Introduction

Scleroderma means "hard skin." The term localized scleroderma refers to the fact that the process in this group of conditions is "localized" to the skin. Sometimes, the term morphea is used interchangeably with localized scleroderma, creating some confusion. Morphea, in fact, is only one particular variant of localized scleroderma. It is important to understand that localized scleroderma is different from the scleroderma affecting internal organs, which is known as systemic sclerosis. Localized scleroderma typically only affects the skin, although in some cases, the underlying muscle and tissue may be involved. Localized scleroderma is not a fatal disease, but quality of life is often adversely affected because of changes in the appearance of the skin, the occurrence of joint contractures, and, rarely, serious deformities of the face and extremities.

The goal of this pamphlet is to inform you about the nature, prognosis and complications of localized scleroderma and to discuss current treatment options. Because localized scleroderma often affects children, a major goal of this pamphlet is to also allay fears about this condition and to explain how it differs from systemic sclerosis.

What Is Localized Scleroderma?

Localized scleroderma is characterized by thickening of the skin from excessive collagen deposits. Collagen is a protein normally present in our skin that provides structural support. However, when too much collagen is made, the skin becomes stiff and hard.

What Causes Localized Scleroderma?

The cause is unknown. It is not infectious. It is not hereditary, though, rarely, similar problems may be present in relatives in some families. It is thought to be an autoimmune disease, but patients have no other known defect in the immune system.

Types of Localized Scleroderma

Names sometimes cause a great deal of confusion in localized scleroderma. Patients may be told they have scleroderma, which may frighten them into thinking that they have systemic sclerosis.

There are four main types of localized scleroderma; each is characterized by the shape and amount of affected skin. The four types are: morphea, generalized morphea, linear scleroderma, and en coup de sabre.

Morphea: Morphea is the most common form and presents as one or more patches of skin thickening with varying degrees of pigment changes. A violet-colored border may be seen when the lesions are still very active and extending. Sometimes, doctors will classify morphea into other sub-types according to the shape or depth of the lesions. For example, "guttate" morphea refers to "drop-like" shaped areas of thickening, whereas "subcutaneous" morphea indicates a predominant involvement of deeper tissues with relative sparing of the overlying skin.

Generalized Morphea: Generalized morphea has larger patches than morphea, often involving more of the body surface. Some patients with generalized morphea may also have a band of thickening on an arm or leg as seen in linear scleroderma. Moreover, individual patches of morphea are common in linear scleroderma. Therefore, although one type of localized scleroderma usually predominates, patients may have a combination of different types of skin involvement.

Linear Scleroderma: Linear scleroderma, as the name implies, shows a band or line of skin thickening. It may extend deep into the skin and even involve the underlying muscle. The bands of skin thickening are more common on the legs and arms and, when crossing the joints, may prevent proper joint motion. On rare occasions linear scleroderma can be a serious problem in children, especially when it extends deep into the skin Sometimes, for reasons we do not yet understand, linear scleroderma delays growth of the underlying bones in children who are still in an active growth phase.

En Coup de Sabre: Linear scleroderma on the face or scalp may appear as a white line referred to as "en coup de sabre." This is a French term meaning "cut from a sword," because of the way it looks. En coup de sabre can be very destructive, as when it results in atrophy (loss of tissue) of the face, which may involve the tongue and mouth. Rarely, the condition is associated with abnormalities in the growth of facial bones, which can potentially lead to considerable deformities.

How Is Localized Scleroderma Diagnosed?

Doctors who are familiar with scleroderma, or who are experts at examining the skin, can arrive at the diagnosis without much difficulty by simply examining the skin. In some cases, further testing may be needed to confirm the diagnosis.

Prognosis: What Will Happen?

Morphea occurs in all age groups, and tends to be more common in women.

Most patients develop only one or two patches of thickening that are frequently darker or lighter than the surrounding skin. A yellow discoloration may also occur. The changes in skin color may last for years, in spite of improvement and softening of the skin. In general, morphea tends to involve only the superficial layers of the skin. Patients with morphea do quite well, but periodic follow-up by a physician is recommended. Rarely, patients will continue to develop new spots and essentially go on to generalized morphea. This evolution to generalized morphea is more common in women.

Generalized morphea also occurs in all age groups, and patients with this type are expected to have more extensive and prolonged periods when the disease is active, in some cases lasting several years. Generalized morphea represents more than just a greater number of morphea lesions, but rather a greater tendency for the condition to spread to more areas of the skin.

The areas of skin thickening in generalized morphea tend to become confluent (joined together), occasionally involving most of the body surface, and the depth of involvement is generally greater than in morphea. Periodic follow-up by a physician is recommended.

Depending on the degree of skin involvement, patients with generalized morphea may have severe disfigurement from the changes caused by extensive skin thickening. Eventually, as with morphea, softening of the skin is expected, but the skin discoloration may last for years or may be permanent. This is because the increased pigment persists in deeper portions of the skin and cannot be removed easily by bleaching agents.

Linear scleroderma is more common in children and adolescents. About 80 percent of patients diagnosed with linear scleroderma are younger than 20. It occurs four times more commonly in women than in men. Linear scleroderma has the potential to cause serious complications.

The linear areas of skin thickening may extend to the underlying tissue and muscle in children, which may impair growth in an affected leg or arm. The surface changes in the skin are similar to those described for morphea and generalized morphea. Extensive lesions of linear scleroderma, when present across joints, can impair motion. Unless continued efforts are made to maintain full motion of the affected joint by physical therapy, this complication may be permanent.

Many patients with linear scleroderma, especially if older at the age of onset of the disease, will have only minor skin changes and minimal thickening. Linear scleroderma remains active for two to five years, but can last longer in some cases.

Sometimes patients develop recurrences after a period of seemingly inactive disease. This is more frequent in patients with "en coup de sabre."

En coup de sabre is potentially the most disfiguring form of localized scleroderma. It can be mild, with only slight atrophy (loss of tissue) of the skin. However, depending on its location on the face, it can lead to considerable problems, especially in children. It is possible that it is an entity by itself, and not truly a sub-type of linear scleroderma.

On the scalp, it can cause some hair loss. When involving the face, it can lead to indentations. The process can extend to the underlying bone. Recurrences can occur, even when it seems the disease has gone into remission.

Does It Go Away?

As a general rule, localized scleroderma is a self-

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When you become a member of the Scleroderma Foundation, you are supporting the organization's mission of support, education and research. Your donation helps pay for programs in each of those three areas, including:

- funding over $1 million in original research grants awarded to investigators annually
- helping patients and their families cope with scleroderma through mutual support groups and physician referrals
- promoting public education of the disease through patient literature, health professional seminars and publicity campaigns

Your membership gives you the following benefits:

- our quarterly magazine, the *Scleroderma Voice*. The magazine includes updates on the latest scleroderma research and treatments, profiles of patients who are overcoming their condition to live productive lives; tips on how to manage your disease
- newsletters and informational and educational offerings from your local chapter
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Please consider joining the Foundation today. A membership form is attached on the reverse side of this panel.

To become a member of the Scleroderma Foundation, fill out this form, tear at perforation and send with your check or credit card information to:

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Attn: Donations
300 Rosewood Drive, Suite 105
Danvers, MA 01923

I would like to become a member and help support the Scleroderma Foundation's efforts to improve the lives of those with scleroderma, and to assist in the search for a cause and cure. Enclosed please find my check (or credit card information) in the amount of $_____.

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limiting problem. Sometimes, new lesions may appear for a few years, but eventually, the process will subside. The one possible exception to this is en coup de sabre, which may run an unpredictable course and become active again, even many years after it first appears.

Are There Other Complications of Localized Scleroderma?

Some patients with localized scleroderma, an estimated 10-20 percent, develop joint pains (arthralgia) during the course of their disease. The pain is not limited to joints beneath the involved skin, but may involve various joints, such as knees, wrists and the spine.

Occasionally, this complication precedes the thickened skin, causing considerable confusion with rheumatoid arthritis. Eventually, the joint pains subside, even in the face of new skin involvement.

Are There Any Tests to Prove the Diagnosis of Localized Scleroderma?

The diagnosis of localized scleroderma is mainly by visual recognition, though a biopsy may often be done to show increased collagen deposits. The biopsy may help determine whether the disease is still active. However, skin lesions may extend even when the biopsy does not show any significant changes.

Several blood tests may be performed, which, when abnormal, help to determine how active the disease is and how extensive or prolonged it may become. These blood tests include the number of blood eosinophils, the amount of blood immune proteins (immunoglobulins), and various blood antibodies: antinuclear antibodies (ANA), antibodies to single-stranded DNA (ssDNA) and antihistones antibodies. These tests are not specific for localized scleroderma and may be abnormal in other conditions, including the internal type of scleroderma. However, specific antibodies that are common in systemic sclerosis, are negative in patients with localized scleroderma.

What Medicines Are Available?

There is no cure for localized scleroderma, although certain drugs and interventions may help halt the spread of the disease or its complications. Specific recommendations should be left to the judgment of the physician, who will discuss the options with the patient and his/her family. Many drugs have been used in the treatment of localized scleroderma, but none has been proven to work in a controlled study.

Many drugs, including phenytoin (dilantin), potassium paminobenzoate (POTABA), systemic corticosteroids (cortisone), antimalarials (plaquenil, chloroquine), and d-penicillamine, alone or in combination, have been used with both favorable and unfavorable results. However, because of their potentially serious complications, these drugs are reserved for patients with active, widespread and rapidly advancing disease. Many physicians continue to advocate the use of oral vitamin E, but there is no proof of its efficacy.

Penicillin and other antibiotics are sometimes used by physicians who still believe localized scleroderma may be caused by the same organism as Lyme disease, but there is no consensus on this relationship.

Methotrexate is another medication that might be useful in many patients, particularly when the skin involvement is widespread or affects the deeper underlying tissues. This medication has been used to treat other autoimmune diseases and is safe and well-tolerated. Cyclosporin, a drug that can depress the immune system, has also been used.

There is, however, definite agreement that physical therapy to preserve full motion of the affected joints is important.

Please note that this brochure is provided for educational purposes only. It is not intended to substitute for informed medical advice.

The Scleroderma Foundation wishes to thank Vincent Falanga, M.D., Boston University, for his contribution to this brochure.

Our Three-Fold Mission Is Support, Education and Research

SCLERODERMA FOUNDATION
SUPPORT · EDUCATION · RESEARCH

Support: To help patients and their families cope with scleroderma through mutual support programs, peer counseling, physician referrals, and educational information.

Education: To promote public awareness and education through patient and health professional seminars, literature, and publicity campaigns.

Research: To stimulate and support research to improve treatment and ultimately find the cause of and cure for scleroderma and related diseases.

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