# A Killer Disease

Richard is 32 years old and is putting up with consistent heartburn, muscle weakness, and some odd-looking patches on his skin. Being a carpenter, he passed it off as being part of the trade, until one morning he awoke to find those skin patches had turned hard and shiny. He went to his doctor and was informed that he had an incurable disease and that he probably wouldn't live to see his 10-year old graduate from high school. Devastating? Of course. The sad reality is that people receive this kind of news every day and are then forced to make major lifestyle changes to live with this disease called scleroderma.

Scleroderma, also known as systemic sclerosis, is the "hardening and thickening of the skin" (Warkentin para 1) and of internal organs, such as the esophagus, lungs, heart and blood vessels. Scleroderma is found to rapidly overdevelop collagen in the connective tissues, thus making the organs involved hard, thick and unable to perform properly (DeWitt para 2-3). The disease is found 3-4 times more often in women than men, occurs between the ages of 30 to 50 years old, and affects people of all nationalities throughout the world (DeWitt para 4). There is no known cause, but research regarding symptoms, treatment and prognosis has given hope to people suffering with this debilitating disease.

Although doctors have not yet found the exact cause of the disease, there are many symptoms associated with scleroderma (Bulpitt para 3). In 90% of victims, the symptoms appear to be associated with an autoimmune disease, in which one's own cells attacks the body (Bulpitt para 4). Other reported symptoms include swelling and inflammation of the fingers, joints, and possibly the mouth. On the arms, there are usually thick, tight patches of skin, which are quite common, and these patches become "hard and numb" (Bulpitt para 5). Furthermore, 66% of patients complain of symptoms such as chest tightness, pneumonia, and general breathing problems that are created when the lung capacity is compressed from the inflammation of tissues (DeWitt para 13).

Not only are all these symptoms prevalent, but also another very common symptom that is usually seen initially in 95% of victims is what is known as Raynaud's phenomenon (DeWitt para 7). This phenomenon occurs when blood vessels constrict in one's hands or feet from coldness or when one is under a lot of stress. The hands or feet then turn white, then blue, then finally (after the constriction is gone) red and become painfully inflamed. This whole process usually occurs well within an hour's time, but some episodes may last longer (Warkentin para 3). After enduring a few of these episodes, open sores may appear on the surface of the skin because of a lack of oxygen being carried through the constricted blood vessels. This may lead to gangrene of the limb and eventual loss of the limb (DeWitt para 7).

In addition to Raynaud's phenomenon, other common symptoms may include heartburn, weakness, carpal tunnel syndrome, and swallowing problems (Bulpitt para 6). On top of these symptoms are hypothyroidism, the decreased working of the thyroid gland, "cottonmouth," decreased eye lubrication, and "enlargement and destruction of the liver" (DeWitt para 16). The most obvious symptom is hard, taut, shiny skin, which can be seen on the fingers and face. A doctor will usually examine a small skin sample to see how thick the skin is and use this information to make a diagnosis (Warkentin para 7).

Once scleroderma is determined, treatment is then started. Unfortunately, treatment can only relax the symptoms; it cannot cure scleroderma. However, there are many options available to treat this disease, depending on the body parts that are affected and the degree to which they are affected (Bulpitt para 10). One of the treatment options is medication. Perhaps one of the most common is Prednisone, a steriod, used to reduce the painful inflammation created in the joints (Bulpitt para 14). Another common drug is D-penicillamine, which slows the spread of damaging collagen and decreases skin thickening progression (DeWitt para 19). In some severe cases, the heart and lungs are affected, and calcium channel blockers, along with rest, are prescribed (Warkentin para 9). Fortunately, there are other drugs being researched to help treat the symptoms of scleroderma.

In addition to medications, there are other alternatives doctors advise to ease the symptoms of the disease. One might benefit from physical therapy to ease joint pain allow for easier movement (Warkentin para 8). Patients suffering from the symptoms associated with Raynaud's disease may find that keeping their extremities always warm and dry greatly reduces chances of having pain and inflammation (DeWitt para 19). Those that suffer from swallowing (acid reflux) problems are urged to eat a lot of small portions throughout the day, refraining from foods that contain caffeine (DeWitt para 19). Dialysis may also need to be considered for the kidneys to function properly (Warkentin para 8).

Even with treatment, the patient's prognosis is never good. In a study performed by the Scleroderma Criteria Cooperative Study, 264 patients with scleroderma were evaluated during approximately a 5.2 year period ("Predictors" para 14). The results of this evaluation showed that 50% had died, 38% had lived, and 12% were unaccounted for (Arthritis para 16). Additional results from the evaluation showed an increased risk of death if one was older than 64 years of age, a woman, not Caucasian, a manual laborer, and/or married (Arthritis para 23). Chances of survival are even less if the kidney or heart is affected. According to a study done by the Cooperative Systematic Studies of Rheumatic Diseases Program, "12 of the 15 (80%) patients who died had at least one abnormal cardiopulmonary sign at baseline compared with 13 of 32 (41%) survivors.... Renal abnormalities were observed in 5 of 32 survivors (16%) and in 6 of 15 patients who died (40%)" (qtd. "Outcome" para 10,15). Without a doubt, there is substantial information supporting the fact that scleroderma is a serious, progressive, and a fatal disease, but let us not forget that this research is constantly being done in hopes of finding a cure. For now though, the best that can be done is to make a patient as comfortable as possible, because the disease is irreversible and the prognosis is always an early death.

Scleroderma is obviously not picky about who it will affect. Despite the symptoms, those who suffer from the disease can hold onto the hope that treatment and future research will improve their prognosis. Perhaps someday Richard will be able to see his kid graduate because he was one of the lucky recipients of a cure from this destructive disease.

**Works Cited**

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