeks to 5mg/day and then was maintained in 5 mg/day and N-acetylcysteine

600 mg 8 hourly. After 3-month follow up patient clinically improved, FVC also increased to 68% and 6 minutes walk test showed significant improvement. So we concluded that patient responded to treatment. Now patient is under follow up 2 monthly.

Last Name: BEGUM	Date of Birth: 1/1/1995	Age: 22			
First Name: MRS.TAHERA	Sex: Female	Weight (Kg): 58			
ID: V1704819337	Ethnic Corr.: 100	Height (cm): 158			
Date: 4/28/2017	Description:	BSA (m <sup>2</sup> ): 1.8			
Predicted: ERS 93	Company:	Smoke: No			
Technician: MD.MOKLESUR RAHMAN Ref. By: DR.MD MOHIUDDINAHMED					
<b>Lung Volumes Summary</b>					
Parameter	UM	Description	Pred.	TEST#5	%Pred.
FRC(DLCO)	l	Functional Residual Capacity during DLCO	2.38	1.43	60.1
RV(DLCO)	l	Residual Volume during DLCO	1.07	1.33	124.3
TLC(DLCO)	l	Total Lung Capacity during DLCO	4.11	2.67	65.0
RV/TLC(DLCO)%		RV(DLCO)/TLC(DLCO)	26.4	49.6	187.8
<b>Lung Diffusing Capacity Summary</b>					
Parameter	UM	Description	Pred.	TEST#5	%Pred.
DLCO	ml/min/mmHg	CO Diffusing Capacity	25.20	8.57	34.0
DLCO(corr)	ml/min/mmHg	DLCO corrected for Hb, Chb and PB	25.20	8.50	33.7
DLCO/VA	ml/min/mmHg/L	DLCO per unit of alveolar volume	6.13	3.18	51.9
VA	l(tpsv)	Alveolar Volume		2.67	

**Figure I:** Markedly reduced DLCO



**Figure II:** Chest X-ray shows inhomogeneous reticular opacities in both mid and lower zones

### Discussion:

Diffuse Parenchymal Lung diseases (DPLD) or Interstitial lung diseases (ILDs) are a diverse group of parenchymal pulmonary disorders characterized by varying degrees of inflammation and fibrosis. When pulmonary fibrosis predominates, ILDs are especially challenging to manage and treat. IPF, the most prevalent idiopathic interstitial pneumonia, has a median survival of 3 to 5 years from diagnosis<sup>1-3</sup>. Although its etiology remains unknown, potential risk factors for IPF includes cigarette smoking, gastro esophageal reflux, and certain environmental exposures<sup>4</sup>. Indications of trial of treatment include severe or



**Figure III:** HRCT shows ground glass opacity with mild reticular shadow

rapidly progressive symptoms, predominantly ground glass appearance on HRCT, reticular pattern on HRCT if lung function significantly impaired or patient requests treatment.

NSIP has distinguished etiologic, radiologic, pathologic, and response to treatment characteristics from other diagnoses in lung fibrosis particularly IPF. Purpose of this paper is to discuss the difficulties in confirming clinical diagnosis of NSIP & response of current treatment. According to the last ATS (American thoracic society) / ERS (European respiratory society) consensus panel<sup>5</sup> regarding idiopathic interstitial pneumonias, the more common causes of DPLD can be easily distinguished clinically, radiologically and pathologically from IPF, granulomatous conditions such as sarcoidosis and other less common causes of DPLD such as lymphangioleiomyomatosis and histiocytosis X.

Among the DPLDs; IPF, chronic hypersensitivity pneumonitis, and fibrotic connective tissue disease-

related ILD are associated with a worse prognosis, with death occurring as a result of both respiratory failure and serious associated co-morbidities<sup>6</sup>.

The distinct term of NSIP was first used by pathologists (Katzenstein) and later separately by radiologists. NSIP patients are frequently women in younger ages. This condition can present with other underlying disorders such as collagen vascular diseases or due to medications and lead to pulmonary fibrosis and at the same time chronic HP. Many times the etiology is idiopathic. In HRCT compared to Usual Interstitial Pneumonia (UIP), there is often less peripheral distribution and ground glass opacities consistent with inflammation seen. Pathologically, there is the uniformity of the interstitium, increased cellularity or fibrosis, but less honey combing. Yet, often it is difficult to distinguish fibrosing NSIP from IPF/UIP<sup>7-9</sup>.

Recent studies have shown that NSIP, even when initially diagnosed as an idiopathic form of NSIP, might be associated with an autoimmune background that later reveals itself as an organ-specific or a systemic autoimmune disease<sup>10</sup>. In a Cohort study of 27 idiopathic NSIP patients, more than 50% of the cases developed an autoimmune disease after a mean follow-up of 22 months<sup>11</sup>. So my patient also needs long term follow up. On the other hand, the response to the question of whether NSIP in rheumatologic diseases is the same as idiopathic NSIP is very difficult, particularly since in pathologic samples the signs of bronchiectasis or granuloma may not be found<sup>12</sup>. Overall, the ATS/ERS workshop committee has used the logical diagnosis of NSIP for middle-aged women and nonsmokers without clinical and serologic evidence for collagen vascular disease<sup>12</sup>. Even in some situations, NSIP has clinically been used in cases of undifferentiated connective tissue disorder<sup>13</sup>.

We believe that using the term NSIP and UIP provides the suggestion that in NSIP, steroids have absolute indication and the prognosis is better. On the other hand, UIP has poor prognosis and steroids are not useful. Yet, this is not the practical approach and the management of patients is individually based. It has previously been supported that NSIP is used for the cases discussed above. In the ATS/ERS 2002 categorization, grey area exists in this differentiation. The purpose of using distinct diagnostic entities due to variability in pathology, clinics and prognostics has been accepted. It can be concluded, that NSIP has been pathologically accepted as a distinct entity, but clinically it is used for middle aged women, nonsmokers with HRCT pattern consistent with NSIP<sup>4</sup> and response is good with steroid.

## Conclusion:

NSIP is a rare variety of DPLD but it is potentially responsive to systemic steroid. We can diagnose it on HRCT finding of ground glass opacity with reticular shadow specially in middle aged female with the help of histopathological findings. NSIP patient may present simultaneously with connective tissue disease, may be diagnosed later on, or may never with connective tissue disease.

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