

Case Report

## Pulmonary Round Lesions in a Twelve-Year-Old Girl with Systemic Lupus Erythematosus

A. Gheisari MD\*, T. Ajoudani MD\*

### ABSTRACT

Systemic lupus Erythematosus (SLE) is a disease with different immunologic abnormalities affecting multiple organs and systems. We describe a case of SLE with multiple pulmonary round lesions, who was admitted due to hemoptysis, mild dyspnea, chest pain and gross hematuria. The patient favorably responded to intravenous immunoglobulin (IVIG).

**Key words:** SLE, Pulmonary round lesion, Chest pain, Dyspnea, Hemoptysis

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Systemic lupus Erythematosus (SLE) is a disease with different Immunologic abnormalities involving multiple mechanisms of dysregulation<sup>1</sup>.

Various autoantibodies have been detected in the sera of SLE patients<sup>1</sup>. Lungs can be affected by immune complex mechanisms followed by inflammation and possibility fibrosis<sup>1</sup>.

Furthermore, acute insult from pulmonary embolism, capillary leak, weakness of diaphragm and serositis may occur<sup>2,3,4,5</sup>.

Pulmonary round lesion is not a common manifestation of SLE.

It may occur due to different etiologies. Here we describe multiple pulmonary round lesions in a twelve-year-old girl with a completely different pattern from common SLE pulmonary lesions.

The patient favorably responded to IVIG.

### Case Report

The patient was a twelve-year-old girl who was suffering from SLE. After performing kidney biopsy, class IV SLE nephritis was confirmed. Histopathological study of the kidney

specimen revealed typical wire loops, and activity index and chronicity index were both equal to 3/12.

Early, at the time of diagnosis of nephritis, the patient received pulse methylprednisolon and the first dose of a six-dose-course of intravenous cyclophosphamide. This therapeutic regimen was followed by oral prednisolon therapy. Five weeks later, while the patient was on 60 mg/m<sup>2</sup>/EOD prednisolon, she demonstrated new symptoms including bloody sputum (mild hemoptysis), blistering lesions on the lips, peripheral edema, gross hematuria and low-grade fever. In spite of these symptoms, however her general condition was good. The first chest X-ray revealed multiple round lesions.

Appropriate antibiotic therapy was started to cover staphylococci and gram-negative bacteria. At first vancomycine plus ceftriaxone were prescribed with doses of 40mg/kg/day and 75 mg /kg/day respectively.

\*Assistant Professor of Pediatric Nephrology, Department of Pediatrics, Isfahan University of Medical Sciences.  
Correspondence to: Dr. Alaleh Gheisari, Department of Pediatrics, Al-Zahra University hospital, Isfahan, Iran.  
E-mail: gheisari@med.mui.ac.ir

Furthermore, acute onset of disease, good condition of the patient and lack of symptoms such as hypoxemia, respiratory distress and severe hemoptysis, all decreased the possibility of Bronchiolitis obliterans, fungal infections, tuberculosis and lupus pneumonitis (table 2).

We continued high dose of oral prednisolon and administered the second dose of intravenous cyclophosphamide.

Also, we changed the previous antibiotics to clindamycin and penicillin to cover anaerobic bacteria as well. These drugs were administered with doses: 25mg/kg/day and 200000 unit/kg/day respectively. Despite these treatments, the number of round lesions in the lungs increased. This led us to administer IVIG with a dose of 400mg/kg/day for four consecutive days.

Three days after starting IVIG, no further pulmonary lesions appeared. On the fifth day of IVIG therapy, hemoptysis and chest pain subsided.

Finally, after two weeks of administering IVIG, round lesions were completely cleared from the chest X-ray.

### Lab data

The following data were obtained from different laboratory and imaging examinations:

Chest X-ray revealed multiple round nodules in both lungs, with cavitation in some of them. One coined lesion with a diameter of 4 cm in the left lower lobe with well defined borders without calcification or rib destruction was noted. Also calcified axillary lymph nodes were obvious.

**Chest CT:** Showed multiple round lesions with probability of necrotic lesions due to Wegener's Granulomatosis disease without signs of abscess or infiltration. However confirmation of necrosis needed biopsy.

WBC= 16500/mm<sup>3</sup>, PMN= 70%, lymph= 22%

Cr= 0.6 mg/dL, Na<sup>++</sup>= 132mEq/dL,

K<sup>+</sup>= 4.5mEq/dL, Ca<sup>++</sup>= 8.5mg/dL

CRP= Negative (checked two times),

ESR 1<sup>st</sup> hr=90 mm

C<sub>3</sub>= 90 (normal range)

Anticardiolipin Ab= Normal range

Antiphospholipid Ab= Normal range

ANA= Positive, Anti ds DNA= strongly positive

ANCA= Negative (checked twice)

**Table 1.** Clinical and Para clinical findings of the patient.

	T (°C)	BP (mmHg)	O <sub>2</sub> - sat. (%)	General condition	New sign / symptom	CXR findings	Treatment
Day of admission	38	130.75	96%	Good	Bloody cough	Patchy infiltration- one round lesion	Previous medications + initiation of antibiotics
3 <sup>rd</sup> day	36.7	125.90	95%	Good	Decreased urine output, normal Dop- pler ultra- sound find- ings		Pulse cyclophosphamide (2nd dose)
10 <sup>th</sup> day	38	125.85	93%		Rhales in bases of both lungs	Increased lung round lesion to 5	No response to antibiotics, IVIG administration
12 <sup>th</sup> day	39	120.85	98%	Relatively bad			IVIG continued
14 <sup>th</sup> day	37	125.85	95%	Good	Decreased cough and hemoptysis and chest pain.	Decreased infiltration and number of round lesions	IVIG continued

**Table 2.** Different features of common lung diseases in SLE

	Chronic interstitial pneumonitis	Acute lupus pneumonitis	Acute alveolar hemorrhage	Bacterial infection	Post primary tuberculosis	Bronchiolitis obliterans	Fungal infection	Patients' condition
Cough	+	+		+	+	+	+	+
Hemoptysis	-		++		+			+
Pleuritic pain		++	+		+			+
dyspnoea	+	+	++			+		+
Hypoxemia			++	even in severe forms	-	++	-	-
General condition	not good	poor	very poor	poor	not good poor subacute chronic	poor -flu like syndrome	poor	Good
Onset	Chronic	acute	acute (+anemia)	acute		subacute	subacute	Acute
Prevalence	3%- 13%	1%- 4%						
Radiological data	unilateral or bilateral interstitial pattern	unilateral or bilateral patchy or lobar infiltration	diffuse alveolar hemorrhage	lobar consolidation pneumatocele abscess formation patchy infiltration	cavitations with air and fluid level tracheal deviation	chest hyper inflation-decreased vascularity in mid and lower zone	cavitation in densities with air-In CT Scan we may see aspergilloma	Bilateral multiple nodular lesion with probability of necrotic center in CT Scan

## Discussion

The mechanisms responsible for the development of an autoimmune disease such as SLE are poorly understood<sup>1</sup>. Autoantibody production and polyclonal B and T cell activities are seen but this polyclonality appears to be restricted to certain autoantigens<sup>14</sup>. While autoimmunity is the hallmark of SLE, the development of tissue injury requires the complex interaction of inflammatory cells and mediators and the vascular endothelium<sup>1</sup>. In most instances the tissue inflammation and the associated vasculitis trigger complement activation, with or without immune complex deposition.

IVIG acts via solubilizing immune complexes and providing anti-idiotypic down regulation of auto antibodies production, thus interfering with T cell and B cell signaling<sup>13</sup>.

Patients with SLE demonstrate different features of thoracic manifestations.

Acute lupus pneumonitis is characterized by alveolar damage which is probably mediated by immune complex deposition<sup>7</sup>.

Such kind of lesions appears in the form of consolidations in chest X-ray<sup>6</sup>.

The chest X-ray often shows one or more areas of bilateral consolidations<sup>8,9,10</sup>. In addition, mixed alveolar and interstitial pattern with nodules were also described<sup>9</sup>.

Bronchiolitis obliterans occurs with rheumatoid disease.

It is characterized with persistent non-productive cough, flu like syndrome, decreased lung vascularity in mid and lower zones, almost always with hypoxemia and in 80% of cases bilateral patchy ground glass densities in CXR<sup>11,12</sup>.

However clinical and radiological pulmonary findings in this case did not match any common manifestations of lung diseases in SLE.

Furthermore, she had good general condition in spite of bilateral multiple round lesions, (Table 2).

Based on probable mechanisms and as we received no response to previous medications, we prescribed IVIG.

The effectiveness of IVIG in the management of lupus induced thrombocytopenia, dermatitis, nephritis and lung disease were already described<sup>15</sup>.

We concluded that our patient's signs and symptoms might be due to Antigen-Antibody complex or complement activation and therefore T cell activation. The favorable response to IVIG further supported the probability of the disease being produced by antigen-antibody complex or complement activation. However we needed lung biopsy to prove necrosis and the patient's parents did not permit us to perform it.

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