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Original Article

Clinical pattern in Egyptian systemic lupus erythematosus patients with pleuropulmonary involvement

Ayman K. El-Garf, Rasha E. Gheith *, Shaimaa N. Badran

Rheumatology and Rehabilitation Department, Faculty of Medicine, Cairo University, Egypt

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ABSTRACT

Aim of the work: To study and analyze the clinical features of Egyptian systemic lupus erythematosus (SLE) patients with pleuro-pulmonary system involvement.

Patients and methods: All SLE patients admitted to the Rheumatology and Rehabilitation inpatient Department, Cairo University Hospitals, during the period from the years 2000 to 2013 were reviewed. Medical records of the patients were revised and data from patients with any clinical, pathological and radiological findings confirming the presence of pleuropulmonary system affection were analyzed.

Results: Pleuro-pulmonary involvement occurred in 265/402 (65.9%) patients. Pleurisy was the most common clinical finding in 163 (61.5%), pulmonary infection in 57.7% and pleural effusion in 24.5% cases. Less common manifestations were pulmonary hypertension (8.3%), interstitial lung fibrosis (4.2%) and diffuse alveolar haemorrhage (n = 8; 3%). The most common clinical symptoms were pleuritic chest pain (50.9%) and cough (49.1%). The most common auto-antibodies were antinuclear antibodies (ANA) (94.7%) and anti-dsDNA in 159/183 (86.8%) cases. In the present study, the muco-cutaneous manifestations was significantly associated with pleuro-pulmonary disease ($p = 0.001$). Pulmonary affection was significantly associated with number of drug intake ($p = 0.003$). Chest infection was significantly related to the presence of other non pleuro-pulmonary infections ($p = 0.034$). Pulmonary infections were more evident in patients with pleural effusion ($P < 0.001$), CNS manifestations ($p = 0.002$) and positive ANA ($p = 0.007$). The number of drugs taken was significantly associated with the incidence of chest infections ($p = 0.002$).

Conclusion: Pleuro-pulmonary system is one of the most commonly affected systems in SLE. Pleurisy was the most common clinical finding followed by pulmonary infection.

1. Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disorder with a complex, multifactorial etiology. It is characterized by multisystem microvascular inflammation with the generation of autoantibodies. Although the specific cause of SLE is unknown, multiple factors are associated with the development of the disease [1]. In 1904, Sir William Osler reported a 24 year old woman with bilateral pulmonary consolidation and haemoptysis associated with skin rash, anaemia and nephritis [2,3]. Later on, several distinct pulmonary manifestations with SLE have been reported with a variable incidence [4]. Pulmonary involvement occurs more commonly in SLE than in any other connective tissue disease [5].

Lungs are commonly involved among the other organs in SLE. Damage and dysfunction are mainly mediated through the action of autoantibodies and immune complex formation. About 50% of patients with systemic lupus erythematosus will show signs of involvement of the lung, its vasculature, the pleura, and diaphragm at some time during the disease course [6]. Pleuritic chest pain, coughing, and shortness of breath are often the first clues to the lung involvement of SLE [4]. Pulmonary involvement is usually in the latter course of the disease [7]. However, pulmonary illness may be the presenting manifestation of SLE [8]. Any part of the pulmonary system can be affected including airways, lung parenchyma, pulmonary vasculature, pleura and diaphragm [7,9]. The pulmonary manifestations may range from sub-clinical abnormalities to life threatening disorders [10]. In an autopsy study of 90 patients diagnosed with SLE, the most frequent findings were pleuritis (78%), bacterial infections (58%), alveolar haemorrhage (26%), followed by distal airway alterations (21%), opportunistic infections (14%) and pulmonary thromboembolism (8%), both acute and chronic [7].

Previous studies [11–16] carried out in the Middle East have suggested the rate of pulmonary involvement in SLE to range from

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* Corresponding author.
E-mail address: rashagheith@hotmail.com (R.E. Gheith).

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4.9% to 30%, with the most common pulmonary abnormality reported to be pleural effusion. Alamoudi and Attar [17] recently conducted a retrospective study analyzing the data of 184 SLE patients admitted to King Abdel Aziz University Hospital assessing the prevalence and independent risk factors for SLE-associated lung abnormalities; pulmonary involvement was present in 33% of the studied patients.

The aim of this work was to study and analyze the prevalence and clinical features of different pleuro-pulmonary manifestations in Egyptian SLE patients following-up at the Rheumatology Inpatient Department, Faculty of Medicine, Cairo University Hospitals.

2. Patients and methods

The study included all the SLE patients admitted to the Rheumatology and Rehabilitation Inpatient Department, Faculty of Medicine, Cairo University Hospitals, during the time period from years 2000 to 2013. We analyzed the medical records of the patients admitted from year 2000 to 2010 and newly admitted SLE cases during the time period of the study (2011–2013) were examined and their history further analyzed. This investigation was conducted in accordance with the Declaration of Helsinki and was approved by the local Ethical Committee of the Rheumatology Department, Faculty of Medicine, Cairo University. Patients included during the study period gave their informed consent to be enrolled in the study.

A chart review was carried out for all the included SLE patients who fulfilled the 1982 revised ACR criteria for the diagnosis of SLE [18]. Demographic data such as age, sex, criteria for diagnosis and any organ or system affected were documented and recorded. The following symptoms, especially those of relevance to the chest were taken into consideration and recorded: cough, shortness of breath, haemoptysis, fever, musculoskeletal pain and pleuritic chest pain. All chest X-rays and high resolution computerized tomography (HRCT) scans were reviewed by a senior radiologist. Furthermore, the presence of the following pleuro-pulmonary manifestations was specially recorded: pleural thickening/effusion, pleurisy (based on the presence of pleuritic chest pain with pleural rub or effusion, or pleural thickening), pneumonia, pneumonitis, interstitial lung disease, bronchiectasis, diaphragmatic dysfunction, pulmonary embolism (PE), adult respiratory distress syndrome (ARDS), diffuse alveolar haemorrhage (DAH), organizing pneumonia and pulmonary oedema. Echocardiography and pulmonary function tests available for the patients were reviewed and the presence of pericardial effusion was also recorded. Only patients with any clinical, pathological and radiological findings confirming the presence of pleuro-pulmonary system affection were included in the study and further analyzed. Accordingly out of 402 patients, the data of the 265 suffering from pleuro-pulmonary manifestations were furtherly analyzed. All pertinent demographic, clinical, laboratory, radiologic and therapeutic data of these patients were abstracted.

2.1. Statistical analysis

All data analyses were performed using statistical package for social sciences (SPSS, SPSS Inc., Chicago, IL, USA; version 16). Means with standard deviations were calculated and range presented for quantitative data. Comparisons between quantitative variables were done using the non-parametric Kruskal-Wallis and Mann-Whitney tests. For comparing categorical data, Chi square test was performed. Exact test was used instead when the expected frequency was less than 5. Results were considered significant at p-value < 0.05.

3. Results

Out of the 402 SLE patients' records, 265 (65.9%) had a confirmed pleuro-pulmonary system involvement and were further analyzed. They were 247 (93.2%) females and 18 (6.8%) males with a female: male 13.7:1. Their mean age was 29.7 ± 8.8 years. Their mean disease duration was 10.5 ± 4 years. Table 1 demonstrates the demographic and clinical characteristics of the SLE patients with pleuro-pulmonary involvement. Muco-cutaneous manifestations, arthritis and lupus nephritis (LN) were the most frequent disease manifestation being present in 90.1%, 70.6% and 64.5% of the patients respectively.

Pleuro-pulmonary manifestations were the presenting SLE features in 15 (3.7%) patients in the form of pleural effusion in 4 and pleurisy in 11. 192 (72.5%) cases were suffering from pleural manifestations while 157 (59.2%) suffered from pulmonary manifestations. Isolated pleural and pulmonary diseases were present in 108 (40.8%) and 73 (27.5%) patients respectively. 84 (31.7%) patients had manifestations of both pleural and pulmonary disease. 244 (92.1%) patients were symptomatic while only 21 (7.9%) were asymptomatic. The most common symptoms were pleuritic chest pain in 50.9%, cough in 49.1% and fever in 25.7%. Table 2 demonstrates the frequencies of pleuro-pulmonary symptoms, autoantibodies and medications used by the studied lupus cohort. Table 3 demonstrated the frequencies of the pleuro-pulmonary manifestations and diagnoses in the studied patients. Pleurisy was the most common manifestation being present in 163 (61.5%) cases while pulmonary infection was the 2nd most common pleuro-pulmonary manifestation detected in 153 (57.7%) throughout the course of the disease.

In the present study, the muco-cutaneous manifestations was significantly associated with pleuro-pulmonary disease ($p = 0.001$). The incidence of chest infections was significantly associated with other non pulmonary infections ($p = 0.034$). Pulmonary infections were more evident in patients with pleural effusion ($p < 0.001$), CNS manifestations ($p = 0.002$) and positive ANA ($p = 0.007$)

Abnormal chest X-rays were found in 102 (38.5%) patients in the form of pleural effusion (24.5%), pericardial effusion (22.6%), elevated copula of the diaphragm (4.9%) and atelectatic bands (4.2%). Less common findings were consolidation, pneumonitis,

Table 1
Demographics, clinical and laboratory characteristics of the 265 systemic lupus erythematosus patients with pleuro-pulmonary involvement.

Parameter mean ± SD or n(%)	SLE patients with PP involvement (n = 265)	
Age (years)	29.7 ± 8.8	
Gender	247	(93.2)
Females		
Males	18	(6.8)
Disease Duration		10.5 ± 4
Manifestations		
Fever	158	(59.6)
Muco-cutaneous	239	(90.1)
Arthritis	187	(70.6)
Nephritis	171	(64.5)
CNS	53	(20)
Non-chest infection	46	(17.4)
Laboratory		
– Leucopenia	100	(37.7)
– Anaemia	150	(56.6)
– ANA	251	(94.7)
– Anti-dsDNA	159/183	(86.8)

SLE: systemic lupus erythematosus, PP: pleuro-pulmonary, CNS: central nervous system, ANA: antinuclear antibodies, ds-DNA: double stranded deoxy-ribonucleic acid.