Scleroderma Quebec™

THE DIFFERENT FORMS OF SCLERODERMA

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The name **"scleroderma"** is derived from the Greek words **"sclero"**, meaning hard and **"derma"**, meaning skin. Thus, the characteristic feature of scleroderma is the hardening of the skin. Scleroderma is generally divided into two main forms: localized scleroderma and systemic scleroderma (or systemic sclerosis). Systemic sclerosis can, in turn, be classified according to the extent of skin hardening (limited or diffuse systemic sclerosis) or according to the presence of specific autoantibodies in the blood.

LOCALIZED SCLERODERMA (OR MORPHEA)

Localized scleroderma is a fibrotic disease of the skin and sometimes of the underlying tissues, but does not affect internal organs. It affects mostly children, but can also occur in adulthood. There are several forms of localized scleroderma, including circumscribed or plaque morphea (involving one or multiple well-defined, oval to round areas of skin thickening), generalized morphea (when at least 4 plaques involving at least 2 anatomical sites are present), linear scleroderma (characterized by tight, thick bands, frequently affecting extremities) and scleroderma en coup de sabre (a type of linear scleroderma that affects the forehead and scalp area on one side of the head, with resemblance to the cut of a saber). Raynaud's phenomenon is usually absent in localized scleroderma.

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SYSTEMIC SCLEROSIS: LIMITED OR DIFFUSE

In contrast, systemic sclerosis (or systemic scleroderma) is a fibrotic skin disease that can also affect internal organs (hence the term "systemic"). When fibrotic skin involvement is limited to the hands, forearms, feet, legs below the knees, face and/or neck, it is referred to as limited systemic sclerosis. When the skin involvement goes up above the elbows and knees, affecting the skin of the upper arms, thighs, trunk and/or abdomen, it is then referred to as diffuse systemic sclerosis. Involvement of the skin and internal organs is generally more common and extensive in the diffuse form of systemic sclerosis. There is also a rarer form of scleroderma called sine scleroderma, in which the skin is not affected, although there is fibrotic damage to the internal organs.

SYSTEMIC SCLEROSIS: BY AUTOANTIBODIES

Systemic sclerosis is an autoimmune disease in which the immune system becomes dysfunctional and turns against oneself. Evidence of this autoimmunity can be found by the presence of autoantibodies in the blood, i.e., antibodies directed against one's own cells. Several systemic sclerosis-related autoantibodies have been identified in recent decades and are useful in predicting potential complications of systemic sclerosis. For example, anti-centromere (or anti-CENP-B) autoantibodies are usually associated with the limited form of systemic sclerosis, a slower disease course at the onset of disease and less severe involvement of internal organs, but with more pulmonary arterial hypertension later in the course of the disease. Anti-topoisomerase I (or anti-Scl-70) autoantibodies are usually associated with the diffuse form of systemic sclerosis, a more rapidly progressive disease course at the onset of disease, and an increased frequency of pulmonary fibrosis. Anti-RNA polymerase III autoantibodies are usually associated with the diffuse form of systemic sclerosis, a more severe disease course and a higher risk of developing scleroderma renal crisis. In these patients, blood pressure should be closely monitored and corticosteroids should be avoided. These three autoantibodies are the most common autoantibodies found in systemic sclerosis, with approximately 75% of patients being positive for one of these three autoantibodies.