Case Report

A case of spontaneous pneumothorax, pneumomediastinum and subcutaneous emphysema in *Pneumocystis jirovecii* pneumonia complicating HIV

S Chatterji[1], D Pal[1], MK Ghosh[1], S Pal[1], A Naskar[1], S Mallick[1], I Bhattacharya[2]

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Abstract:

Spontaneous pneumothorax and subcutaneous emphysema are well-known but less common complications of *pneumocystis* pneumonia, whereas pnuemomediastinum is rare. We had a patient known to be HIV seropositive, who developed *pneumocystis jirovecii* pneumonia. During the course of treatment, he developed spontaneous pneumothorax, pneumomediastinum and subcutaneous emphysema. He had no predisposing factors known to cause this complication.

Key words: *Pneumomediastinum, pneumothorax, subcutaneous emphysema, Pneumocystis jirovecii, AIDS.*

Introduction:

Pneumocystis jirovecii is an opportunistic fungus found ubiquitously. It causes interstitial pneumonia in immunocompromised hosts. It is acquired by inhalation and can cause fatal disease if not recognized early, especially in persons with some form of immunodeficiency. In severe disease, spontaneous pneumothorax and subcutaneous emphysema can occur but pnuemomediastinum occurs only rarely.[1] Doppman et. al[2] in a review of atypical radiographic features in 30 cases of *pneumocystis* pneumonia specifically noted a total absence of spontaneous pneumothorax in adults. Recent reports[3,4] however have shown that *Pneumocystis jirovecii* pneumonia can cause lung tissue destruction and spontaneous pneumothorax in patients with the acquired immunodeficiency syndrome (AIDS).

Case Report:

A 36 year old gentleman was suffering from low grade, intermittent fever with loose motions for one month. It was associated with non-productive cough and exertional shortness of breath for fifteen days. His breathlessness was insidious in onset and progressive in nature. He received anti-tubercular therapy for pulmonary tuberculosis seven years back and was found to be seropositive for HIV-1 (by rapid antibody card test, test kit batch no - 4000012651, date of expiry - 28.05.2015) three weeks before the present illness. He was a non-smoker and height was 162cm.

On examination, the patient was alert, conscious and had evidence of oropharyngeal candidiasis. He was tachypnoeic (respiratory rate 40/min), with a heart rate of 104/minute and normal blood

[1] *Department of Tropical Medicine,[2]Department.of Microbiology, School of Tropical Medicine, Kolkata, India*

Address for correspondence: Dr.Dipankar Pal, Department of Tropical Medicine, School of Tropical Medicine, Kolkata, India. E-mail : dipankarpal.2009@gmail.com Phone- 91-9432113713

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pressure, single breath count was 8, cyanosis was present and SpO2 was 90% at rest without oxygen. Except a small area of diminished breath sounds in the right infra-axillary area, the chest examination was unremarkable. Chest x-ray done 15 days previously was normal. However, the x-ray done on admission revealed right lower zone infiltrations. No cavity/cyst was apparent. His sputum grew *Streptococcus pyogenes* but acid fast bacilli were absent. Blood CD4 count was 91cells/µL. He was given moist oxygen inhalation, oral amoxicillin for streptococcal infection and fluconazole for oropharyngeal candidiasis.

The clinical presentation and a low CD4 count in an HIV infected subject, led to the suspicion of *Pneumocystis jirovecii* pneumonia (PJP). Sputum induction done with hypertonic saline nebulization showed the presence of *Pneumocystis jirovecii* cysts in Giemsa stained smear under microscope.(Fig.1)

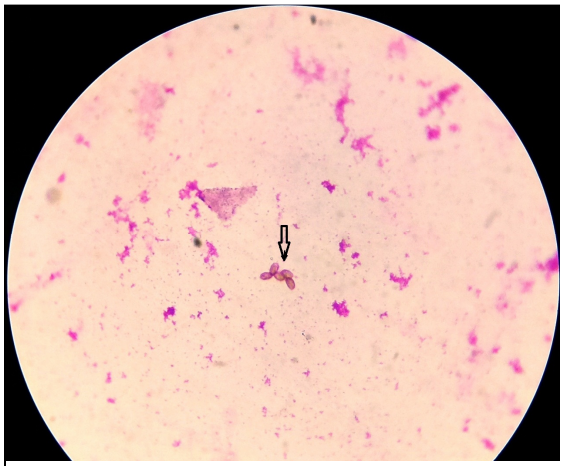

Figure. 1: Sputum smear showing *Pneumocystis jirovecii* cysts (black arrow) in Giemsa stained smear 100X magnification.

He was immediately shifted to the critical care unit and closely monitored. Repeat x-ray revealed subcutaneous emphysema and right sided small pneumothorax with pneumomediastinum (Fig.2). Contrast enhanced CT (CECT) thorax confirmed the presence of pneumomediastinum and also revealed the presence of some cystic lesions and bronchiectatic changes in both lungs with ground glass opacity (Fig.3,4).

The patient was kept on conservative treatment with close monitoring. His subcutaneous emphysema, pneumothorax and pneumomediastinum resolved in about two weeks. He was returned to the general ward. Anti-retroviral therapy (ART) was initiated and he was discharged on ART and secondary prophylaxis for

Treatment with co-trimoxazole and prednisolone was started in the recommended doses (trimethoprim 15mg/kg/day).[5] He started to improve gradually. Cyanosis was absent on day three and he became ambulatory.

On day ten of admission, he complained of sudden increase in shortness of breath started at rest and became cyanotic again. Subcutaneous emphysema was noted on both sides of neck. Trachea and apex beat were normal in position and there was slight diminution of vesicular breath sound bilaterally.

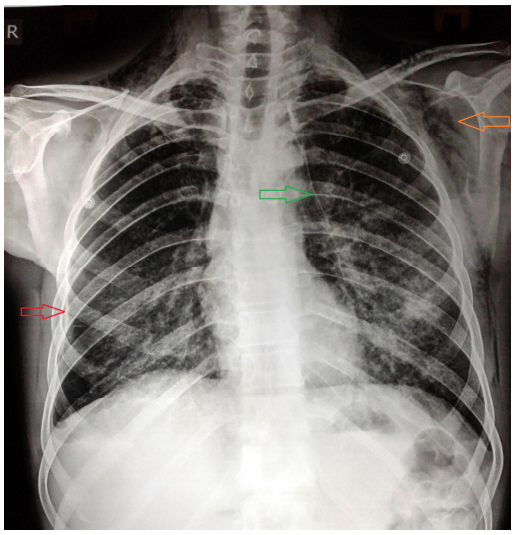

Figure. 2: Chest x-ray showing
a) Collapsed lung border (red arrow),
b) Subcutaneous emphysema (yellow arrow)
c) Pneumomediastinum (green arrow).

Pneumocystis jirovecii with co-trimoxazole. The major events in the process of disease evolution are depicted in Fig 5.

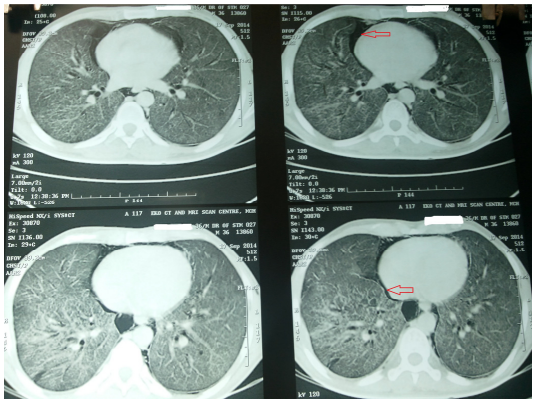

Figure. 3: CECT thorax showing ground glass opacity and pneumomediastinum (red arrows)

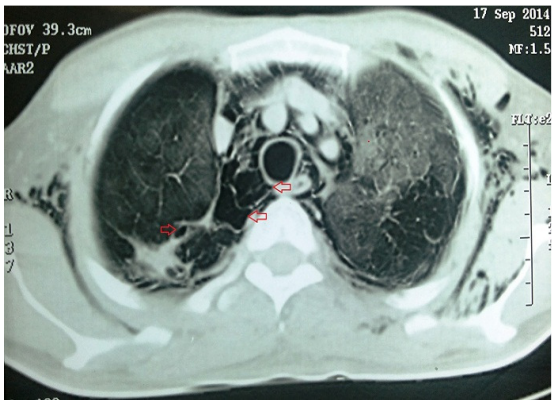

Figure. 4: CECT thorax showing cysts in apices of both lungs (red arrows)

History of Pulmonary Tuberculosis (treated)

↓7years later

Diagnosed to be seropositive for HIV-1

↓3weeks later

Presented with shortness of breath , tachypnea.
Diagnosed to be PJP & treatment started.

↓10days later

Developed pneumomediastinum, pneumothorax & subcutaneous emphysema.

↓ 2 weeks

Pneumomediastinum, pneumothorax & subcutaneous emphysema resolved, anti-retrovirals (ART) initiated & patient discharged on co-trimoxazole prophylaxis & ART.

Figure 5: Timeline of events in evolution of disease.

Discussion:

Subcutaneous emphysema with pneumomediastinum is a well known complication of blunt or penetrating chest trauma, soft tissue infections or any condition that creates a pressure gradient between the perivascular interstitial and intra-alveolar spaces. Mediastinal emphysema has been described in a variety of clinical situations including parturition[6], administration of general anaesthesia[7], acute bronchial asthma[8] as well as in apparently healthy subjects with no underlying lung disease.[9] *Pneumocystis* has been associated with pneumatocele and cyst formation in 7-34% of cases in AIDS patients.[10,11,12] Pneumomediastinum is rare in AIDS

patients[1] infected with *Pneumocystis jirovecii* and there are no recent reports of this complication.

Several mechanisms have been suggested to explain the pathogenesis of lung cavities in *Pneumocystis jirovecii*[1] pneumonia. These include check valve bronchiolar obstruction with distal cyst formation[13], elastase release from macrophages with subsequent destruction of the alveolar tissue[14], direct or indirect cytotoxic effects of HIV[15] and arterial invasion with thrombosis of the vessels, necrosis and cavitation[16] among others. Aerosolized Pentamidine used to treat *Pneumocystis jirovecii* has also been related to pulmonary cysts and pneumothorax[17] development**.** Reports also suggest that some of the changes may even be reversible with treatment**.**[18] The cysts are typically apical and subpleural, lined by fibrosis and/or alveolar parenchyma with little inflammation in the acute stage. Necrotizing, thin-walled, smaller intraparenchymal cavities lined by organisms, exudate and chronic inflammation can also be seen**.**[19] Some propose that the mechanism of spontaneous pneumothorax/pneumomediastinum is spontaneous rupture of necrotic lung tissue occurring in a subgroup of AIDS patients in which the interstitial inflammatory response to *Pneumocystis* has been accelerated**.**[20] Patients at risk of pneumothorax/pneumomediastinum include those with cystic lesions on chest radiograph and those patients with a history of *Pneumocystis* pneumonia. Patients with AIDS and *Pneumocystis* pneumonia who develop pneumothorax during mechanical ventilation have a poorer outcome.[21]

In our case, there was no past history of *Pneumocystis* pneumonia and he is non-smoker. His arm span (152cm) is shorter than height (162cm) suggesting that he does not have a marfanoid habitus which is a known predisposing factor for developing pneumothorax in young adults. There was no similar history of pneumothorax in his family. Chest radiograph had infiltrative changes in the right lower zone, but no cyst/cavity could be seen and the apices of both lungs appeared normal. However, CECT thorax revealed fibrotic, cystic and bronchiectatic changes. There was also no history of aerosolized Pentamidine therapy. The patient developed pneumomediastinum as well as subcutaneous emphysema while he was recovering. Thus physicians should be alert about this complication in acute as well as in recovery phase.

Conclusion:

The unique features of our case are as follows: firstly, there was spontaneous pneumomediastinum, pneumothorax and subcutaneous emphysema without any known risk factors as discussed above. Secondly, these complications developed when the patient was in the recovery phase. Treating physicians should therefore have a high index of suspicion while treating patients with *Pneumocystis* pneumonia and should be vigilant during the whole treatment duration as this potentially fatal but treatable complication can occur anytime during the disease course.

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