KEY WORDS:
Adult onset still's disease, respiratory distress, interstitial pneumonitis, steroid.

INTRODUCTION
Adult onset Still’s disease is a multisystem inflammatory disorder characterized by high spiking fever, rash, polyarthritis, lymphadenopathy and hepatosplenomegaly. Polymorphonuclear leukocytosis in the absence of infection, elevated erythrocyte sedimentation rate (ESR) and high serum ferritin are characteristic features. Rashes are typically faint salmon colored, transient, appearing along with spike of fever. Pleural effusion, upper lobar pneumonia, pericardial effusion and myocarditis are also reported.

CASE REPORT
A 30 year old lady was admitted with high grade intermittent fever and non productive cough of 18 days and dyspnoea of 2 days. There was no chest pain or haemoptysis. She was a non-smoker, did not have pets, neither had any travel or contact history in recent past. She had a temperature of 39°C; mild pallor; respiratory rate was 30 breaths/min and had no lymphadenopathy, clubbing or cyanosis. Her trachea was normal in position. Chest auscultation revealed few crackles over left mammary and axillary region. She gave a history of intermittent fever, rash and hepatosplenomegaly. Pulmonary involvement in the form of pneumonitis, as a presenting feature is very rare. We report a case of a young lady who presented with fever, cough and respiratory distress. Chest X-ray revealed patchy infiltration in left upper lung zone. She was subsequently diagnosed as Adult onset Still’s disease. There was no improvement in clinical condition despite five days of antibiotics. On trans-bronchial lung biopsy (TBLB) proved she had interstitial pneumonitis and responded dramatically to steroid treatment.

SUMMARY
Adult onset still’s disease usually presents with high grade intermittent fever, polyarthritis, salmon pink evanescent rash and hepatosplenomegaly. Pulmonary involvement in the form of pneumonitis, as a presenting feature is very rare. We report a case of a young lady who presented with fever, cough and respiratory distress. Chest X-ray revealed patchy infiltration in left upper lung zone. She was subsequently diagnosed as Adult onset Still’s disease. There was no improvement in clinical condition despite five days of antibiotics. On trans-bronchial lung biopsy (TBLB) proved she had interstitial pneumonitis and responded dramatically to steroid treatment.

As the clinical and radiological findings of chest showed no signs of resolution even after seven days of treatment, a flexible bronchoscopy was performed. Bronchoalveolar lavage and bronchial brush specimens were negative in acid-fast smear test, culture and polymerase chain reaction for tubercle bacillus. Histological examination of the transbronchial lung biopsy specimen disclosed interstitial pneumonia pattern and focal areas of atelectasis (Figure: 2).

DISCUSSION
Pulmonary manifestation of AOSD is reported to be 12-53% for pleuritis and 0-27% for interstitial pneumonia.  Transient pulmonary infiltrates and pleural effusion are the most common pulmonary diseases. Occasional cases of cryptogenic organizing pneumonia and diffuse pulmonary hemorrhage (DAH) are also reported.
But AOSD presentation as pulmonary involvement is quite rare. Our patient presented with long continued fever and clinical as well as radiological features of pneumonia. After excluding common etiologies, diagnosis of AOSD was entertained on the basis of more than 2 weeks of fever, polyarthritis, rash, persistent neutrophilic leukocytosis and elevated liver enzymes. Hyperferritinaemia, negative ANA and Rheumatoid factor further helped in diagnosis.

Interleukin-18 (IL-18) is the most important proinflammatory cytokine in AOSD pathogenesis. It is overproduced in the acute phase of the disease and is believed to initiate the inflammatory cascade that includes interferon gamma, IL-6, and tumor necrosis factor alpha. Increased IL-18 levels is found in both blood and lung tissue in endotoxemia-related lung injury. This may lead to interstitial lymphoplasmacytic infiltration and pulmonary capillaritis resulting in DAH.

Hyper-ferritinemia in AOSD is probably a consequence of cytokine secretion induced by the reticuloendothelial system or hepatic damage. A fivefold increase in serum ferritin has 41% specificity and 80% sensitivity. Ferritin levels correlate with disease activity; and after remission get normalized. Liver function abnormalities occur in the form of elevation of serum transaminases. Ferritin was raised in our case as were serum transaminases.

Aspirin or non steroidal anti inflammatory drugs are recommended as initial treatment of AOSD, but response rate is low. Most patients are therefore treated with corticosteroids in the course of their disease, with an efficacy of up to 95%. Steroid should also be used for those suffering from persistent anemia, pericarditis, serositis and marked elevation of liver enzymes. We treated our patient with high dose methylprednisolone for her non resolving systemic and pulmonary manifestations and got excellent result.

We therefore conclude that pulmonary involvement in the form of interstitial pneumonia can be an initial presentation of Adult-onset Still’s disease which responds dramatically to glucocorticoid treatment. AOSD diagnosis should be considered by clinicians in a case of non resolving pneumonia after exclusion of common infective, malignant and inflammatory etiologies.

REFERENCES