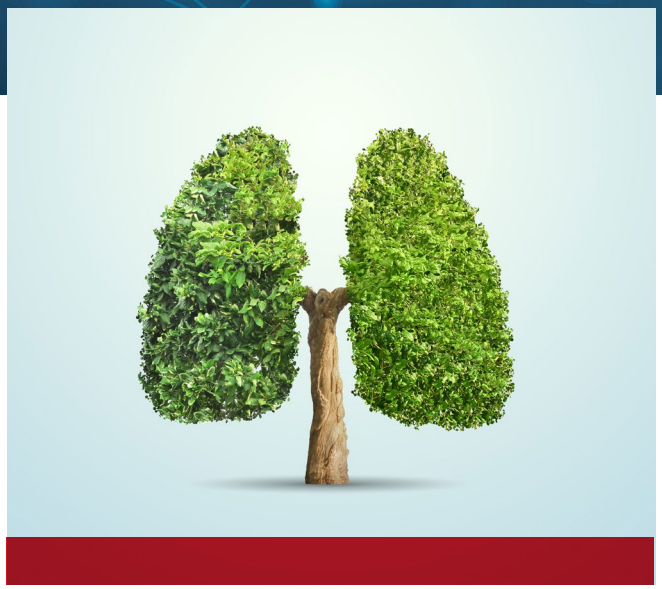
Scleroderma Association B.C.

PULMONARY FIBROSIS IN SYSTEMIC SCLEROSIS

Dr. Sabrina Hoa, MD MSC,
Rheumatologist, clinical researcher at the CHUM Research Centre

Systemic sclerosis (SSc, systemic scleroderma) is a disease characterized by abnormalities in the functioning of small blood vessels and of the immune system, ultimately leading to inflammation and excessive fibrosis (hardening) of the skin and various organs. When the inflammation and fibrosis reach the lungs, it is called "interstitial lung disease" (ILD) or "pulmonary fibrosis".



WHO IS AT RISK OF DEVELOPING PULMONARY FIBROSIS IN SYSTEMIC SCLEROSIS?

Pulmonary fibrosis is a common manifestation of systemic sclerosis, being present in about half of the patients. However, pulmonary fibrosis is severe in only about 15% of patients. Patients who are men, of Black race, with the diffuse form of systemic sclerosis, with anti-topoisomerase I (Scl-70) autoantibodies or who have cardiac, muscular or upper gastrointestinal disease may be at higher risk of developing severe pulmonary fibrosis.

WHAT ARE THE SYMPTOMS OF PULMONARY FIBROSIS?

Pulmonary fibrosis often presents itself silently in the early stages of the disease. With more severe involvement, patients may have symptoms of fatigue, decreased exercise capacity, shortness of breath and a persistent dry cough. However, in a sedentary patient, pulmonary fibrosis may remain asymptomatic even in more advanced stages of the disease.

HOW IS PULMONARY FIBROSIS DIAGNOSED?

Physical examination may reveal abnormalities during the auscultation of the lungs with the stethoscope. A chest X-ray may reveal more advanced changes in pulmonary fibrosis, but a high-resolution chest CT scan is a better test to detect pulmonary fibrosis in its earliest stages. Pulmonary function tests (PFTs) are useful to measure the severity of lung function impairment. The six-minute walk test, during which blood oxygenation and walking distance reached after 6 minutes are measured, can also be useful in assessing the severity of the disease.

Because pulmonary fibrosis in systemic sclerosis is often silent, periodic screening is recommended in all patients, knowing that appropriate treatment started early will result in better outcomes.

WHAT ARE THE TREATMENTS FOR PULMONARY FIBROSIS?

According to current guidelines, indications for initiating treatment for pulmonary fibrosis are:

- the presence of respiratory symptoms attributable to pulmonary fibrosis;
- moderate to severe involvement as evidenced by thoracic CT scan and pulmonary function tests;
- worsening of pulmonary fibrosis as evidenced by thoracic CT scan or lung function tests.

There are now two classes of medications used in the treatment of pulmonary fibrosis associated with systemic sclerosis. First, immunosuppressive medications, such as mycophenolate mofetil (Cellcept®), cyclophosphamide and rituximab, work by decreasing the activity of the immune cells responsible for inflammation. These medications can slow the progression of pulmonary fibrosis associated with systemic sclerosis. Mycophenolate is commonly used as a first-line treatment because a randomized trial showed that it is as effective as cyclophosphamide and has a better safety profile.

PULMONARY FIBROSIS IN SYSTEMIC SCLEROSIS

Recently, a randomized trial also showed that an anti-fibrotic medication, nintedanib (Ofev®), is effective in slowing the progression of systemic sclerosis-associated pulmonary fibrosis compared to placebo. Nintedanib is now approved by Health Canada. Although this medication is new in its use in systemic sclerosis, it has already been used for several years in the treatment of idiopathic pulmonary fibrosis (IPF).

When a medication is started for the treatment of pulmonary fibrosis, the patient is assessed monthly with blood tests to detect side effects of the medication. Pulmonary function tests are repeated every 3 to 6 months to determine the effectiveness of the treatment. Success is currently defined as stabilization of the disease.

Research studies are underway to determine whether earlier treatment in the mild stage of pulmonary fibrosis could be effective in preventing more severe involvement over the years. Considering that immunosuppressive and anti-fibrotic treatments may cause side effects, the decision to begin treatment should be made after assessing the risk of toxicity of the treatment compared to the expected benefits.

To reduce the risk of complications associated with certain infections when taking immunosuppressive medications, it is recommended to get the flu shot once a year and to get vaccinated against the pneumococcal bacteria (a cause of severe pneumonia) every five years. If the patient is a smoker, it is recommended to quit smoking to avoid further damage to the lungs. In patients with gastroesophageal reflux disease (GERD), it is recommended that this condition be treated aggressively in order to prevent further damage to the lungs due to reflux and aspiration of gastric contents into the lungs.

Home oxygen administration can be used for patients with very severe pulmonary fibrosis. Finally, in very advanced cases not responding to treatment, an autologous stem cell transplant or a lung or heart-lung transplant may be considered after a detailed medical and multidisciplinary assessment.

IN SUMMARY

Pulmonary fibrosis is a common and potentially serious complication of systemic sclerosis. Careful monitoring by treating physicians and appropriate early treatment can improve the quality of life and life expectancy of patients with systemic sclerosis.