**Understanfing Systemic Scleroderma and CREST**

Clinicians Corner Paper

A patient came into the office for an evaluation of her left foot. She reported she has also been diagnosed with CREST syndrome. She then paused and said, “Have you ever heard of CREST?” I responded that I had and in fact had one other patient who was diagnosed. She had a sigh of relief and stated, “Oh good, no one ever knows what it is, they think of the toothpaste.” This statement made me reflect on how refreshing our knowledge of different diagnoses, even if it’s not the primary, can mean to a lot of patients. Considering this interaction, I felt that highlighting a rare disease would be crucial for expanding our knowledge as clinicians.

Scleroderma is broken into localized and systemic scleroderma. Systemic scleroderma includes CREST syndrome and diffuse scleroderma. CREST syndrome is a localized variant of limited cutaneous systemic sclerosis (lcSSc) which is characterized by calcinosis causing tightness mostly in the hands and face, Raynaud’s phenomenon, esophageal dysmotility, sclerodactyl, and Telangiectasias.1,2,3 Patients can have 2-5 of these symptoms and have lower incidence of internal involvement.1,3 Diffuse scleroderma (dSSc) can occur more rapidly and can affect the organs, such as the heart, leading to heart, kidney, and lung involvement. Conditions including pulmonary fibrosis and hypertension can result as well as weakness, joint pain and myopathy. Diffuse scleroderma can worsen rapidly in the first five years. There is also another form of systemic sclerodermas which is an overlap of limited and diffuse. 1,2,4

Localized scleroderma can include morphea, which includes oval shaped patches on the trunk. Linear scleroderma includes band like lesions on the skin. This form primarily affects the skin but not the organs and may impact the joints and muscles. The use of blood tests, skin biopsy, x-rays of joints and lungs, specific organ tests such as lung function, GI series, and distinctive serum antibodies which are found in 90% of cases can aid in confirmation of scleroderma.2,5

Physical therapy can assist in the improvement of ROM and improve joint mobility in these patients. Patients can experience contractures, arthralgias and tendon friction rubs. In patients with dSSc management of ROM and stretching to decrease contracture risks is very important. Regular exercise as well as active and passive stretching are important in maintaining flexibility and have been shown effective. Skin can be sclerotic and may be sensitive to pressures placed on it. Aquatics can also be effective for exercise to aid in the sensitvity.4,5 Self-administered stretching programs have shown to improve ROM of the fingers. Manual stretching and ROM exercises which consist of exaggerated facial movement have also been effective in mouth opening which can affect oral health. Massage and joint mobilizations were shown to improve fist closure. Paraffin wax with exercise has been shown to be effective in treatment of the hands with improving extensibility of collagen. This has been shown to be more effective than exercise alone.5

Protection of swollen and painful joints can also be important and lightweight splints may be used for joint protection however dynamic splints have not been shown effective. 4,5 Cardiopulmonary involvement must be considered and may present with limitations with aerobic and endurance capacity, however resistance and stretching programs as well as breathing and aerobic exercises showed positive results.5 Patient education on taking blood pressure three times a week to assess for acute hypertension is also very important for patients.4 It is important to take into consideration the effect of these other diagnoses may have on our patients that can affect their POC and tolerance of treatment.

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