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Pneumocystis jiroveci Pnemonia in Systemic Lupus Erythematosus: A Case report

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Abstract
Pneumocystis jiroveci pneumonia (PJP), formerly known as *Pneumocystis carinii* pneumonia (PCP) is the most common opportunistic infection in immunocompromised patients including patients with systemic lupus eythematosus (SLE). To our knowledge there was no case report on *Pneumocystis jiroveci* pneumonia among the patients with SLE in Bangladesh. We report on a case of SLE who developed PJP. A 37-year-old housewife with a 12-year history of SLE presented with dry cough, breathlessness and fever for one month. The patient had been treated with varying doses of corticosteroids and/or cytotoxic drugs within the last 12 years and had been taking cyclosporine 200 mg daily along with prednisolone 0.5 mg/ kg body weight with gradual tapered dose to 7.5 mg/day for 3 months before presentation. Her Chest X-ray revealed bilateral diffuse infiltrate, CT scan of chest also disclosed bilateral diffuse infiltrate. Diagnosis was established based on the findings of Geimsa stain of induced sputum. After treatment with Trimethoprim-sulfamethoxazole (TMP-SMX; TMP: 5mg/kg, SMX: 25mg/kg 8 hourly) the patient improved clinically and discharged to home. Patients with SLE like other immunocompromised patients presenting with fever, cough and breathlessness should arouse a high suspicion for *Pneumocystis Jiroveci* pneumonia. Patients with SLE treated with corticosteroids and cytotoxic drugs may be at increased risk for this opportunistic infection.

Keywords: *pneumocystis jiroveci* pneumonia (PJP), erythematosus

Introduction

Case summary

A 37- year-old housewife with a 12-year history of Systemic Lupus Erythematosus (SLE) presented with cough, breathlessness and fever for one month. SLE was diagnosed on the basis of malar rash, oral ulcer, photosensitivity, joint pain, positive anti-nuclear antibody and high titer of anti-double stranded DNA antibodies. She had history of taking methotrexate 20 mg weekly for 8 years with good response, but it was stooped due to thrombocytopenia. She also had history of taking azathioprine 100 mg daily for one year with inadequate response. Her joint pain increased three months prior to her admission, she was prescribed cyclosporine 200 mg daily along with Tab. Prednisolone 0.5 mg /kg body weight with gradual tapering dose. On examination the patient appeared very sick, She was mildly anaemic, temperature was 103^0 F, ulcers were present on her tongue and hard palate, pulse was 142 beat per minute, respiratory rate 42 per minute, blood pressure 80/60 mm of Hg, and the oxygen saturation was 81% while she was breathing ambient air. Bilateral crackles were heard on lung auscultation. She was mildly anaemic. Initial laboratory investigation showed a total white cell count of 9.5 x 10^9/L, neutrophil 93%, lymphocyte 06%, eosinophil 01%, hemoglobin 11.2 g/dL, platelet count 300 x 10^9/L, ESR-120 mm in first hour, serum ALT-26 u/L, anti dsDNA –negative, sputum for AFB-negative, sputum for c/s revealed normal flora, chest X-ray showed bilateral interstitial and alveolar infiltrates (Fig. 1). CT scan of chest also disclosed bilateral diffuse infiltrate (Fig.2). Empirical treatment with Injection ceftazidime and amikacin was started. Considering clinical scenario and the high prevalence of tuberculosis in our country anti-tubercular drugs were also started according to the advice of a pulmonologist. No significant improvement occurred despite these therapies. Giemsa staining of sputum induced with 3% NaCl revealed *Pneumocystis jiroveci*. TMP-SMX (TMP: 5mg/kg, SMX: 25mg/kg 8 hourly) was started. Anti HIV 1 & 2 were negative. Sputum for GeneXpert MTB/RIF was also negative. Anti-tubercular drugs were stopped. The patient improved clinically. She was discharged with instructions to complete twenty-one days treatment with TMP-SMX. She was counseled to follow-up in our SLE clinic.

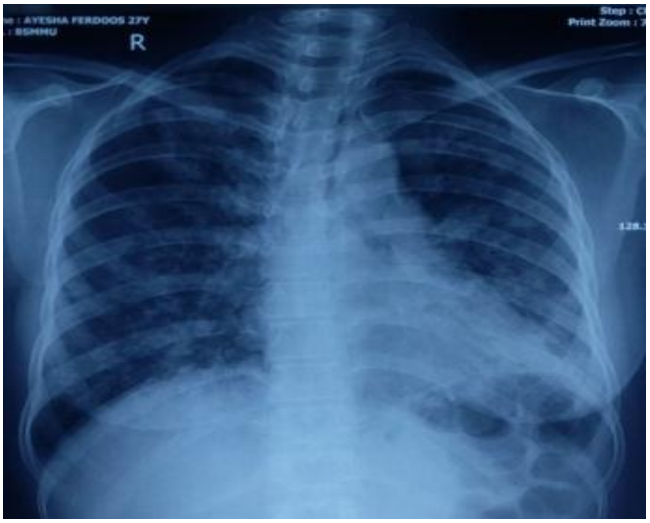

Fig 1: X-ray chest pstero-anterior view Showing Diffuse bilateral pulmonary infiltrate

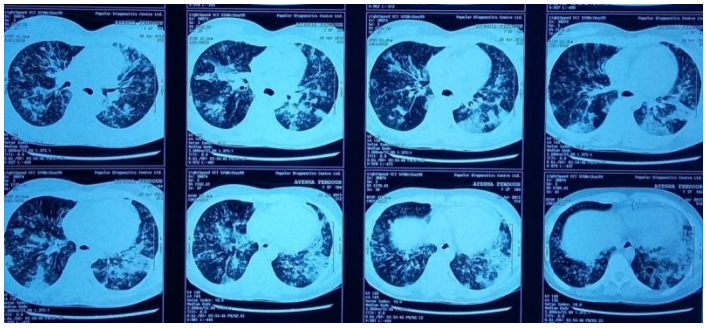

Fig 2: CT scan of the chest Showing Diffuse bilateral pulmonary infiltrate

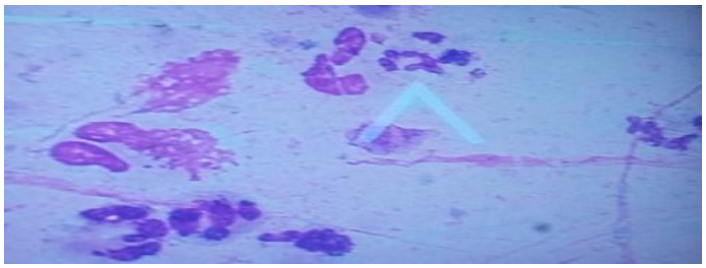

Fig 3: Sputum with Giemsa Stain revealed Pneumocystis Jiroveci.

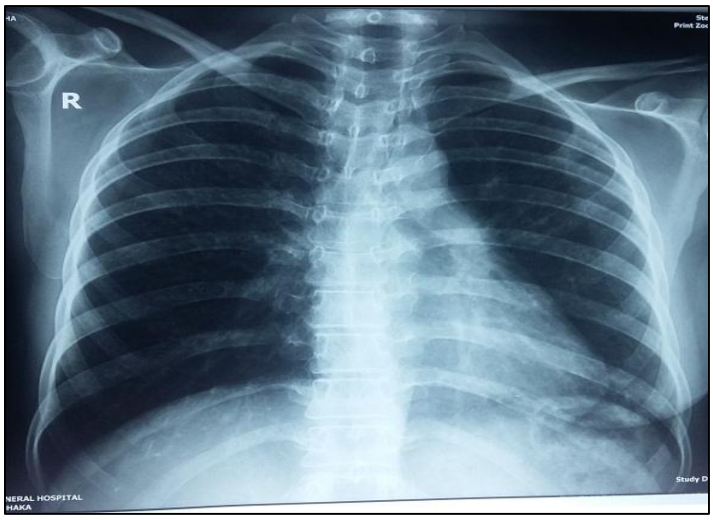

Fig 4: X-ray chest showing normal findings after six weeks of treatment withTMP-SMX

Discussion

Lung involvement in patients with SLE treated with immunosuppressive agents has a broad differential diagnosis including opportunistic lung infection, *Pneumocystis Jirovecis* among the most frequent of these opportunistic infections. Connective tissue diseases are associated with *PJP* in about 5% of cases [1, 2]. Fatal infections in SLE patients are correlated with the use of prednisolone and cytotoxic drugs in the 3 months before death and with prednisolone doses greater than 40 mg/day [3, 4]. Yale and Limper reported respiratory failure in 50 of 116 patients with HIV-negative *PJP* (43%), and these patients had an in-hospital mortality rate of 66% [5]. Table 1 demonstrates the reported larger-scale studies on lupus patients with PCP from different geographical areas in the recent 2 decades [6, 7] Five patients achieved the definite diagnosis of PCP out of 858 lupus patients in a institute of Southern Taiwan [7]

Table 1: Reported Large-Scale Studies on Lupus Patients with PCP in Different Geographical Areas Since 1992[7]

Serial No.	Geographical Location	Publication Year	Type of Study	Total No. Patientsin Study	PCPIncidence n (%)
1	Malaysia	1992	S, R	351	9 (2.56)
2	France	1994	S, R	750	6 (0.80)
3	United States	1996	S, R	100	3 (3.0)
4	Japan	1996	S, R	59	1 (1.69)
5	United States	1999	M, R	72,816	94 (0.13)

6	Korea	1999	S, R	544	0 (0)
7	Hong Kong	2000	S, R	186	0 (0)
8	France	2001	S, R	87	0 (0)
9	Canada	2002	S, R	363	3 (0.83)
10	United States	2005	S, P	59	1 (1.69)
11	United States	2005	M, P	140	0 (0)
12	Hong Kong	2006	S, P	212	0 (0)
13	China	2006	S, R	283	2 (0.71)
14	Spain	2006	S, P	110	2(1.81)
15	Thailand	2007	S, R	542	2 (0.37)
16	Argentina	2009	S, A	90	1 (1.11)
17	Thailand	2011	S, R	119	3 (2.52)
18	Taiwan	2013	S, R	858	5 (0.58)

Published reports with total patient number of more than 50 were selected in this table.
A indicates autopsy; M, multicenter; P, prospective; R, retrospective; S, single institute

The mechanism of immune suppression in patients with SLE who have PJP is usually multi-factorial, and may be related to underlying diseases, cytotoxic therapies, or malnutrition. However, the development of PJP in most patients with SLE is associated with daily administration of corticosteroids and with the development of lymphopenia.

McGonigle [8] *et al* concluded in a study that cylosporin A increases the incidence and adversely affects the prognosis of immunocompromised patients with P. carinii infection. They also found that patients with PJP had significant hypoxia. Hanauer[9] reported a case of PJP while he had been receiving 500 mg of cyclosporine A and 30 mg of prednisolone per day for two months, the patient developed dyspnea, non-productive cough and fever. His chest film revealed a diffuse pulmonary infiltrate. Our patient presented with dyspnoea, fever and cough. She had been taking 200 mg of cyclosporine and 0.5 mg/kg of body weight of prednisolonewith tapered dose for three months before developing her symptoms. Our patient was hypoxic (SPO_2- 81%) and lymhopanic (Lymhocyte- 06%). Her chest X-ray showed diffuse pulmonary infiltrate. CT scan of chest also revealed bilateral diffuse infiltrate. 3% NaCl induced sputum for Giemsa stain confirmed the presence of *Pneumocystis Jiroveci*. She was treated with a standard treatment of PJP consisting of TMP-SMX, prednisolone along with supplemented oxygen. She was symptomatically improved, her SPO_2 became normal. She was discharged and followed up for several months. Our important differential diagnoses were pulmonary tuberculosis, bacterial pneumonia and lupus pneumonitis. Though initially we empirically started anti-tubercular drugs along with ceftazidime and amikacine, we stopped these drugs soon after confident exclusion of the differentials.

Conclusions

Immunocompromised patients like SLE with or without immunosuppressive therapy presenting with dyspnoea, cough, fever and hypoxia should arouse a high suspicion for PJP and mandate urgent investigation as well as management.

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