Journal of Contemporary Medical Education, 2022
VOL 12, NO. 11, PAGES 2

PERSPECTIVE

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Structure of Connective Tissue Diseases and its Causes

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ARTICLE HISTORY
Received: 28-Oct-2022, Manuscript No. JCMEDU-22-79865;
Editor assigned: 01-Nov-2022, PreQC No. JCMEDU-22-79865 (PQ);
Reviewed: 15-Nov-2022, QC No. JCMEDU-22-79865;
Revised: 22-Nov-2022, Manuscript No. JCMEDU-22-79865 (R);
Published: 29-Nov-2022

Description

A connective tissue disease (collagenosis) is any disease whose pathology targets the connective tissues of the body. Connective tissue is any type of biological tissue with an extensive extracellular matrix that supports, binds, and protects organs. These tissues form the framework, or matrix, for the body and are composed of two main structural protein molecules, collagen and elastin. There are many different types of collagen protein in each of the body's tissues. Elastin has the ability to stretch and return to its original length like a spring or rubber band. Elastin is a major component of ligaments (the tissue that attaches bone to bone) and skin. In patients with connective tissue disease, it is common for collagen and elastin to be injured by inflammation (ICT). Many connective tissue diseases are characterized by abnormal activity of the immune system with inflammation in the tissues due to the immune system being directed against the body's own tissues (autoimmunity).

Diseases in which there is inflammation or weakness of collagen are also referred to as collagen diseases. Collagen vascular diseases may (but not necessarily) be associated with collagen and blood vessel abnormalities that are autoimmune in nature. See also vasculitis.

Connective tissue disease can have a strong or weak hereditary risk and can also be caused by environmental factors.

Structure

Loose and dense connective tissue consists of the following three fibers: collagen fibers, reticular fibers, and elastin fibers.

Collagen fibers consist of tightly packed thin collagen fibrils that have a wavy course in the tissues. These parallel fibrils are bundles of flexible proteoglycans that offer fundamental mechanical properties. They offer flexible yet strong tensile strength resistance. Specifically, in loose connective tissue, collagen runs in a parallel course and then joins to form a larger bundle. They separate from each other and connect again at different places, creating a three-dimensional network. Dense connective tissue such as ligaments and tendons are compromised mainly by densely packed collagen fibers.

Reticular fibers, also called argyrophilic fibers, have a limited amount in the human body. They are primarily present in the basal epithelial tissue, fat cells, Schwann and muscle cells, lymphoid tissue and the endothelium of the liver sinusoids. Under the microscope, these reticular fibers are fine, dark fibrils that are continuous with the collegiate fibers described above. The arrangement of these fibers forms a network that lies beneath the basal lamina layer. There is a firm attachment of these fibers to the basal lamina, suggesting that, together with the collagen fibers, these fibers form a functional and structural unit that serves to support the tissues. The loose arrangement of these fibers also provides room for molecular movement in the extracellular fluid.

The last component to discuss is elastin fibers. These fibers have the characteristic property of elastic recoil. In loose connective tissue, elastin is typically a loose network. Their organization and distribution depends on the type of tissue. Concentric elastin fibers are present in the vessel wall to help maintain an even blood pressure. Fibers are also present in extensible and contractible organs such as the lungs and bladder.

Causes

Mixed connective tissue disease is an autoimmune disorder, although the cause is unknown. In autoimmune disorders, the immune system responsible for fighting disease mistakenly attacks healthy cells.

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In connective tissue disease, your immune system attacks the fibers that provide framework and support for the body. Some people with mixed connective tissue disease have a family history of the condition. But the role of genetics in the disease remains unclear.