

Lupus and the Lung

By: *Thomas J. Lang, MD, PhD*

Assistant Professor, Division of Rheumatology, University of Maryland School of Medicine & Baltimore Veterans Administration Medical Center

Lupus can affect various tissues of the lung including the pleura, which covers the lung, the blood vessels of the lung, and the actual lung tissue. By far the most common lung manifestation of lupus is **pleuritis**. Pleuritis involves the inflammation of the thin tissue-like layer which covers the lung and lies between the lung and the inside of the chest wall. When this tissue, the pleura, becomes inflamed, pain is produced when one breathes. This is because breathing produces movement of the inflamed pleura relative to the wall of the chest. Typically an individual will complain of chest pain, which is worsened when taking a deep breath. This is called pleuritic chest pain. Pleuritis can also result in the accumulation of fluid between the lung and the chest, and this is called a pleural effusion. When large amounts of fluid accumulate, it can contribute to the sensation of breathlessness sometimes made worse when lying down. Pleuritis has been described in 20 to 60% of lupus patients, making it a very common diagnosis. Treatment depends on the severity of the symptoms. Frequently the use of a non-steroidal anti-inflammatory is adequate to treat the pain symptoms until inflammation resolves. Occasionally the initiation of an increased dose of corticosteroids such as prednisone is required. Rarely, if fluid accumulation is large enough to cause shortness of breath, drainage of the fluid is required to relieve symptoms. Pleuritis is not a life-threatening condition, but primarily results in significant discomfort.

While pleuritis involves the tissue that covers the lungs, lupus can also affect the lung tissue itself in a number of ways. Most of these are fairly rare. **Chronic interstitial pneumonitis** is frequently without symptoms and is rarely severe. It is due to inflammation of the lung tissue and its primary symptom is shortness of breath. Disease can be found on x-rays of the chest, or with specific lung function tests. Though it can progress to severe disease, symptoms are commonly mild or absent. A more severe effect of lupus on the lung is called **acute lupus pneumonitis**. Individuals experience the sudden development of cough, shortness of breath, chest pain, and fever. Its symptoms are very similar to the pneumonia caused by an infection. Though this is a rare manifestation of lupus it can be severe and life-threatening. Because it looks so much like an infectious pneumonia, whether an infection is present needs to be determined before treating. If no infection is present, it is generally treated with high doses of corticosteroids. Another rare but serious complication of lupus is **acute alveolar hemorrhage**. The individual with this will develop bleeding within the air sacs of the lung, called alveoli, and will experience sudden shortness of breath, and if the bleeding is severe enough, may cough up blood. Though this most commonly occurs in someone already diagnosed with lupus, for nearly 20% of individuals with bleeding it is the first manifestation of lupus.

Lupus can cause **pulmonary hypertension** similar to the high blood pressure which is so common, except it only involves the blood vessels in the lung. It is relatively rare in lupus. Most patients with this also have Raynaud's phenomenon. Individuals complain primarily of shortness of breath. Pulmonary hypertension can be a severe problem because it can lead to

heart failure and severe shortness of breath requiring the use of oxygen. A second way in which lupus can affect the blood vessels of the lung is the development of **blood clots**.

Symptomatically individuals complain of sudden onset of shortness of breath with occasional chest pain. If these symptoms are experienced, the individual should seek immediate medical evaluation. Blood clots are associated with the presence of lupus anti-coagulant, which is an abnormal antibody that increases the clotting of blood. Treatment involves use of drugs such as warfarin, which reduce the ability of blood to clot.

In conclusion, since lupus can affect the lung in many different ways, if an individual develops chest pain or increased shortness of breath, they should be promptly evaluated by their physician so that more serious diseases of the lung can be quickly identified and treated.

This article is reproduced with permission from Lupus Update, quarterly newsletter of the Maryland Lupus Foundation, Volume 117, March 2003.