

Localized Scleroderma

Scleroderma means “hard skin.” In localized scleroderma, only the skin and sometimes the underlying muscle and tissue are involved. The internal organs are spared. Localized scleroderma is not a fatal disease, but quality of life is sometimes adversely affected because of the appearance of the skin, 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 the internal form of scleroderma (systemic sclerosis).

What Is Localized Scleroderma?

Localized scleroderma is characterized by thickening of the skin from excessive collagen deposition. Collagen is a protein normally present in our skin. However, when too much of it is made, the skin becomes stiff and hard. Because it affects only the skin, localized scleroderma is very different from systemic sclerosis, which almost always involves internal organs. Although there may be isolated reports suggesting that it is possible, no well-documented evidence of the transition from localized scleroderma to systemic sclerosis exists. Patients and parents of children with this condition should rest assured that localized scleroderma remains limited to the skin and underlying muscle and tissue.

What Causes Localized Scleroderma?

No one knows. It is not infectious (it cannot spread to someone else). 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. An association between Lyme disease or tick bites and localized scleroderma has been reported, but this linkage is uncertain. There have also been reports of localized scleroderma occurring after minor trauma to the involved skin, but this may be purely coincidental.

How Many Types of Localized Scleroderma Exist?

Names 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, the internal type of scleroderma. Depending on the shape and amount of affected skin, there are four main types of localized scleroderma: 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 active and extending. Sometimes, doctors will classify morphea into other subtypes 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 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 (see below). Moreover, 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 muscle. The bands of skin thickening are more common on the legs and arms and, when crossing the joints, may prevent proper joint motion. Rarely, linear scleroderma can be a serious problem in children, especially when the process extends deep into the skin. Sometimes, for reasons we do not yet understand, linear scleroderma delays growth of the underlying bones.

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 “blow 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.

Recently it has been suggested that a condition known as eosinophilic fasciitis is also part of the localized scleroderma family of disorders. In eosinophilic fasciitis, there is initially involvement of limbs, swelling, and decreased range of

motion. The skin may also have an “orange-peel”-like appearance. Lesions of eosinophilic fasciitis may follow trauma or physical exertion. Biopsy specimens or MRI evaluation would reveal inflammation of fascia, which is the name given to tissues that surround muscles and other deeper tissues.

Regardless of the type, these variants all represent localized scleroderma. Even when most of the body surface is involved (very rare), there are no associated findings of internal organ involvement.

How Is Localized Scleroderma Diagnosed?

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

Is a Skin Biopsy Necessary?

Generally, this is done to confirm the diagnosis of localized scleroderma. A biopsy cannot distinguish localized scleroderma from systemic sclerosis, a distinction that needs to be made by other means. For a skin biopsy, the skin is first made numb by injecting a small amount of xylocaine into it. This causes a slight burning sensation, which lasts only a few seconds. Thereafter, there is no further sensation and a small sample of skin, generally about one quarter of an inch, is taken painlessly. A stitch may or may not be necessary. Generally, no further pain is experienced, even after the numbing effect of the medicine wears off. The results of the biopsy, if improperly interpreted, can cause confusion and apprehension.

Typically, the biopsy report may state “scleroderma,” which may lead patients to believe that they have the internal type of the disease. It is important to recognize that it is difficult, if not impossible, to establish whether someone has localized scleroderma or systemic sclerosis based on a skin biopsy, as other diagnostic measures are usually required in order to render a diagnosis of systemic sclerosis.

Prognosis: What Will Happen?

As a rule, patients with localized scleroderma do not have internal organ involvement. As discussed below, patients with localized scleroderma may have certain abnormal blood tests, which can cause anxiety in both patients and physicians.

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 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 (but still without internal disease). 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 disease, in some cases lasting several years. Generalized morphea represents more than just a greater number of morphea lesions. 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. Nevertheless, these patients do not have internal disease. Periodic follow-up by a physician is definitely 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 be permanent.

Linear scleroderma is more common in children and adolescents. About 80% of patients diagnosed with linear scleroderma are younger than 20 years. 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 other 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 may 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. On the scalp, it can cause some hair loss. When involving the face, it can lead to unsightly indentations. The process can extend to the underlying bone. Rarely, an area of the face, even though not showing surface changes, can become indented. Recurrences can occur, even when it seems that the process has become quiet.

Does It Go Away?

As a general rule, localized scleroderma is a self-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 years after it first appears.

Are There Other Complications of Localized Scleroderma?

Some patients with localized scleroderma, an estimated 10–20%, 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 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 both by visual recognition and biopsy. The biopsy may help determine whether the disease is still active.

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. Antibodies that are common in the internal type of scleroderma (systemic sclerosis) are negative in patients with localized scleroderma.

What Medicines Are Available?

There is no cure for localized scleroderma, although certain drugs may help halt the spread of the disease. 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 proved to work in a controlled study.

Many drugs, including phenytoin (dilantin), potassium p-aminobenzoate (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 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. This medication has been used to treat other autoimmune diseases and is safe and well-tolerated. It is important to note that patients on methotrexate should have periodic blood tests to watch for possible side effects. Oral and topical vitamin D may be helpful to some patients. There has also been evidence that specific types of ultraviolet light phototherapy, such as UVA1 and PUVA, may help the condition.

There is, however, definite agreement that physical therapy to preserve full motion of the affected joints is important. Of help, also, is the use of moisturizers for the affected skin. The skin darkening may lessen with time. Because sun exposure darkens the lesions further, the use of sun-screens and shielding of the areas from the sun are recommended. Various camouflage products are now available which, when properly applied, will render the discolored areas of skin cosmetically more acceptable. Chemical peels, laser treatment, and topical agents may decrease the amount of pigment in some patients. In patients with extensive facial involvement, corrective plastic surgery has been successful.

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Disclaimer: The information provided is for educational purposes only. Any drugs or treatments mentioned should be discussed with your own physician(s).

The Scleroderma Foundation's mission is three-fold:

- ☞ To help patients and their families cope with scleroderma through mutual support programs, peer counseling, physician referrals, and educational information.
- ☞ To promote public awareness and education through patient and health professional seminars, literature, and publicity campaigns.
- ☞ To stimulate and support research to improve treatment and ultimately find the cause and cure of scleroderma and related diseases.

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