AJPMHS
GLOBAL RESEARCH NETWORK

AMERICAN Journal of Pediatric Medicine and Health Sciences
Volume 02, Issue 03, 2024 ISSN (E): 2993-2149

Clinical and Immunological Features of the Diffuse Form of Systemic Scleroderma

Mukhitdinova Gulshan Zohirovna

assistant of the Department of Internal Diseases, Faculty of Pediatrics, SamSMU

Khudoiberdiev Tokhir Abdurashid Ugli

student of 508 groups of the Faculty of Pediatrics

Bozorova Chekhroza Sodik kizi

student of group 515 of the pediatric faculty

Abstract *: In today's article, Scleroderma appears: limited; systematic.*

Systemic scleroderma is a problem associated with characteristic damage to the skin, blood vessels, musculoskeletal system and internal organs (lungs, heart, digestive system, kidneys).

keyword : *Systemic scleroderma - skin, blood vessels, musculoskeletal system and internal organs lungs, heart, digestive system, kidneys.*

Scleroderma -´ (Greek: - "hard" and - "skin") is a progressive disease from the group of collagenoses, autoimmune in nature. Scleroderma occurs: limited; systematic. Systemic scleroderma is an autoimmune disease of connective tissues associated with characteristic damage to the skin, blood vessels, musculoskeletal system and internal organs (lungs, heart, digestive system, kidneys), based on microcirculation disorders, inflammation and general fibrosis. The disease has a genetic predisposition. . However, reliable triggers for its occurrence are external harmful factors such as hypothermia, vibration at work, and past infections of the nervous system. The development of inflammation of small vessels leads to the growth of collagen and fibrous tissue around them, as well as specific changes in their walls - thickening, loss of elasticity, and perhaps even complete closure of small vessels. These changes, in turn, lead to disruption of blood supply to all organs and tissues involved in the pathological process. The lack of blood supply to the tissues leads to their thinning (for example, the mucous membranes of the esophagus and stomach) or, on the contrary, to their thickening (the walls of the alveoli in the lungs), disruption of their main functions (absorption in the gastrointestinal tract, removal of carbon dioxide by the lungs, contraction of muscle fibers).

Diagnostics

Clinical manifestations of scleroderma are very diverse, as the disease affects almost all organs and tissues. A characteristic skin lesion is seen in most patients with scleroderma. Diagnostic features are a mask-like face (greatly reduced facial expressions that give the impression of tension on the skin) and changes in the hands (thin and immobile fingers, large claws, and thickening of the terminal phalanges).A diagnosis of systemic scleroderma is definite if one "major" or two "minor" criteria are met.

"Large" criteria:

Proximal scleroderma: asymmetric thickening of the skin in the area of the fingers, extending proximally from the foot-palm joints. Skin changes can be observed on the face, neck, chest, abdomen.

"Small" criteria:

Sclerodactyly: the skin changes listed above are limited to the fingers. Digital scars are skin retraction or loss of pad substance on the distal phalanges of the fingers. Bilateral basal pneumofibrosis; mesh or linear-nodular shadows are most often detected in the lower parts of the lungs by standard X-ray examination; A type of "cellular lung" may appear.

Clinic

A change in the veins of the hands leads to the development of Raynaud's syndrome - an acute vascular spasm with coldness and pain in the fingers. Another distinctive disorder is the joint changes in scleroderma. They are manifested by inflammation with irreversible stiffness due to very rapid loss of joint mobility and the formation of so-called contractures, i.e., the growth of fibrous tissue in the joint and the loss of elasticity of its capsule. Internal organs such as the kidneys, lungs, and heart are often affected in scleroderma. Changes in them lead to a decrease in all body functions. For example, with kidney damage, it is manifested by increased kidney failure. As a result, general intoxication of the body develops along with the loss of proteins and ions. Many of the changes in scleroderma, such as muscle and bone pain, can mimic rheumatism or rheumatoid arthritis. Differential diagnosis with these diseases is carried out on the basis of X-ray examination and immune tests.

Treatment

The main means in the treatment of scleroderma are drugs that reduce fibrotic changes in blood vessels - enzyme preparations based on lidase or ronidase [the site does not work], hyaluronic acid derivatives, etc. Pharmacological treatment is combined with physical therapy and exercise.

Prognosis

The prognosis of the disease is conditionally unfavorable, modern medicine cannot eliminate the cause of the disease by acting only on its symptoms. The disease is chronic, slowly growing, adequate treatment only improves the quality of life and slows down the progression of the disease, over time the ability to work is completely lost and Microorganisms or toxins that enter the body collide with the cells and mechanisms of the innate immune system. The innate response is usually initiated when microbes are recognized by pattern recognition receptors or when damaged or stressed cells send warning signals. Innate immune defenses are nonspecific, meaning that these

systems respond to pathogens in a general way. This immune system does not provide long-term immunity against the pathogen. The innate immune system is the dominant host defense system in most organisms and the only system in plants.

Immune sensitivity

Cells in the innate immune system use pattern recognition receptors to recognize molecular structures produced by pathogens. They are mainly proteins secreted by cells of the innate immune system, such as dendritic cells, macrophages, monocytes, neutrophils, and epithelial cells, and serve to detect two groups of molecules. They are pathogen-associated molecular patterns (PAMQs)—injury-associated molecular patterns (JAMQs)—associated with host cell components that are associated with microbial pathogens and released during cell injury or cell death. Recognition of extracellular or endosomal PAMQs is mediated by transmembrane proteins known as toll-like receptors (TLRs). TLRs have leucine-rich repeats (LRRs) that give them an oblique structural motif. Toll-like receptors were first discovered in Drosophila and activate the synthesis and secretion of cytokines and other host defense programs required for innate or adaptive immune responses. Ten toll-like receptors have been identified in humans. Cells in the innate immune system contain pattern recognition receptors (PRRs) that recognize infection or cell damage. The three main classes of these "cytosolic" receptors are NOD-like receptors, RIG-like receptors, and cytosolic DNA sensors.

Innate immune cells See caption

Scanning electron microscope image of normal circulating human blood. The image shows red blood cells, a few small white blood cells, including lymphocytes, monocytes, neutrophils, and many small disc-shaped platelets. Some leukocytes (white blood cells) act as independent, single-celled organisms and are a second branch of the innate immune system. Innate leukocytes include "professional" phagocytes (macrophages, neutrophils, and dendritic cells). These cells recognize and destroy pathogens by physically attacking larger pathogens or by engulfing and then killing microorganisms. Other cells involved in the innate response include innate lymphoid cells, mast cells, eosinophils, basophils, and natural killer cells. Phagocytosis is an important feature of cellular innate immunity, performed by phagocytes that engulf pathogens or particles. Phagocytes usually scout the body for pathogens, but can also be called to specific locations by cytokines. After a pathogen is engulfed by a phagocyte, it enters a cellular vesicle called a phagosome, which then fuses with another vesicle called a lysosome to form a phagolysosome. The pathogen is killed by free radicals released by the activity of digestive enzymes or respiratory burst into the phagolysosome. Phagocytosis evolved as a method of nutrient uptake, and its role expanded into a defense mechanism by engulfing pathogens. Phagocytosis is the oldest form of host defense, as phagocytes have been identified in both vertebrates and invertebrates. Neutrophils and macrophages are phagocytes that travel through the body to hunt down pathogens. Neutrophils are normally found in the bloodstream and are the most abundant type of phagocyte, accounting for 50-60% of the total circulating leukocytes. During the acute phase of inflammation, neutrophils migrate to the site of inflammation by chemotaxis and are usually the first cells to arrive at the site of infection. Macrophages are multifunctional tissue-based cells that produce a range of chemicals, including enzymes, complement proteins, and cytokines, while also activating the adaptive immune system as antigen-presenting and scavenging cells from the body. Dendritic cells are tissues that communicate with the external environment

REFERENCES

1. Шодикулова Г. З., Мухитдинова Г. З., Атоев Т. Т. ОЦЕНКА СОСТОЯНИЯ ЦЕНТРАЛЬНОЙ ГЕМОДИНАМИКИ У БОЛЬНЫХ С ХРОНИЧЕСКИМИ БОЛЕЗНЯМИ ПОЧЕК В САМАРКАНДСКОМ РЕГИОНЕ //Достижения науки и образования. – 2021. – №. 8 (80). – С. 59-63.
2. Babamuradova Z. B., Shavazi N. N. Assessment of the efficacy and safety of biological agents in rheumatoid arthritis //Journal of Advanced Medical and Dental Sciences Research. – 2021. – Т. 9. – №. 6. – С. 26-31.
3. Babamuradova Z., Isokova S., Shopulotova Z. THE IMPORTANCE OF STUDYING THE STRUCTURE OF BLOOD SERUM IN GYNECOLOGY //Science and innovation. – 2023. – Т. 2. – №. D12. – С. 718-721.
4. Babamuradova Z. B., Shodikulova G. Z., Mirzaev O. V. Treatment of patients with undifferentiated connective tissue dysplasia in mitral valve prolapse with varying degrees of mitral regurgitation //European science review. – 2018. – №. 3-4. – С. 140-143.
5. Kizi S. I. K., Babamuradova Z. B. IMMEDIATE OUTCOMES OF UNSTABLE ANGINA IN METABOLIC SYNDROME //International Journal of Medical Sciences And Clinical Research. – 2023. – Т. 3. – №. 02. – С. 44-49.
6. Najmiddinov A., Ibragimova N., Muxitdinova G. SILIKOZ PATOGENEZIDA KREMNIY DIOKSIDI VA YALLIG‘LANISH MEDIATORLARINING O‘RNI //Евразийский журнал медицинских и естественных наук. – 2023. – Т. 3. – №. 12. – С. 63-77.
7. Nasirova A. et al. Immunodiagnosis of chronic obstructive pulmonary disease and associated bronchial asthma //Annals of the Romanian Society for Cell Biology. – 2021. – С. 13090-13097.
8. Zikriyayevna S. G., Zoxirovna M. G., To’raqulovna Q. S. PREVENTION OF CHRONIC KIDNEY DISEASE AND CHRONIC HEART FAILURE //Spectrum Journal of Innovation, Reforms and Development. – 2022. – Т. 4. – С. 408-414.
9. Zikiryayevna S. G. et al. Kidney Damage in Patients with Chronic Cardiac Insufficiency and Obesity //Texas Journal of Medical Science. – 2022. – Т. 13. – С. 72-78.
10. Shodikulova G. Z., Babamuradova Z. B., Atoev T. T. SIGNIFICANCE OF TYPE I COLLAGEN ANTIBODIES IN ISCHAEMIC CARDIOMYOPATHY //Thematics Journal of Education. – 2022. – Т. 7. – №. 3.