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Systemic Sclerosis: A Multiorgan Disease, Types and Symptoms

Chateen I Ali Pambuk* and Fatma Mustafa Muhammad

College of Dentistry, University of Tikrit, Iraq

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***Corresponding author:** ChateenIzaddin Ali Pambuk, Lecturer in College of Dentistry, University of Tikrit, Iraq, Tel: 07701808805; Email: dr.chatin2@yahoo.com

Abstract

Systemic Sclerosis is a chronic condition caused by excessive secretion of collagen inside the body. Some consider it a chronic condition of the connective tissue in the body. Although it is not contagious or cancerous, it may threaten life depending on the severity of the disease. Syatemic sclerosis affects the body's organs as one of the rheumatic autoimmune diseases, which means that the body's immune system behaves abnormally. The main result of this is the so-called hardening of the skin and the appearance of infections and ulcers in many parts of the body, Lungs, kidneys, heart, intestinal tract and other organs, and yet there is no cure for hardened skin but alternative treatments are effective for some forms of disease available. This mini review aims to disclose some important aspects of this multiorgan disease.

Introduction

Systemic sclerosis is considered one of the rare immune diseases that afflict the connective tissue. Skin stiffness is the most important characteristic of this disease, although hardening of the skin may occur in another type of disease known as the sclerosis, which is different from the general sclerosis in that it does not affect the organs Interior [1]. The disease is characterized by vasoconstriction, with the proliferation of cells lining the wall and increased vascularity of constriction, which may expose the organs infected with vascular insufficiency of severe circulation in the blood, in addition to the presence of inflammation of infected organs and cirrhosis of the skin and internal organs.

The disease affects the females three to five times its effect on males, especially in the age of fifteen to forty years of age. The disease usually affects people between 30 and 50 years of age and rarely develops in children. Is not fully aware of the actual cause of the disease and genetic factors are involved in a limited extent in the cause of the disease. Infection with the cytomegalovirus may affect the immune system of the disease. A close relationship has been found between exposure to organic solvents, chemicals and general sclerosis [1,2]

Types

There are two types of the disease and the division depends on two following types of characteristics: on the area and location of the affected skin where the disease may affect a limited part of the skin such as the limbs in the bottom of the elbows and knees in the specified type, while it may affect the large area of the skin like the parties in what is Above the elbows and knees or trunk injury in the diffuse type. The face may be affected in either way. In addition to the above, the spread of the disease is characterized by the rapid onset of the disease at the beginning and the rapid development of skin injury in the first five years of the disease, and may prove the disease after this period or improve in part, in addition to the early and severe effects of the internal organs of the body That these patients are more likely than others to have the renal cirrhosis which will be mentioned later. It is also characterized by the presence of anti-topoisomerase I / Scleroderma 70 antibodies [3].

From the above we conclude that the prevalence of the spread type is indicative of the patient's poor condition compared to the specific type. On the contrary, the specific type is characterized by delayed development of the disease after a long period ranging from months to years after the occurrence of the phenomenon of Reynod with internal organs affected after a long period of onset of the disease and usually less severe impact of the spread and the presence of antibodies to Centromir Anticentromere antibodies. The average life expectancy of these patients is often greater for patients with diffuse sex [3,4].

Symptoms

I. Skin symptoms: Scleroderma is a hallmark of the disease. The patient may suffer from swelling of the skin at the beginning of the disease and then develop to cirrhosis and hardening of the skin with a change in color to light or dark or both and loss of skin moisture, which may lead to the feeling of the patient itching and skin peeling [5]. Skin stiffness may lead to narrowing of the mouth, the appearance of the teeth, and the appearance of the nose. Skin sclerosis may accompany blood vessel aneurysms that appear as red spots in the skin. This rash can also affect the mucous membranes of the digestive tract, mouth and gum. Usually, vasodilatation does not cause symptoms other than poor appearance, but in a few cases patients may cause an enlarged blood vessel of the digestive tract,

mouth and gums to cause hemorrhage, which can lead to anemia [5,6].

The patient may suffer from calcification of the skin due to deposition of calcium in subcutaneous nodules, which are often found around the elbow, fingers, knees and other joints and may cause skin ulcers and difficult to cure this type of complication. The stiffness of the limbs leads to deformity of the joints such as flexion of the fingers and knees and other joints and elbows with the inability of the entire individual hip joints, which limits the movement of those joints and lead to muscle atrophy Patients may suffer from ulcers above the great protrusion due to the tightening and dystrophy of the skin and poor circulation of the skin feeding and exposure to bruises and may be exposed to ulcers of infection and may be difficult to heal those ulcers The severity of injury to the skin is usually proportional to the severity of injury to the internal organs of the body.

Rarely, some patients suffer from general stiffness of the body and internal organs without hardening of the skin. This condition requires a good doctor to the diagnosis of the disease [2,5,6]. Vascular symptoms: Approximately 80% of patients suffer from Raynond's phenomenon, which means the contraction of the blood vessels of the fingers and others when exposed to cold weather, which leads to the transformation of the color of the fingers to the pale and then the blue color and dark red . This phenomenon may also affect the ear and nose. This is usually the first symptom of the disease and is often preceded by the occurrence of other symptoms of the disease. This phenomenon can cause finger ulcers and tissue necrosis in severe cases. These ulcers may be exposed to infection, making them very painful [7,8]. With the occurrence of this phenomenon and vascular dysfunction, the affected organ may be exposed to severe blood circulation shortage, especially to the limbs, which may lead to bone decomposition, shortness of the fingers and sometimes progressive loss of the fingers [8,9].

Symptoms of musculoskeletal injury: In addition to bone degeneration, deformation of joints and muscle atrophy caused by hardening of the skin, the patient may suffer from several other symptoms such as: 1-Joint pain, which is common in many patients.2-Inflammation and swelling of the joints. 3-Inflammation and weakness of muscles with high level of muscle enzymes in blood. Muscle weakness due to the illness and lack of use due to joint injury, malnutrition, muscle fibrosis or the use of certain drugs such as steroid hormones [8,10].

II. Lung injury: Most patients with diffuse skin sclerosis suffer from respiratory dysfunction due to the weakness of the lungs to expand due to stiffness of the trunk.Some patients, especially those with severe skin sclerosis, may suffer from elevated pulmonary artery pressure, which occurs either due to vasculopathy, accompanying pulmonary cirrhosis, or pulmonary embolism, as is common in those the presence of centromere antibodies. Increased pulmonary artery pressure may lead to failure of the right side of the heart. While those with diffused type, in particular, may suffer fromintraintresticial inflammation of the lungs, which may lead to cirrhosis of the lungs [11,12]. Other complications of the disease is the pneumonia may develop from the inhalation of contagious contents in the lungs due to reflux of the esophagus and ventricular resuscitation and bleeding of the peritoneum (bleeding in the pulmonary alveolar). Lung disease is the leading cause of death in systemic sclerosis [13].

III. Gastrointestinal infection: The digestive system is the second organ affected by the skin after the disease. Generalized sclerosis can affect any part of the digestive system from the mouth to the anus in terms of fibrosis of the wall and weakness of the ability to contract. The upper part of the digestive tract is considered to be more affected than the lower part, although treating the symptoms of the lower part is more difficult. The most common parts of the digestive system affected is esophagus and the one of the most common symptomis the difficulty swallowing and the return of gastric contents from the stomach to the esophagus, which causes pain in the chest due to the weakness of the motor capacity of the pharynx or esophagus, in the first, and dysfunction of the lower valve which leads to the inability to constrict to prevent the return of the contents of the stomach to the intestines, in the second. The reflex of the esophagus may cause a fibrosis of the lower part of the esophagus, which may lead to difficulty of swallowing. In some cases, repeated reflux may lead to cells mutating, lining the lower part of the esophagus from one type to another, which may in turn cause malignancy in the lower part of the esophagus. The patient may suffer from gastritis due to the stagnation of infectious contents in the stomach for a long periods.

Poor intestinal and bowel movements may result in an early satisfaction of the patient with swelling and chronic constipation in some cases and in other cases may lead to an increase in the proliferation of cells that causing intestinal diseases, which may lead to chronic diarrhea and malabsorption of various nutrients [14]. As for the infection of the large intestine (colon), the weakness of the worm may lead to the accumulation of waste in the intestines leading to constipation and also the occurrence of seizures of false intestinal obstruction, the patient suffers frequent vomiting and the retention of waste and gases with abdominal pain. In some rare cases, the intestinal wall may be implicated with the cause of intestinal air vesicles form in the intestinal wall known as pneumosisintestinaliscystoides. The weakness may also result in the forming the small bags or vesicles in the lining wall is Known as colonic diverticulosis, which may rupture and the air leakage to the peritoneal cavity of the abdomen. Rarely, the patient may suffer from intestinal vasculitis, which may lead to abdominal cramps after eating a meal in the so-called intestinal angina and in neglected cases may lead to intestinal gangrene [14-16].

As explained above, the general sclerosis patient may suffer from poor nutritional status and loss of many of its weight. The patient may also suffer from the dilatation of blood vessels located in the lining of the intestine, which appear in the form of a diagonal shape around the pyloric opining of the stomach gives the stomach form of watermelon shell from the inside and the vessels may expand in the form of points. These blood vessels can cause bleeding from the digestive system and anemia in some cases [15]. Some cases may complain the less control of the removal of waste due to dysfunction of the anus and some cases may suffer from

fibrosis of hepatic bile ducts, which results from the severe attack of the immune system to the liver bile ducts.

IV. Kidney injury: Kidney injury is an indicator of poor patient's future condition. Kidney injury was the most important cause of death in the past.The most common kidney disease in this disease is renal failure in patients with general sclerosis, which affects 5-10% of patients with general sclerosis, especially in the spread type and at the onset of the disease with the rapid development of skin injury and in the case of the presence of antibodies to the polymerase special enzyme with anti-RNA polymerase [8,17]. The renal crisis is characterized by a violent increase in blood pressure and associated with other body organs such as headache, visual impairment, convulsions, stroke and heart muscle failure. The level of waste increase in the blood that is eliminated by the kidneys , such as creatinine, urea, with the destruction of red blood cells in the microcircles of the kidney, causing anemia and decrease the number of platelets in the blood and the emergence of albumin and red blood cells in urine analysis [17].

The patient's condition may extend to develop kidney failure and the need for dialysis. In the past, the renal failure patient was usually killed by the disease and now the patients' future has improved significantly after the detection of the angiotensin-converting enzyme inhibitors. Kidney function in renal dialysis patients may improve to the extent that these patients stop renal dialysis even after the next three years of renal failure. This improvement occurs in about one-third of the patients. And the age of the patient and the lack of high blood pressure in the incidence of renal crisis is one of the future indicators to the inability to stop the renal dialysis.

V. Heart injury: The heart is affected mainly by late stages of general sclerosis, especially the spread type, and the impact of the heart is one of the indicators of poor future condition of the patient. The affected heart includes both the dysfunction of the heart electricity transforming system leading to cardiac arrhythmias and irregular heartbeat, as well as the involvement of the heart muscle itself due to the lack of blood circulation, which may lead to infarction, fibrosis, and heart failure [8,18].

VI. Neurological symptoms: The patient may suffer from symptoms of pressure on the peripheral nerve [8]. Dry mouth and eyes.

VII. Sexual dysfunction: Males with ED may have erectile dysfunction, which usually responds to phosphodiesterase inhibitors. Females may experience pain during intercourse due to vaginal dryness or vaginal opening [8].

Conclusion

Scleroderma is considered to be a disease that harms many organs of the body. It is therefore necessary to stay under constant surveillance and follow-up of multiple medical specialists: Rheumatologist, Cardiologist, Cardiologist, Gastroenterologist, Dermatologist and Kidney specialist. Early sclerosis can be diagnosed with frequent and permanent lung function (Spirometry), CT (CT) of the lungs, Echocardiography and kidney performance. Early treatment of scleroderma may alter its normal course and protect against complications and irreversible effects.

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