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Rheumatology: Current Research

Commentary

The Connection between Systemic Sclerosis and Raynaud's Phenomenon

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ABOUT THE STUDY

Systemic Sclerosis (SSc) is a chronic autoimmune disease that affects connective tissue in the skin and internal organs. Raynaud's Phenomenon (RP) is a clinical manifestation characterized by episodic color changes in fingers, toes, or both, in response to cold or emotional stress. SSc and RP are closely related and often coexist in patients, with RP being an early sign of SSc in many cases.

Pathophysiology

The pathophysiology of SSc is complex and multifactorial, involving immune dysregulation, vascular damage, and fibrosis. The immune dysregulation leads to the activation of T and B cells, cytokine production, and autoantibody production, which contribute to vascular damage and fibrosis. Vascular damage is a key feature of SSc, resulting from endothelial cell injury, intimal proliferation, and perivascular inflammation. Vascular damage leads to reduced blood flow and tissue hypoxia, which in turn triggers fibroblast activation and collagen deposition, leading to fibrosis.

RP is also characterized by vascular dysfunction, but its pathophysiology is more focused on the peripheral vasculature. RP is caused by an exaggerated vasoconstrictor response to cold or emotional stress, leading to narrowing of small arteries and arterioles in the fingers and toes. This vasoconstriction is mediated by the sympathetic nervous system and involves activation of alpha-adrenergic receptors on smooth muscle cells in the vessel wall. The vasoconstriction leads to reduced blood flow, tissue hypoxia, and tissue damage, which can lead to digital ulcers, gangrene, and even amputation in severe cases.

Clinical features

SSc is a heterogeneous disease that can affect multiple organs, including the skin, lungs, heart, kidneys, and gastrointestinal tract. The clinical features of SSc vary depending on the subtype of the disease and the organs involved. The two main subtypes of SSc are limited cutaneous SSc (lcSSc) and diffuse cutaneous SSc (dcSSc). lcSSc is characterized by skin involvement limited to the fingers, face, and neck, while dcSSc is characterized by skin involvement extending proximally to involve the trunk and limbs. RP is a common feature of both subtypes, affecting up to 90% of patients with SSc.

RP is characterized by episodic color changes in the fingers or toes, usually in response to cold or emotional stress. The color changes typically involve a triphasic pattern of pallor, cyanosis, and rubor, lasting for minutes to hours. RP can be painful and disabling, especially in severe cases, where it can lead to digital ulcers and tissue necrosis.

Diagnosis

The diagnosis of SSc is based on clinical and laboratory criteria, including skin changes, Raynaud's phenomenon, nailfold capillaroscopy, autoantibody testing, and organ involvement. The American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) have proposed classification criteria for SSc, which require the presence of RP and one of the following:

- Skin involvement proximal to the MCP joints
- Sclerodactyly
- Digital pitting scars or loss of tissue from the fingertips
- Bilateral interstitial lung disease on chest radiography or high-resolution computed tomography
- Pulmonary arterial hypertension on echocardiography or right heart catheterization

Nailfold capillaroscopy is a non-invasive technique that allows visualization of the microcirculation in the nailfold, which can provide valuable information for the diagnosis and prognosis of SSc. Nailfold capillaroscopy can reveal abnormalities in the density, morphology, and distribution of capillaries, such as giant capillaries, hemorrhages, and loss of capillaries, which are characteristic of SSc. The presence of early nailfold capillaroscopy abnormalities is a strong predictor of future development of SSc in patients with RP.

Autoantibody testing is also an important diagnostic tool in SSc, as up to 95% of patients with SSc have detectable autoantibodies. The most common autoantibodies in SSc are

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Anti-Centromere Antibodies (ACA), Anti-Topoisomerase Antibodies (ATA), and anti-RNA polymerase III antibodies (anti-RNA pol III). The presence of specific autoantibodies can also predict the clinical phenotype and prognosis of SSc. For example, patients with ACA tend to have lcSSc with a good prognosis, while patients with ATA or anti-RNA pol III tend to have dcSSc with a worse prognosis. The diagnosis of RP is based on clinical criteria, including the characteristic triphasic color changes in response to cold or emotional stress. However, other causes of RP should be excluded, such as secondary RP due to other autoimmune diseases, vasculitis, or occupational exposure to vibration or cold. Laboratory tests, including autoantibody testing, may also be helpful in distinguishing primary RP from secondary RP.

Treatment

There is currently no cure for SSc, and treatment is mainly focused on symptom management and prevention of complications. The treatment of RP is also aimed at symptom relief and prevention of digital ulcers and tissue damage. The management of SSc and RP is multidisciplinary and requires a coordinated approach between rheumatologists, dermatologists, pulmonologists, cardiologists, and other specialists, depending on the organs involved.

Treatment options for RP include lifestyle modifications, such as avoiding cold exposure and emotional stress, and pharmacological interventions, such as vasodilators and calcium channel blockers.

Nifedipine is a commonly used calcium channel blocker in the treatment of RP, which can reduce the frequency and severity of attacks. Other vasodilators, such as sildenafil, iloprost, and bosentan, may also be effective in reducing RP symptoms and preventing digital ulcers.

The treatment of SSc depends on the subtype of the disease and the organs involved. In lcSSc, skin involvement is usually limited, and treatment may focus on symptom relief and prevention of complications, such as digital ulcers, pulmonary hypertension, and renal crisis. In dcSSc, skin involvement is more extensive, and treatment may require immunosuppressive therapy to prevent or reduce organ damage.

Immunosuppressive therapy in SSc includes corticosteroids, methotrexate, mycophenolate mofetil, cyclophosphamide, and rituximab, depending on the severity and activity of the disease.

Biologic agents, such as tocilizumab and abatacept, have also shown promising results in the treatment of SSc, especially in patients with lung involvement.