

VASOSPASTIC DISORDERS

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It is extremely important for the podiatric surgeon to be familiar with circulation and its inherent abnormalities when evaluating a patient with a known or suspected vasospastic disorder. Not only is it essential that the surgeon know whether the circulation will sustain adequate and prompt healing, but it is also imperative that he or she be familiar with the etiologies, pathophysiology and methods of treatment should therapeutic intervention become necessary.

Vasospastic disorders of the foot consist of functional arterial problems caused by excessive constriction or dilation of reflexive vessels. These circulatory problems may be the result of an abnormality in the vessel walls, sympathetic nervous system, or a product of a pathological vascular or systemic disease.

The following classical vasospastic conditions must be considered in the perioperative management of the surgical patient. Raynaud's phenomenon is seen with intermittent episodes of blanching or cyanosis of the digits of the hands and/or feet, brought on by exposure to cold temperature or emotional stress. This initial blanching is followed by a period of a bright red hyperemic phase upon rewarming. Clinically, there is a progression from extreme pallor to a bluish phase of cyanosis to a bright red flush upon rewarming. Attacks last for minutes or hours.

When pain is present in the ischemic phase, a secondary cause should be suspected. This phenomenon may be idiopathic (Raynaud's disease) or secondary to connective tissue disorders (eg. SLE, RA, scleroderma), obstructive arterial diseases (arteriosclerosis obliterans, thromboangiitis obliterans, thoracic outlet syndrome), or neurogenic conditions which result in sympathetic nervous system disturbances. Drug intoxications (ergot and methysergide), beta adrenergic receptor blockers, bleomycin, Vinblastin and Cisplatin, myxedema, primary pulmonary hypertension and

trauma of occupational origin are other associated inducers of this vasospastic condition.

Patients with Raynaud's phenomenon frequently also have a history of migraine headaches or variant angina. This association suggests that there may be a common predisposing cause for the vasospasm.

The majority of patients with Raynaud's disease are young women, five times more common than men. The usual age of presentation is between 20 and 40 years. The fingers are frequently involved more than the toes, although the toes are affected in 40 percent of the patients. The earlobes and tip of nose have been involved but very infrequently.

Raynaud's disease is differentiated from Raynaud's phenomenon by bilateral involvement, a history of symptoms for at least two years without progression and no evidence of an underlying cause. In Raynaud's disease, trophic skin changes and gangrene are either absent or present, and if present they are only in minimal areas. The physical exam of the radial, ulnar, and pedal pulses is entirely normal. In Raynaud's phenomenon associated with scleroderma, there may be tightness and thickening of the skin, telangiectases of the hands, arms, or face, difficulty swallowing, painful trophic ulcers of the digital tips, and symptoms referable to other systems.

The diagnosis is usually made from the history of typical episodic attack of a demarcated vasospasm of the digits upon cold exposure. Attacks are very difficult to induce in patients with an idiopathic disorder, despite immersion of the hand in cold ice water. However, patients with sufficient symptoms should have a complete work-up to exclude secondary causes.

A detailed history will elicit symptoms of collagen vascular disease (arthralgia, arthritis, dysphagia, heartburn, facial rash from the sun, persistent tans), a drug etiology, symptoms of

obstructive arterial disease, or vibrational arterial injury which may occur with use of industrial tools. Physical examination should note all pulses, blood pressure in both arms, subcutaneous nodules, swollen or deformed joints, skin texture, discoloration of eyelids, bruits in the neck, thyroid size, relaxation time of reflexes, neurologic deficits especially with sympathetic nervous system overactivity including multiple sclerosis. The Tinel's sign should be sought.

Blood analysis for anemia, polycythemia, leukopenia, sedimentation rate, serum protein electrophoresis, and antinuclear bodies are necessary. A urinalysis should be done, specifically looking for proteinuria and red blood cells casts. Recently a new technique employing a high frequency A mode ultrasound scanner has been successfully utilized in detecting digital arterial closure with a high degree of reproducibility. This technique may prove useful in monitoring patients with Raynaud's phenomenon.

The treatment of patients with episodic digital vasospasm is primarily based on avoidance of cold exposure, other known causes of reflex sympathetic vasoconstriction, and pressure on the digits. Tobacco smoking should be avoided since nicotine induces intense cutaneous vasoconstriction; smokers are a major percentage of patients with Raynaud's disease and with traumatic vasospastic disease. In few patients, relaxation techniques such as biofeedback may reduce vasospastic episodes. Therapy for Raynaud's phenomenon depends on recognition and treatment of the underlying disorder.

Drug therapy is successful in about 50 percent of the patients with the more severe phenomenon or disease. Both reserpine and guanethidine, sympatholytic drugs, have been shown to increase capillary blood flow but have a side effect of depression which may prevent its use. The drugs of choice are the newer vasodilating agents pazasin and calcium channel blockers, especially nifedipine. Encouraging reports on the use of pentoxifylline, a rheologic agent have appeared in the literature. There is also some evidence to suggest that serotonin is a contributory element in the maintenance of cold induced vasospasm. Research with encouraging results is in progress studying the effects of prostaglandins (thromboxane) in the treatment of Raynaud's phenomenon. Occasionally, regional surgical

sympathectomy is proposed for patients with progressive disability and appear to be refractory to all medical therapy. Often surgical sympathectomy alleviates symptoms, but relief may only last as long as two years. Results of this type of procedure are more successful in the patient with Raynaud's disease.

In addition, there is encouraging evidence that the use of chronic beta blockers, at low dosages, can prevent cold induced vasospasm. Better response to atenolol is seen than with propranolol. There is also recent data showing relief of vasospasm from a topical glyceryl trinitrate. Digital plethysmography in patients with secondary Raynaud's phenomenon and an underlying connective tissue disorder has demonstrated the effect of this topical drug when applied locally.

See Table 1 for classification of Raynaud's phenomenon.

ACROCYANOSIS

Acrocyanosis is arterial vasoconstriction and secondary vasodilation of capillaries and venules which results in persistent, painless symmetric cyanosis of the hands and less commonly the feet. The digits are cool and often excessively moist. Color abnormality is intensified by cold or emotional upset. Puffiness and numbness may accompany the cyanosis. This disorder usually occurs in women and is not associated with occlusive vascular disease.

Clinically the disorder can be distinguished from Raynaud's phenomenon by lack of episodic, well-demarcated color changes and blanching does not occur.

Acrocyanosis is considered to be a benign disorder. Treatment is usually unnecessary. Patients should be advised to dress warmly and avoid the cold. Vasodilators have been used with little success and surgical sympathectomy is not usually indicated.

ERYTHROMELALGIA (ERYTHERMALGIA)

This disorder occurs with sudden episodes of vasodilation causing increased erythema, skin temperature, and pain. It more commonly involves the feet as compared to the hands. The etiology of primary erythromelalgia is unknown. It may be secondary to myeloproliferative disorder.

TABLE 1
CLASSIFICATION OF RAYNAUD'S
PHENOMENON

- I. Primary or idiopathic Raynaud's phenomenon: Raynaud's disease
- II. Secondary Raynaud's diseases:
 - A. Collagen vascular diseases:
 - 1. Scleroderma.
 - 2. SLE
 - 3. RA
 - 4. Dermatomyositis.
 - 5. Polymyositis.
 - B. Arterial occlusive diseases.
 - 1. Atherosclerosis of the extremities.
 - 2. Thromboangitis obliterans.
 - 3. Acute arterial occlusion.
 - 4. Thoracic outlet syndrome.
 - C. Pulmonary hypertension
 - D. Neurologic disorders.
 - 1. Intervertebral disk disease.
 - 2. Syringomyelia.
 - 3. Spinal cord tumors.
 - 4. Stroke.
 - 5. Poliomyelitis.
 - 6. Carpel tunnel syndrome.
 - E. Blood dyscrasias.
 - 1. Cold agglutinins.
 - 2. Cryoglobulinemia.
 - 3. Myeloproliferative disorders.
 - 4. Waldenstrom's macroglobulinemia.
 - F. Trauma.
 - 1. Vibration injury.
 - 2. Hammer hand syndrome.
 - 3. Electric shock.
 - 4. Typing.
 - 5. Piano playing.
 - G. Drugs.
 - 1. Ergot derivatives.
 - 2. Methysergide.
 - 3. Beta adrenergic receptor blockers.
 - 4. Bleomycin.
 - 5. Vinblastin.
 - 6. Cisplastin.

ders, hypertension, venous insufficiency or diabetes mellitus. There is no sex predilection and the disease may occur at any age. Patients complain of attacks of intense burning pain with hot, red feet or hands, precipitated and aggravated by warm environment and a dependent position.

The onset of the disorder is gradual, but with progression, the frequency and duration of the attacks become more pronounced. Eventually, symptoms become almost continuous and cause total disability. It is usually bilateral and can extend to the level of the knee. Trophic changes are rare. Examination shows bounding pulses, redness and heat of the part. Relief is characteristically sought by exposing the part to cold or standing on a cold tile floor.

Peripheral neuropathy may cause a burning pain similar to erythromelalgia. The pain may be accompanied by cutaneous vasodilation. Detection of associated sensory and motor manifestations of peripheral neuropathy should help distinguish this condition from erythromelalgia.

Treatment involves avoidance of heat exposure. Attacks can be avoided by rest, elevation, and cold application. In primary erythromelalgia, doses of aspirin 600 - 650 mg one to four times a day can produce relief in most cases. This response may be of diagnostic value. Vasoconstrictive agents, such as ephedrine, propranolol and methysergide may also produce relief in some patients. In secondary manifestations of erythromelalgia, correction of the underlying disease may prevent further attacks.

SUMMARY

After a complete history and physical has been performed and a diagnosis of a vasospastic condition is determined, it is imperative that the proper perioperative measures be taken to avoid vascular crisis and catastrophe. The prudent podiatric surgeon must keep current with the ever-advancing research and technology available to assist in the prompt and accurate detection of vasospastic conditions and effective perioperative management and treatment.

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