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Scleroderma

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Scleroderma

INTRODUCTION

Scleroderma is one of the least common connective tissue diseases. However, it is a clinically relevant disease to both the general practitioner and the rheumatologist because almost all patients with scleroderma experience Raynaud's phenomenon, a condition that has a high prevalence in any given patient population. Physicians must be able to distinguish between patients with primary Raynaud's phenomenon and those who either have or are at risk for developing a connective tissue disorder.

The major pathologic events in scleroderma—vascular damage, fibrosis, and vasomotor instability—result in a wide spectrum of clinical manifestations, including skin thickening, cardiac arrhythmias, respiratory insufficiency, esophageal and small intestine dysfunction, and digital ischemia. These features are related to the distribution of fibrosis and the relative ratio of fibrosis and vasomotor instability. The extent of skin involvement and pattern of internal involvement are used to classify subsets of scleroderma. By distinguishing among the subsets, the rheumatologist can anticipate problems and institute treatment to change the course of the disease, particularly as it pertains to scleroderma renal crisis and the prevention of esophageal complications.

CREST SYNDROME

INITIAL PRESENTATION

An otherwise healthy 38-year-old woman is referred to a rheumatologist by her primary care physician for evaluation of reversible color changes and pain in her fingers on cold exposure. These symptoms have occurred since her teenage years. She is planning a winter vacation at a ski resort and is concerned that her symptoms will limit her outdoor activities.

- What is Raynaud's phenomenon?
- What evaluations are appropriate for this patient?

DISCUSSION

Raynaud's Phenomenon

Raynaud's phenomenon is defined as reversible 2- or

3-phase color changes of the digits. Typically, blanching is followed by bluish discoloration, and erythema develops on rewarming. Raynaud's phenomenon has a characteristic distribution in which the most distal segments of the digits are involved to the greatest extent. Several digits may be completely affected while others are spared (**Figure 1**). Although all 3 color phases are usually noted, the diagnosis of Raynaud's phenomenon may be made if blanching plus either a blue or red phase is observed. Accompanying ischemic pain and numbness in the affected digits are common. Each episode usually lasts 5 to 30 minutes, and digital color returns entirely to normal between attacks. The features of Raynaud's phenomenon occur because of vasospasm of the vessels that provide circulation to the fingers or toes. Vasospasm can occur in response to a variety of stimuli, including cold, stress, and anxiety.

Other conditions may be confused with Raynaud's phenomenon. Blanching on cold exposure without a subsequent blue or red phase simply results from normal compensatory vasoconstriction in response to cold and is not sufficient to establish the diagnosis of Raynaud's phenomenon. Acrocyanosis is a violaceous discoloration of the entire hand or foot that extends proximal to the metacarpophalangeal or metatarsophalangeal joints and is often exaggerated by temperature extremes. Acrocyanotic changes are a result of vascular insufficiency proximal to the digits, which may occur in peripheral atherosclerotic disease. Fixed digital ischemia (lasting more than 10 to 15 minutes upon rewarming) is seen with thromboembolic disease, vasculopathy, or vasculitis.

Appropriate Evaluations

Although more than 90% of people with Raynaud's phenomenon have no underlying medical condition and are thus classified as having Raynaud's disease, all should be evaluated for underlying conditions (**Table 1**). At a minimum, an occupation/avocation history and a thorough review of systems should be performed, including questions about signs and symptoms of a rheumatic disease (such as systemic lupus erythematosus [SLE], rheumatoid arthritis, scleroderma, and polymyositis) and toxin exposure. In addition to having an increased sensitivity to stimuli, the blood vessels in connective tissue disease are often structurally abnormal, with intimal hyperplasia causing narrowing and