Pulmonary Involvement in Childhood Systemic Lupus Erythematosus

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Abstract

Twenty-four children (aged 6-15 years, M : F = 1 : 11) with systemic lupus erythematosus (SLE), who had respiratory symptoms, were retrospectively reviewed. Chest radiographs obtained from all patients revealed pleural effusion in 13, alveolar infiltration in 9, pericardial effusion and cardiomegaly in 6, interstitial infiltration in 4, hilar adenopathy in 3, lung abscess in 2 and pneumatocele with pneumothorax in 1. Etiologic organisms were identified in 7 cases; (3 cases of nocardia isolated from pleural effusion and sputum, 2 cases of tuberculosis, 1 case with staphylococcus aureus septicemia and 1 case with salmonella septicemia). All except one patient improved with medical treatment. One patient died from pneumonitis. Although pulmonary involvement is increasingly recognized in children with SLE, neither roentgenogram nor clinical findings were specific. The differentiation of pulmonary infiltrates caused by lupus lung disease from pulmonary infection should be carefully evaluated.

Key word : Pulmonary Disease, SLE

Systemic lupus erythematosus (SLE) is characterized by the production of autoantibodies that can damage any organ system. Respiratory abnormalities are less common than other system involvements. The pathological findings in the respiratory system of adults with SLE have been well described[1-5] but relatively little attention has been paid to this problem in childhood[6-10]. In this study, we report our experience in 24 children with SLE who had respiratory symptoms and were evaluated for possible respiratory system involvement.

PATIENTS AND METHOD

SLE was diagnosed according to the revised criteria for the classification of systemic lupus erythematosus, 1982[11]. From 1979 to 1996, 24 children with SLE, who had respiratory symp-

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toms, were evaluated for respiratory involvement at Department of Pediatrics, Ramathibodi Hospital. The evaluation included physical examination and chest X-ray in all cases, sputum and/or pleural fluid cultures in particular cases suspicious of infection.

RESULTS

Of 24 patients, 22 (91.7%) were females. Ages ranged from 6 to 15 years, with the mean of 11 7/12 years. The onset of pulmonary symptoms ranged from 20 days to 3 years after the onset of SLE, with the mean of 8 months. Physical examination and radiological findings are shown in Table 1. Respiratory symptoms generally consisted of one or more of the following: cough, dyspnea, tachypnea (respiratory rate > 40 breaths per min) and chest pain. Abnormal respiratory examination included crepitations, rhonchi or diminished breath sound. It is of note that cough was the most common respiratory symptoms presenting in 17 patients (71%) and chest pain was the least common symptom presenting in only 5 patients (21%). Crepitations were observed in 15 patients (62.5%).

Chest X-ray examinations revealed abnormalities in all patients including pleural effusion in 13 (54.2%), alveolar infiltration in 9 (37.5%), pericardial effusion and cardiomegaly in 6 (25%), interstitial infiltration in 3 (12.5%), hilar adenopathy in 3 (12.5%), lung abscess in 2 (8.3%) and pneumatocele with pneumothorax in 1 (4.2%). No respiratory signs and symptoms were specific for any radiologic findings. In 5 cases presenting with chest pain, chest radiography revealed pleural effusion in 4 cases and lung abscess in one case. Diminished breath sound was observed in patients either with or without pleural effusion. Not all patients with crepitations had infiltration on the chest radiographs.

Pleural fluid and sputum culture were performed in 13 and 24 patients respectively. Nocardia was identified in 3 cases and tuberculosis in 2 cases. Blood culture was positive in 2 patients, one for staphylococcus aureus and the other for Salmonella group D. All patients except one were recovered from the pulmonary insults caused by either infectious agents or underlying SLE. One died from pneumonitis and was suspected of having candida pneumonia as the terminal event.

DISCUSSION

The prevalence of pulmonary involvement in childhood SLE reported varies considerably from 5 per cent to 77 per cent[8,12–14]. Respiratory abnormalities include interstitial or localized infiltrations, pleural effusion, restrictive lung disorder and dysfunction of the diaphragm. There are, however, rare instances in which pulmonary symptoms occur at the onset of disease in the absence of other organ system involvement. The onset of pulmonary symptoms in this study ranged from 20 days to 3 years after the onset of disease which was similar to other reports[12,15,16]. In our study, cough, tachypnea and chest pain were common presenting pulmonary symptoms which may result from pleuritis, pleural effusion and pneumonitis[12,14].

Nadorra et al[14], reported the pathological findings in the lung and related organs of 26 patients with SLE, with onset of disease before 20 years of age. Chronic interstitial pneumonia was present in all 26 patients and was severe in 5. Acute pneumonia was present in 20. Alveolar hemorrhage was found in 18 patients and pulmonary edema was found in 13 patients.

In our report, 13 out of 24 (54%) had pleural effusion in which some were bilateral. Nine out of 24 cases (37.5%) had alveolar infiltration shown in the chest X-ray. The pulmonary infiltration that is caused by lupus lung disease may be difficult to differentiate from pulmonary infection.

Among 3 cases of nocardia, one was isolated from pleural effusion and two were isolated from sputum of the patients with lung abscess. This organism is increasingly recognized as a significant opportunistic infection in immunologically incompetent hosts[17]. All 3 patients were successfully treated with trimethoprim-sulfamethoxazole. Unfortunately one patient died six months later from meningitis and acute renal failure.

In the 2 cases with tuberculosis, one presented in concomitant with the SLE disease and the other developed 3 years after the onset of SLE. Both had positive acid-fast bacilli stained sputum and responded well with antituberculous therapy. The association of SLE and tuberculosis was reported and the incidence varied from 3.6 per cent to 19 per cent[18-20]. The onset of tuberculosis can precede that of SLE, be concomitant with, or occur later in the course of SLE evolution.

The case with Staphylococcus septicemia presented with superficial abscess and pneumatocele on the chest X-ray. Pneumonectomy was performed in order to control lung infection and correct abnormal gas exchange. The patient recovered after

Table 1. Clinical and laboratory data of patients.

Patient number	Sex	Age (yr) of onset of		Respiratory symptoms/signs	Radiological findings
		SLE	Lung disease		
1	F	12	14	c, d, t / crep, di	- Interstitial infiltration
2	F	13	13 6/12	c, d, t / crep	- Interstitial infiltration
3	F	9	12	c / crep	- Alveolar infiltration
4	M	10	10 3/12	d, t / crep	- Alveolar infiltration
5	F	13	13	c / crep, di	- Alveolar infiltration - Rt paratracheal node enlargement
6	F	13	13	c / crep	- Combined interstitial and alveolar infiltration
7	F	10	12	c / crep	- Combined interstitial and alveolar infiltration - Lung abscess at RLL
8	F	12	12 1/12	c, d, t / crep, di	- Alveolar infiltration - Pleural effusion - Pneumothorax - Pneumatocele
9	F	13	13	c, d, t, ch / crep, di	- Alveolar infiltration - Rt pleural effusion - Cardiomegaly - Pericardial effusion
10	F	14	14	d, t / crep	- Alveolar infiltration - Minimal pleural effusion, pericardial effusion - Rt hilar node enlargement
11	F	14 9/12	15	d, t / crep	- Alveolar infiltration - Pleural effusion - Cardiomegaly
12	F	13	13	c, t / crep	- Lt pleural effusion - Pericardial effusion, alveolar infiltration, atelectasis
13	F	14	15	c , t, ch / di	- Rt pleural effusion
14	M	9	11	c, ch / crep, di	- Lt pleural effusion
15	F	9	9 3/12	c, t / crep, rh, di	- Rt pleural effusion
16	F	13	13 4/12	c, d, t, ch / di	- Pleural effusion - Cardiomegaly - Pericardial effusion
17	F	6	6	c / di	- Bilateral pleural effusion - Segmental atelectasis - Fibronodular infiltration both lower lobes - Multiple calcified hilar nodes bilaterally
18	F	12	12 1/12	c, d / crep	- Bilateral pleural effusion
19	F	12	13	c, d, t / crep, rh	- Bilateral pleural effusion - LLL atelectasis - Pericardial effusion
20	F	7	7	-	- Cardiomegaly
21	F	14	14	d, t	- Cardiomegaly
22	F	9	10	-	- Pericardial effusion
23	F	14	14 5/12	c, t, ch / di	- Lung abscess with air fluid level
24	F	12	14	-	- Prominent main pulmonary artery

Respiratory symptoms

c =cough
d = dyspnea
t = tachypnea
ch = chest pain

Signs

crep = crepitation
di = diminished breath sound
rh = rhonchi

treatment with intravenous antibiotics and was discharged home safely. Another case with Salmonella group D septicemia presenting with interstitial pneumonia was treated successfully as well.

Chest X-ray and echocardiography of 6 patients (24%) revealed cardiomegaly and pericardial effusion. Two patients had signs and symptoms of cardiac tamponade, for which pericardial tapping and pericardial drainage were subsequently performed. No etiologic agents of pericardial effusion could be identified. This may be caused by the involvement of endocardium, myocardium and pericardium by lupus itself or other associated abnormalities, i.e. anemia, hypertension and renal abnormalities. All responded well with treatment.

Interstitial lung disease, which commonly occurrs in viral, mycoplasmal, or chlamydial infection were found in 3 out of 24 patients (12.5%). The frequency of interstitial lung lesion in SLE was reported differently. In one study the reported frequency was as high as 98 per cent[21], while other autopsy reports have recorded 13-80 per cent occurrence of this lesion[22-24]. Since interstitial pneumonia can easily be obscured by other pulmonary lesion, there may be a tendency to understate this lesion in patients in whom acute pneumonia or pulmonary hemorrhage are also found. Lung biopsy, which may be necessary to exclude infection, was recommended as the high diagnostic yield and low morbidity in the evaluation of pulmonary disease in SLE[25]. The pathologic finding of primary lung disease in SLE usually shows alveolar damage with interstitial edema, hyaline membranes and perivascular lymphocytic and plasma cells infiltrates both in adults and children[4,14,26]. This primary lung disease is almost always dramatically responsive to high dose steroid therapy and should be differentiated from infectious process[12,27].

In conclusion, respiratory involvement in childhood onset of SLE is not uncommon. It may be insidious or presented as a life threatening event. Pulmonary symptoms are present in the majority of children at some time during the disease course. Infection was one of the most important causes of lung lesions and can cause diagnostic confusion. Effort should be exerted promptly to determine as accurately as possible the cause of pulmonary symptoms in the patients with SLE. Lung biopsy should be done when noninvasive examinations are uninformative since definite diagnosis and treatment are crucial to the survival of SLE patients.

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ปัญหาทางระบบหายใจในเด็กที่เป็นโรคเอสแอลอี

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ผู้วิจัยได้ทำการศึกษาย้อนหลังในผู้ป่วยเด็กที่ได้รับการวินิจฉัยว่าเป็น systemic lupus erythematosus (SLE) และตรวจพบว่ามีอาการหรือความผิดปกติทางระบบหายใจที่มาตรวจรักษาที่ภาควิชากุมารเวชศาสตร์ โรงพยาบาลรามาธิบดี ตั้งแต่ปี พ.ศ.2522 จนถึง พ.ศ.2539 พบผู้ป่วยทั้งหมด 24 ราย ความผิดปกติที่พบจากการตรวจภาพรังสีทรวงอกมีดังนี้ พบน้ำในเยื่อหุ้มปอดทั้งหมด 13 ราย, alveolar infiltration 9 ราย, pericardial effusion และหัวใจโต 6 ราย, interstitial infiltration 4 ราย, hilar node โต 2 ราย, ฝีในปอด 2 ราย และ 1 รายพบลมในเยื่อหุ้มปอด ตรวจพบเชื้อที่เป็นสาเหตุของโรคทางระบบหายใจทั้งหมด 7 ราย พบ nocardia จากน้ำในเยื่อหุ้มปอดและเสมหะ 3 ราย เชื้อวัณโรคจากเสมหะ 2 ราย, เชื้อ Staphylococcus aureus และเชื้อ Salmonella group D ในเลือดอย่างละ 1 ราย ตามลำดับ ผู้ป่วยทุกรายตอบสนองต่อการรักษาดี มีเพียงรายเดียวที่เสียชีวิต การวินิจฉัยหาสาเหตุของความผิดปกติทางระบบหายใจในผู้ป่วย SLE โดยใช้ภาพรังสีทรวงอกและอาการหรืออาการแสดงทางคลินิกเพื่อแยกแยะระหว่างความผิดปกติที่เกิดจากโรค SLE เอง หรือจากการติดเชื้ออื่น ๆ เป็นสิ่งที่จะต้องพิจารณาด้วยความระมัดระวัง

คำสำคัญ : ระบบหายใจ, เอสแอลอี

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