Raynaud Phenomenon - Patient Page

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Abstract

Raynaud phenomenon is a condition in which some of the body's blood vessels (most commonly those in the fingers and toes) constrict (narrow down) excessively in response to cold or any emotional stress. Those patients who have Raynaud phenomenon without any related disease are said to have primary Raynaud phenomenon. Those patients who have a related underlying illness like scleroderma or systemic lupus erythematosus (SLE) are said to have secondary Raynaud phenomenon, also known as Raynaud syndrome. Most often, in a typical attack, suddenly the fingers become cold and the skin color changes markedly. Raynaud phenomenon is diagnosed clinically if the patient provides a history of the sudden onset of symptoms characteristic of a Raynaud attack. Patients with primary or secondary Raynaud phenomenon usually benefit by avoiding cold exposure, smoking, stress and anxiety. If general measures are not sufficient, medications like calcium channel blockers (include amlodipine, nifedipine, diltiazem) may be prescribed which can reduce both the frequency and severity of attacks.

Introduction

The circulatory system in our body involves the heart that pumps blood in our body, and specialized tubes (known as blood vessels), through which the blood flows. There are three major types of blood vessels: arteries, veins and capillaries. From the left side of the heart, the arteries carry blood rich in oxygen and nutrients to all organs of the body. The arteries connect to the capillaries which are very narrow channels that enable transfer of the oxygen and nutrients carried by blood in arteries to the tissues. The capillaries further connect to the veins that return the deoxygenated (i.e. devoid of oxygen), blood from these organs back to right side of heart. This cycle repeats itself thousands of times each day. This article focuses on a disorder of the arteries known as Raynaud phenomenon.

WHAT IS RAYNAUD PHENOMENON

Normally, blood vessels supplying the skin constrict in response to cold temperatures. This process, called "vasoconstriction," is necessary to decrease blood flow to the skin, which in turn helps to minimize heat loss from the body and preserve a normal range of internal temperature of the body. In warm temperatures the same blood vessels open up or dilate, allowing excess of heat to leave the body, thus again preserving internal body temperature. Raynaud phenomenon is a condition in which some of the body's blood vessels (most commonly those in the fingers and toes) constrict excessively in response to cold or any emotional stress. This is due to defects in mechanisms that control the normal constriction and dilation of blood vessels. This exaggerated/severe vasoconstriction restricts blood flow to the affected areas and causes the skin to change color to purple or blue, when the oxygen supplied is low, or white, when the blood flow in the vessel ceases. Eventually the vessel dilates again, thus allowing blood flow to resume and the skin color to turn normal. Sometimes the skin may even become very pink or red and throb due to excessive blood flow.

WHAT ARE THE TYPES AND CAUSES OF RAYNAUD PHENOMENON

Primary disease - Those patients who have Raynaud phenomenon without any related disease are said to have primary Raynaud phenomenon or Raynaud's disease. More than half of the patient's with Raynaud's phenomenon have primary disease. It generally presents at a younger age, mainly between 20-40 years and is more common in women (nearly five times more common than in men). Often there is a family history of Raynaud phenomenon. The exact underlying reason for this phenomenon is not known. The exaggerated response of blood vessels to cold may be genetically determined. Patients with Raynaud's disease generally have milder forms of Raynaud's phenomenon.

Secondary disease – Those patients who have a related underlying illness like scleroderma or SLE are said to have secondary Raynaud phenomenon, also known as Raynaud syndrome. Underlying illnesses include

- Connective tissue diseases like scleroderma (occurs in 80-90% of patients and can be the first symptom in 30%), systemic lupus erythematosus (SLE), dermatomyositis, polymyositis and rheumatoid arthritis.
- Occlusive vascular diseases like atherosclerosis, Buerger’s disease (or, thromboangiitis obliterans) and Idiopathic (primary) pulmonary hypertension.
- Blood disorders like cold agglutinin disease, cryoglobulinemia, waldenstrom's macroglobulinemia, etc.
- Side effect of drugs like ergot preparations, triptans, methysergide, beta-adrenergic receptor antagonists (beta-blockers), and the cancer chemotherapy agents like bleomycin, vinblastine, and cisplatin.
- Patients whose work includes the use of vibrating hand tools such as jackhammers and power drills. Pianists and keyboard operators also have higher incidence of this disease.
- Injury to hands like electric shock and frostbite.

Secondary Raynaud phenomenon can be more difficult to manage because it is linked to an underlying condition that can physically damage and occlude the blood vessels. Due to lack of oxygen and nutrient supply to the tissue, fingertip ulcers may develop and can also progress to gangrene and loss of parts of fingers and toes.

WHAT CAN PRECIPITATE AN ATTACK
Patients often report that exposure to cold temperature precipitate Raynaud phenomenon. More commonly, the phenomenon occurs during shift from warmer to cooler temperatures and even shifting to air-conditioned rooms may induce an attack. An attack may also occur after stimulation of the sympathetic nervous system (such as emotional stress). It is not uncommon for the patients to report an attack when they are nervous or anxious.

WHAT ARE THE SYMPTOMS OF RAYNAUD PHENOMENON
Most often, in a typical attack, suddenly the fingers become cold and the skin color changes markedly, and may become pale in what is called a "white attack" or purple or blue in a "blue attack". An attack of Raynaud phenomenon may begin in a single finger and then spread to other fingers in both hands. Patients may experience
- pain
- discomfort
- feeling of pins and needles
- numbness, or
- clumsiness of the affected hand.

Blood vessels supplying the skin of the ears, nose and face can also be affected. Occasionally patients with secondary Raynaud phenomenon may experience pain or ulceration at the tips of their fingers and/or toes after prolonged or repeated loss of blood flow to the tissues.

Mottling (a bluish discoloration) of the skin of the arms and legs might also appear. Attacks affecting the toes are also common, although patients tend to complain of these less frequently. Symptoms typically resolve when the precipitating factor is removed and as normal blood flow returns the skin becomes pink or red.

HOW IS RAYNAUD PHENOMENON DIAGNOSED
Raynaud phenomenon is diagnosed clinically if the patient provides a history of the sudden onset of symptoms characteristic of a Raynaud attack. No simple maneuver consistently triggers an attack so attempts to induce an attack are not recommended. The complaint of cold hands or feet is very common in many people and must be distinguished from Raynaud phenomenon, which leads to both cool skin and color changes. Normal individuals can have cold fingers and toes but they do not develop clearly visible color changes in skin.

Some investigations like nailfold capillaroscopy, thermal imaging (thermography), and laser Doppler imaging can often distinguish between healthy people and those with Raynaud's disease (primary Raynaud phenomenon), from those with secondary Raynaud phenomenon.

HOW CAN RAYNAUD PHENOMENON BE PREVENTED
Patients with primary or secondary Raynaud phenomenon usually benefit from the following measures:
- Avoiding cold exposure by wearing gloves and mittens during cold weather as well as in other situations where one can be exposed to cold temperature like while getting things out of a freezer or handling ice. In addition to gloves and mittens, patients should protect the trunk, head, and feet with warm clothing to prevent constriction of blood vessels in fingers and toes.
- Avoiding smoking which is strongly associated with atherosclerosis and thromboangitis obliterans
- Reducing stress and anxiety
- Avoiding medications that cause vasoconstriction as listed before.

HOW CAN RAYNAUD PHENOMENON BE TREATED
Primary Raynaud phenomenon — General measures listed above are tried alone initially and most of the patients benefit from them. If general measures are not sufficient, medications like calcium channel blockers (e.g. amlopidine, nifedipine, diltiazem) may be prescribed which can reduce both the frequency and severity of attacks.

Secondary Raynaud phenomenon — Patients with secondary Raynaud phenomenon may require more aggressive treatment. Medications are usually needed to reduce the frequency and severity of attacks and calcium channel blockers are used most frequently. Properly managing underlying illnesses is also necessary reduce the frequency and severity of Secondary Raynaud Phenomenon.

Other drugs can also be used in patients not responding to or having side effects from the calcium
channel blockers. These include nitrates (nitroglycerin), phosphodiesterase inhibitors (cilostazol, pentoxifylline, sildenafil), angiotensin receptor inhibitors (losartan), endothelin-1 inhibitors (bosentan), or prostaglandins (prostacyclin). Occasionally, for severe attacks of Raynaud’s phenomenon, or for intractable pain, ulceration, or gangrene related to compromised blood flow to the fingers (severe digital ischemia), hospitalization may be required in patients not responding to the above medications alone. In some cases, surgery (sympathectomy) may be used to block the nerves that trigger constriction of blood vessels in the fingers. If all other treatments have failed, surgical amputation of the affected finger or toe may be the only available option for advanced and irreversible damage where gangrene is inevitable.

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