OASIS DERMATOLOGY GROUP

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Scleroderma

Scleroderma, also known as systemic sclerosis (SSc), is a chronic autoimmune disorder characterized by fibrosis and thickening of the skin and internal organs due to excessive collagen production. The disease can manifest in a limited form, predominantly affecting the skin, or as a systemic disorder with involvement of multiple organ systems. Scleroderma is classified into two primary subtypes: limited cutaneous systemic sclerosis (lcSSc) and diffuse cutaneous systemic sclerosis (dcSSc), with disease progression and organ involvement differing significantly between these forms.

Clinical Features and Classification

- ***Systemic Sclerosis***:
  Systemic sclerosis is characterized by progressive fibrosis of the skin and internal organs. The disease may initially present with Raynaud's phenomenon, a vasospastic disorder causing intermittent episodes of discoloration of the fingers and toes in response to cold or stress. This abnormal circulation results in pallor, cyanosis, and, occasionally, hyperemia as blood flow is restored. The disease can affect various organ systems, leading to fibrotic and vascular complications in the musculoskeletal, renal, pulmonary, cardiac, and gastrointestinal systems.
  - ***Skin Involvement***:
    Skin involvement is universal in SSc, with varying degrees of severity and distribution. Initial signs often include edematous swelling and erythema, which can later progress to scleroderma (thickening and hardening of the skin). The hands, fingers, and face are typically the earliest areas affected. Sclerodactyly, or the thickening of the skin on the fingers, can result in limited mobility and deformities. Other features include digital ulcers, telangiectasia, and calcinosis cutis (deposits of calcium in the skin).
- ***Classification by Extent of Skin Involvement***:
  - ***Limited Cutaneous SSc (lcSSc)***:
    This subtype typically involves the skin of the hands, face, distal forearms, and neck. Severe Raynaud's phenomenon is common, and telangiectasia (visible small blood vessels) may develop. Patients with limited SSc can also exhibit CREST syndrome, an acronym representing the features of the disease: Calcinosis cutis, Raynaud's phenomenon, Esophageal dysmotility, Sclerodactyly, and Telangiectasia.
  - ***Diffuse Cutaneous SSc (dcSSc)***:
    In contrast to the limited form, diffuse SSc involves a broader distribution of

sclerotic skin, often affecting the chest, abdomen, and upper arms. This form is associated with a higher likelihood of significant internal organ damage, including fibrosis and ischemic injury affecting the lungs, heart, kidneys, and gastrointestinal tract. Internal organ involvement is typically more severe and can lead to life-threatening complications.

Diagnosis

The diagnosis of systemic sclerosis is primarily clinical but can be supported by specific autoantibodies, including anticentromere antibodies (often present in limited cutaneous SSc) and anti-topoisomerase I antibodies (more common in diffuse cutaneous SSc). Capillaroscopy of the nailfold capillaries and pulmonary function tests may help assess vascular and respiratory involvement. Additionally, skin involvement can be quantified using the Rodnan skin score, which rates the extent of skin thickening and induration on a scale of 0 (normal) to 3 (most severe) across 17 different body areas.

Treatment

Treatment strategies for systemic sclerosis are multifaceted and tailored to the individual's symptoms, disease subtype, and organ involvement. While there is no cure, various therapies aim to manage symptoms, slow disease progression, and prevent complications.

- **Skin and Symptom Management**:
  - For pruritus (itching), topical agents such as camphor and menthol have been used, alongside PUVA (psoralen + ultraviolet A light) therapy, which has shown efficacy in alleviating skin thickening and improving flexibility.
  - Calcinosis cutis can be managed through surgical excision or, in some cases, with pharmacologic interventions, though outcomes vary.
- **Vascular and Raynaud's Phenomenon**:
  - Calcium channel blockers (e.g., nifedipine) are commonly prescribed to manage Raynaud's phenomenon, reducing the frequency and severity of vasospastic attacks. In severe cases, intravenous prostaglandins (e.g., epoprostenol) may be used for their vasodilatory effects.
  - Smoking cessation is critical, as tobacco exacerbates vascular dysfunction and contributes to poor outcomes in systemic sclerosis.
- **Gastrointestinal Involvement**:
  - For esophageal dysmotility and related reflux symptoms, proton pump inhibitors (PPIs) and H2 blockers are effective in managing acid reflux and improving swallowing function.
- **Pulmonary Involvement**:
  - Interstitial lung disease (ILD) is common in SSc, and treatment often includes cyclophosphamide or mycophenolate mofetil to manage inflammation and prevent

fibrosis. Calcium channel blockers and prostacyclin analogs (e.g., iloprost) may be used in cases with pulmonary hypertension.

- **Renal Involvement**:
  - Renal crisis, characterized by acute hypertension and renal failure, is a serious complication of diffuse cutaneous SSc. Angiotensin-converting enzyme (ACE) inhibitors, such as enalapril, are the treatment of choice, helping to control blood pressure and protect kidney function.
- **Diffuse Cutaneous SSc Management**:
  - Mycophenolate mofetil has shown promise in treating progressive, diffuse SSc by reducing fibrosis. Methotrexate and azathioprine are also frequently used, although their efficacy varies among patients.

Conclusion

Scleroderma, or systemic sclerosis, is a complex autoimmune disorder that presents with both skin and internal organ involvement. The two major subtypes, limited and diffuse cutaneous systemic sclerosis, differ in their severity, organ involvement, and prognosis. While no cure exists, early intervention and symptom-specific treatments, including immunosuppressants, vasodilators, and supportive therapies, can improve quality of life and prevent severe complications. In many cases, a multispecialist team is necessary to appropriately manage this condition. Continued research is needed to optimize therapeutic approaches and improve outcomes for individuals with scleroderma.

References

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