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Raynaud's Phenomenon Involving all Four Limbs Post - Cold Exposure: A Rare Case Report

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Abstract: ***Raynaud's phenomenon (RP) is described in the scientific literature as a clinical syndrome considered common, but poorly recognized, causing changes in the color of the fingers due to vasospasm. Such changes occur due to exposure to a cold environment, emotional stress or other physical or drug exposures. Primary RP usually don't need pharmacological treatment, but investigation to secondary causes must be performed.***

Keywords: Autonomic nervous system, Raynaud disease, Therapeutics, Vasospasm

1. Introduction

Primary Raynaud's phenomenon (RP) is a common sign characterized by episodic color changes of acral parts of the body (pallor, cyanosis, rubor) lasting from a few minutes to hours, which are usually triggered by cold temperature and/or stress. Secondary RP is a symptom of an underlying disease. Raynaud's phenomenon has to be distinguished from other color changes of the distal extremities like acrocyanosis, erythromelalgia, perniosis, and Chilblain - Lupus.

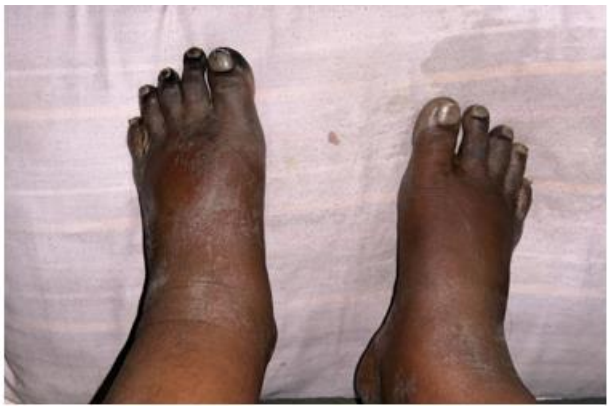
Autoimmune rheumatic diseases like systemic sclerosis and systemic lupus erythematosus, as well as vascular diseases like arterial occlusions and compression syndromes, are also possibilities that should be taken into account in the differential diagnosis. The rheumatologist or general practitioner, therefore, should be aware of clinical findings that may differentiate to benign causes or syndromes/diseases that require specific treatment.

On the basis of physical examination, as well as the use of complementary exams, such as nail capillaroscopy and immunological tests, it is possible to differentiate primary from secondary RP situations. Treatment is aimed at preventing RP, thus avoiding the possibility of irreversible ischemic damage. We present a case of Raynaud's phenomenon associated with exposure to cold, but also a discussion of its pathophysiology and differential diagnosis.

2. Case Report

A 43 - year - old female patient reported to civil hospital, Rajkot with blackening of the fingers and toes. She had history of putting limbs in ice cold water for long hours. A progressive worsening of the condition was observed, initially, the clinical picture was located only in the little finger and, later, the second, third and fourth metacarpals of both the hands and toes of both the foots were also affected. No family history had been referred.

At the time of onset of symptoms, laboratory tests were performed to rule out secondary causes. Inflammatory and immune - mediated diseases such as systemic lupus erythematosus and other rheumatologic diseases had been excluded. She was submitted to electrocardiogram and echodopplercardiogram. All these exams showed no significant changes.



Patient was started aspirin, pentoxifylline and cilostazol tablets for months.

With strict avoidance to the environmental factors. And was on regular follow ups for 3months.

3. Discussion

Raynaud's phenomenon (RP), as described in 1862 by Maurice Raynaud, is characterized by reversible episodes of extremity vasospasms, associated with changes in typical coloring that occur after exposure to cold or in stress situations.

Color changes are classically described in three successive phases: paleness (ischemic phase), cyanosis (caused by venostasis and deoxygenation), and flushing (hyperemia reactive/reperfusion). Pain and/or paresthesia may also be associated with attacks, causing discomfort to the individual.

Mainly in RP secondary to systemic sclerosis (SS), the vasospastic events are usually more intense and frequent, and often associated with ischemic ulcers. Additionally, progressive resorption of the extremities is the manifestation. In recent years, advances in the study of the pathophysiology of the RP and vascular disease in SS, for example, led to the onset of new therapeutic options for this manifestation.

The average age of onset of primary RP is 14 years of age and only 27% of cases start with around 40 years old or more. In contrast, the secondary RP tends to start in adulthood. The frequency and severity of episodes are influenced by daily temperature variations, with clear exacerbation during the winter and cold water.

The vascular tone Is controlled by the interaction between endothelial cells, vascular wall smooth muscle, soluble mediators, and neuronal stimulation. An imbalance between vasoconstriction and vasodilation, favoring vasoconstriction, is a central event in the pathophysiology of RP; although part of this mechanism still has question gaps. Patients who present primary Raynaud's phenomena should only be instructed about the triggers for the beginning of the clinical process.

4. Conclusion

In patients with primary RP, pharmacological treatment is generally not necessary, and non - drug measures such as patient education and protection from cold are sufficient. But drugs like aspirin, trental, cilostazol may slow down the progression of disease and may decrease the complications. On the other hand, RP secondary to autoimmune rheumatic diseases will often require drug treatment. In these cases, the severity and associated complications must be evaluated and the treatment must be stratified for each case. We emphasize the need for differential diagnosis.

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