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Muscles and Joints - Scleroderma

Connective Tissues

The body is comprised of millions of cells and these cells are held together by connective tissue. Early in evolution nature had to solve the problem of how cells could be held together to create a multi-cellular organism. A major solution was the building of extra-cellular (outside the cell) collagen fibrils and fibres. Collagen fibrils have the same tensile strength as steel wires and they act to hold together the cells of various tissues. There are several types of collagen and different forms are found in different parts of the body. Collagen is produced by cells called fibroblasts.

Connective tissue is also comprised of a related fibrous protein called elastin, which has elastic properties. Elastin is important in tissues that expand such as the lung and arterial wall. The other major components of connective tissue are the proteoglycans, which are sugar-rich substances that also have elastic properties and can expand and resist compression. Large amounts of proteoglycans are found in cartilage.

The fibrosis (tightening) of the skin and internal organs that occurs in scleroderma is the result of too much collagen being produced by the fibroblasts. The cause of this is unknown. The type of collagen produced, however, is normal.

Bone and Joint Involvement

Several types of joint involvement can occur in scleroderma.

1. Mild joint pain and stiffness involving the fingers, wrists, knees and ankles may occur early in scleroderma. This may be associated with generalised (overall) swelling of the fingers, the joints may be a little swollen and examination of joint fluid shows only mild inflammation. On biopsy excess collagen is found in the lining of the joint (the synovium). Sometimes calcification (calcinosis) can also occur in and around the joint. Joint erosions do not usually occur. Later flexion contractures (bowed fingers) can occur. These are caused by tight tendons, skin and fibrosis of connective tissues around the joint with increased amounts of collagen being present in these structures.
2. Occasionally a more severe joint swelling and inflammation can occur resembling rheumatoid arthritis, but this is very uncommon.
3. Because of lack of movement of the joints involved as a result of the stiffness, osteoporosis can occur but this does not usually cause symptoms.
4. The end bones of the fingers (distal phalanges) may partly become resorbed and disappear leaving shortened fingers. This is called osteolysis. This may occur in later stages of severe disease.

The joint involvement of scleroderma usually settles with physiotherapy and anti-inflammatory drugs.

Muscle Involvement

Muscle involvement is common in scleroderma.

1. The weakness and wasting of muscle commonly found in scleroderma is a result of disuse owing to joint contractures as a result of the skin involvement.

2. About 20% of patients with scleroderma develop a bland non-progressive muscle disorder (myopathy). Weakness is not usually noticed by the patient, but it can be detected on physical examination by the physician and the tests of muscle inflammation are either normal or only minimally elevated. The muscle biopsy shows areas of replacement of muscle fibres with collagen tissue without evidence of inflammation. This muscle involvement does not usually require treatment.
3. A very small minority of patients develop acute inflammation of their muscles with severe muscle pain and weakness especially around the shoulders and hips causing problems in arising from chairs and elevating the arms. This may occur in patients who have an overlap syndrome with features of scleroderma, polymyositis and S.L.E. (Mixed connective tissue disease). This form of muscle inflammation requires high dose Prednisone (Cortisone) for control.

Tendon Involvement

1. Some patients become aware of creaking noises on movement of their fingers or wrists, knees or ankles. They note a peculiar type of coarse leathery grating that may be felt over these areas during movement. The noise is due to fibrous deposits on the surface of tendon sheaths (the lining of tendons) and this often indicates a more severe form of scleroderma.
2. Symptoms of carpal tunnel compression may occur early in the disease with tingling and numbness in the hands at night or aching in the hands at night. This is caused by swelling of the tendons in the wrists causing pressure on the median nerve in the wrist. This may need treatment with a resting splint worn at night or a local steroid injection if necessary into the carpal tunnel, or surgical decompression.

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