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**Case Report**

# Nonspecific interstitial pneumonia presenting with high grade fever: a case report

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## ABSTRACT

Nonspecific interstitial pneumonia (NSIP), one subtype of idiopathic interstitial pneumonias is an autoimmune disease presenting usually at a younger age, most commonly in women. It can be distinguished clinically and pathologically from other subgroups of idiopathic interstitial pneumonias. Many cases of this entity occur in the context of an underlying disorder, such as a connective tissue disease, drug induced interstitial lung disease, or chronic hypersensitivity pneumonitis. Symptoms are non-specific and include insidious onset of dyspnoea and dry cough with restrictive pattern of decreased lung function and reduced gas exchange capacity. Fever or a flu-like syndrome can occur in up to one third of patients with NSIP. We report a case of 50 years old female who presented with breathlessness, cough and high grade fever. She did not have any features suggestive of connective tissue disease. On computed tomography of chest, diagnosis of NSIP was made and she responded to steroids.

**Keywords:** Idiopathic interstitial pneumonia, Nonspecific interstitial pneumonia, Cough, Fever

## INTRODUCTION

Nonspecific interstitial pneumonia (NSIP) is a subgroup of the idiopathic interstitial pneumonias that can be distinguished clinically and pathologically from other subgroups like usual interstitial pneumonia (UIP) or idiopathic pulmonary fibrosis (IPF), acute interstitial pneumonia (AIP), cryptogenic organizing pneumonia (COP) or bronchiolitis obliterans with organizing pneumonia (BOOP).[1] Idiopathic NSIP is a sub-acute restrictive process with a presentation similar to that of IPF but usually at a younger age, most commonly in women who have never smoked. It is often associated with a febrile illness. HRCT shows bilateral, sub pleural ground-glass opacities, often associated with lower lobe volume loss.[2] We report a case of NSIP who presented with high grade fever, such case has not been found in the literature reported earlier.

## CASE REPORT

50 years old female was admitted on 16$^{th}$ February, 2015 with complaint of fever for last two months. On history, her illness started with cough and intermittent fever two months back. Fever was initially low grade, which became high grade for the last two weeks. Cough was dry with no haemoptysis or wheezing chest. She developed dyspnoea on moderate exertion for the last one week. There was no history of smoking and drug intake prior to the episode and no history of radiation in the past. No arthralgia or other symptoms suggestive of connective tissue disease were present. On examination, she had pulse 100/minute regular, respiratory rate 30/minute, normal blood pressure, no lymphadenopathy, no discharge, no sinus formation, normal thyroid, no other swelling. JVP was normal. Chest examination revealed bibasilar fine crepitations. CVS, CNS and abdomen

examination were normal. On investigations, Hb 8.8 gm%, total and differential leucocyte count normal, platelet count normal and ESR 130 mm in 1st hour. Tests for malaria, dengue, and typhoid were negative. Viral markers were negative and LFT, RFT were normal. ANA was positive. On x-ray chest, there were bilateral reticular shadows at bases as in Figure 1. Patient could not perform spirometry. Oxygen saturation on room air was 90%. She was put on oxygen and given broad spectrum I/V antibiotics empirically. Patient did not improve for one week. She had intermittent fever with maximum temperature ranging from 101°F to 104°F. HRCT chest showed multiple patchy ground glass opacities and interstitial septal thickening predominantly in lung bases suggestive of interstitial inflammation – NSIP pattern as shown in Figure 2. BAL and lung biopsy were not done. With the diagnosis of NSIP, she was put on prednisolone 1mg/kg body weight and antibiotics were stopped. Patient responded in 24 hours with no fever and cough. She was discharged on 3rd day. She followed our OPD after 10 days with no complaint of fever or cough.

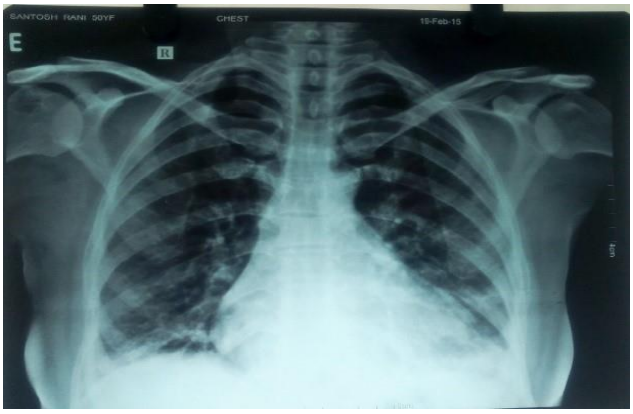

**Figure 1: Chest X-ray showing reticular shadows in both lower zones.**

## DISCUSSION

Nonspecific interstitial pneumonia (NSIP) was first described in 1994 by Katzenstein et al.[3] It is an uncommon idiopathic interstitial pneumonia. Most patients are women, between the ages of 40 and 50, and have no known cause or association. However, a similar pathologic process can occur in patients with a connective tissue disorder (in particular, systemic sclerosis or polymyositis/dermatomyositis), in some forms of drug-induced lung injury, and in patients with hypersensitivity pneumonitis.[4] The reported patient was 50 years old female, no history of prior drug exposure, or radiation exposure or exposure to environmental pollution. However, her ANA test was positive but no other autoantibodies tests were done because the patient could not afford it. She did not have any features suggestive of connective tissue disease.

Generally, a high-resolution computed tomography (CT) scan is performed to help diagnose NSIP. CT scan of patient with NSIP shows a typical "ground glass" pattern that represents interstitial inflammation, and is usually seen in the cellular form. Scarring or fibrosis is seen in the fibrotic form.[5] HRCT done in this patient showed features suggestive of NSIP.

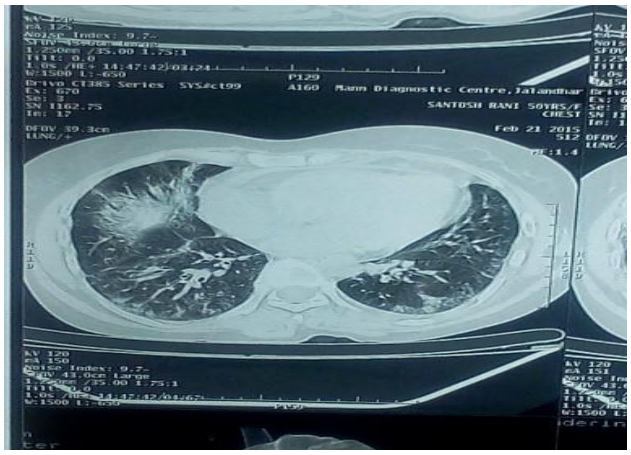

**Figure 2: HRCT Chest showing multiple patchy ground glass opacities and interstitial septal thickening in both lung bases.**

Most patients with the cellular type of NSIP respond well to treatment with oral corticosteroids, such as prednisone. However, patients who do not respond to corticosteroid therapy may require additional treatment with immunosuppressive drugs. Patients with the fibrotic type of NSIP might benefit from the use of both types of drugs to prevent further irreversible fibrosis. The overall prognosis and response to steroid therapy is generally favourable in patients with NSIP.[6,7] However, not all patients respond well to treatment. In some patients, relapse can occur when steroids are tapered or stopped. In our patient, fever and cough subsided within 48 hours of starting prednisolone and she is doing well on follow up.

## CONCLUSION

NSIP is a very rare entity encountered in daily practice. A high degree of suspicion for this diagnosis is to be kept in mind in patients who present with dry cough and fever. Though BAL and lung biopsy is needed to confirm the diagnosis of nonspecific interstitial pneumonia. The invasive procedures are usually associated with complications. But HRCT chest and clinical acumen can guide us in keeping this differential diagnosis. And the rapid response of the patient to steroid therapy as in our case within a period of 48 hours from a long illness confirms the diagnosis.

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