By Vincent M. Vacca, Jr., MSN, RN, CCRN, SCRN

MS. J, 30, IS A HAIR STYLIST who owns a small salon. Recently, she's been experiencing episodic attacks of cold fingers of both hands associated with sharply demarcated skin pallor (white attack). It usually begins in one of her index fingers and spreads to her middle and ring fingers symmetrically in both hands. The pallor, which lasts for several minutes, is followed by cyanosis (blue attack), then by erythema with “a painful burning sensation.” Ms. J's attacks occur suddenly following exposure to cold temperatures, and her hands don't return to normal for up to several hours. During these attacks, she can’t use her hands skillfully or precisely, which inconveniences her customers and hurts her business.

In an attempt to rewarm her hands so she can resume working, Ms. J rubs them together, holds them under warm running water, and places them under her axillae. Ms. J is very concerned that these attacks could be caused by a serious disease that may permanently damage her hands, leaving her unable to work. Because of her concerns, Ms. J hasn’t been sleeping well and has increased her cigarette smoking from ½ pack to 1 pack per day.
One day a nurse who’s a customer is present during an attack. Suspecting that Ms. J may be experiencing Raynaud phenomenon (RP), she recommends immediate supportive treatment, including rewarming her hands, stopping smoking, and avoiding exposure to second-hand smoke, and suggests that Ms. J make an appointment with her primary care provider (PCP) for definitive diagnosis and treatment.

Realizing that RP attacks are episodic and may not be present during a clinic appointment, the nurse recommends that Ms. J have photos taken of her hands during an attack to show her PCP. Ms. J gives her cellphone to the nurse, who takes photos during this current attack.

A world of colors

RP causes repeated episodes of biphasic (sometimes uniphasic) color changes, usually in the hands upon exposure to stimuli such as cold environmental temperature or emotional stress. (See Who’s affected?) Although typically primary (idiopathic), meaning no underlying cause can be identified, RP can also be secondary to repetitive injuries to susceptible hands or an underlying disease such as a connective tissue disease (CTD).

Who’s affected?
The onset of RP is most common during the teenage years, with the median age of onset being 14. About 27% of people presenting with the condition for the first time are age 40 or older. RP seems to have a familial or genetic predisposition. The Raynaud’s Association has reported that the incidence of RP in adults is 4% to 20% worldwide. About 5% to 10% of all Americans suffer from RP, with about nine times as many women affected as men. Some researchers estimate as many as 20% of all women in their childbearing years have RP.

Pathophysiology points

Impaired vasodilation, sustained vasoconstriction, endothelial dysfunction, and platelet activation and aggregation are all implicated in the signs and symptoms of RP. Vasoconstriction in RP is an exaggerated physiologic response of the small muscular arteries and arterioles in the digits or other affected area to a trigger, such as cold or stress. Arterial vasoconstriction is associated with an increase in alpha-2 adrenergic responses in the digital vessels leading to arterial vasospasm. The vascular endothelium normally releases vasodilating substances such as nitric oxide, prostaglandins, and neuropeptides, but capillary damage associated with RP can impair release of these substances, leading to sustained and unopposed vasoconstriction.

In the digits, temperature and color changes are usually symmetrical and reversible, often with a clear line of demarcation; the thumbs are usually unaffected. Although signs and symptoms usually resolve within an hour, they may last for several hours.

If the thumbs are affected, this can be a sign of an underlying CTD. Unilateral signs and symptoms such as pallor suggest an obstructive arterial lesion, thoracic outlet syndrome, or injury.

Primary Raynaud phenomenon

Primary Raynaud phenomenon (PRP) is related to functional alterations of the vascular endothelium that cause episodic vasospasm of peripheral arteries. PRP is considered benign because it doesn’t progress to irreversible tissue injury and damage that can lead to digital ulceration and gangrene.

When no underlying illness has been identified, the condition is known as PRP. However, patients with PRP may go on to develop secondary Raynaud phenomenon (SRP).

Understanding SRP

SRP occurs in certain autoimmune rheumatic diseases, for example, it’s...
seen in scleroderma (90% to 95% of these patients have SRP), mixed CTD (85%), systemic lupus erythematosus (40%), antisyntetase syndrome (40%), and sometimes in patients with other autoimmune rheumatic diseases. It may also be seen in hematologic disorders such as cryoglobulinemia, cryofibrinogenemia, paraproteinemia, cold agglutinin disease, and polycythemia vera. SRP can also result from environmental and occupational exposures (such as frostbite or use of vibrating tools) and from exposure to certain drugs and toxins (such as polyvinyl chloride). Recurrent episodes of peripheral arterial ischemia and vasodilatory reperfusion can lead to significant morbidity and multiple complications, including digital ulceration, necrosis, tissue loss, gangrene, and even amputation. SRP may even become life-threatening.

**Diagnostic process**

Ms. J presents to her PCP for evaluation and treatment of her suspected RP. Because she isn’t having an episode during her visit, the photographs taken by the nurse during the event are especially valuable to her PCP in addition to her history.

RP is diagnosed based on patient history, including characteristic demarcated skin pallor and/or cyanosis of digit(s) upon cold exposure, followed by reperfusion erythema. Diagnostic studies to help rule out SRP may include a complete blood cell count, erythrocyte sedimentation rate, antinuclear antibodies, and rheumatoid factor if CTD or autoimmune disease is suspected. Nailfold video capillaroscopy, also known as nailfold capillary microscopy (NCM), helps detect abnormal capillaries associated with CTD, such as systemic sclerosis.

Results of Ms. J’s blood work and NCM don’t suggest underlying CTD or other underlying disorder, so Ms. J will be treated for PRP.

**Management of RP**

General measures to avoid RP triggers, such as avoiding emotional stress and cold exposure, maintaining a normal core body temperature, wearing gloves in cold weather, stopping vasoconstrictive medications, and smoking cessation, may be sufficient in patients with PRP. Although these measures are also important with SRP, these patients are more likely to require pharmacologic or even surgical treatment.

Patients with SRP should also follow general recommendations to prevent attacks, but they’ll most likely need pharmacologic therapy as well. Medications to inhibit vasoconstriction, promote vasodilation, support endothelial function, and reduce platelet aggregation may be included in the treatment plan. Calcium channel blockers are most commonly used for patients with RP who don’t respond to nonpharmacologic management. Additional pharmacologic therapies may include phosphodiesterase inhibitors, such as sildenafil, and topical nitrates. Additional management strategies are usually indicated for patients with ischemic ulcers or other thrombotic events. For some patients who fail conservative and pharmacologic management strategies, and have critical ischemia, surgical intervention such as a thoracoscopic sympathectomy may be beneficial.

**Warming up to treatment**

Ms. J’s treatment plan includes conservative measures such as using hand-warming techniques, avoiding cold temperatures, limiting exposure to vibrations, and avoiding sympathomimetic medications and substances, including tobacco.

Following diagnosis and patient education, Ms. J can manage her PRP and return to her salon to provide skilled styling services for her customers and successfully manage her business.

**REFERENCES**


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