



When cold-induced vasospasm is the tip of the iceberg

For practitioners who see a lot of patients, particularly a lot of young women, patients describing cold-induced color changes of the fingers sometimes accompanied by tingling or burning are common. For most patients it is mild, but for some the discoloration or dysesthesia may be striking and disconcerting. For a minority, this reversible vasoconstrictive phenomenon (Raynaud "disease" if it occurs in isolation, without any associated underlying condition) may be the presenting sign of a systemic disorder.

For many patients, Raynaud symptoms are mild enough to not even mention to their primary care provider, and conversely, there is little reason for most clinicians to routinely inquire about such symptoms. So it may surprise some readers to read about the nuances of diagnosis and treatment discussed by Shapiro and Wigley in this issue of the *Journal* (page 797).

To a rheumatologist, Raynaud phenomenon, particularly of recent onset in an adult, raises the specter of an underlying systemic inflammatory disease. The phenomenon is not linked to a specific diagnosis; it is associated with lupus, rheumatoid arthritis, cryoglobulinemia, inflammatory myopathy, Sjögren syndrome, and, in its severe form, with the scleroderma syndromes. We focus on differentiating between these rheumatic disorders once we have discarded nonrheumatic causes such as atherosclerotic arterial disease, carcinoma, embolism, Buerger disease, medications, smoking, or thrombosis.

But rheumatologists are toward the bottom of the diagnostic funnel—we see these patients when an underlying disease is already suspected. The real challenge is for the primary care providers who first recognize the digital vasospasm on examination or are told of the symptoms by their patient. These clinicians need to know which initial reflexive actions are warranted and which can wait, for, as noted by Shapiro and Wigley, there are several options.

The first action is to try to determine the timeline, although Raynaud disease often has an insidious onset or the patient doesn't recall the onset. New and sudden onset likely has a stronger association with an underlying disease. A focused physical examination should look for digital stigmata of ischemic damage; the presence of digital ulcers or healed digital pits indicates a possible vascular occlusive component in addition to the vascular spasm. This strongly suggests scleroderma or Buerger disease, as tissue damage doesn't occur in (primary) Raynaud disease or generally even with Raynaud phenomenon associated with lupus or other rheumatic disorders. Sclerodactyly should be looked for: diffuse finger puffiness, skin-tightening, or early signs such as loss of the usual finger skin creases. Telangiectasia (not vascular spiders or cherry angioma) should be searched for, particularly on the palms, face, and inner lips, as these vascular lesions are common in patients with limited scleroderma. Careful auscultation for

basilar lung crackles should be done. Distal pulses should all be assessed, and bruits in the neck, abdomen and inguinal areas should be carefully sought.

Patients should be questioned about any symptom-associated reduction in exercise tolerance and particularly about trouble swallowing, "heartburn," and symptoms of reflux. Although patients with Raynaud disease may have demonstrable esophageal dysmotility, the presence of significant, new, or worsened symptoms raises the concern of scleroderma. Patients should be asked about symptoms of malabsorption. Specific questioning should be directed at eliciting a history of joint stiffness and especially muscle weakness. The latter can be approached by inquiring about new or progressive difficulty in specific tasks such as walking up steps, brushing hair, and arising from low chairs or the toilet. Distinguishing muscle weakness from general fatigue is not always easy, but it is important.

Shapiro and Wigley discuss the extremely useful evaluation of nailfold capillaries, which can be done with a standard magnifier or ophthalmoscope. This is very valuable to help predict the development or current presence of a systemic rheumatic disease. But this is not a technique that most clinicians are familiar with. A potentially useful surrogate or adjunctive test, especially in the setting of new-onset Raynaud, is the antinuclear antibody (ANA) test; I prefer the immunofluorescent assay. While a positive test alone (with Raynaud) does not define the presence of any rheumatic disease, several older studies suggest that patients with a new onset of Raynaud phenomenon and a positive ANA test are more likely to develop a systemic autoimmune disorder than if the test is negative. Those who do so (and this is far from all) are most likely to have the disease manifest within a few years. Hence, if the ANA test is positive but the history, physical examination, and limited laboratory testing (complete blood cell count with differential, complete metabolic panel, creatine kinase, and urinalysis) are normal, it is reasonable to reexamine the patient in 3 months and then every 6 months for 2 to 3 years, repeating the focused history and physical examination. It is also reasonable at some point to refer these patients to a rheumatologist.

Since Raynaud phenomenon is common, and the associated severe rheumatic disorders associated with it are rare, it is easy to not recognize Raynaud phenomenon as a clue to the onset of a potentially severe systemic disease. Yet with a few simple questions, a focused examination, and minimal laboratory testing, patients who are more likely to harbor a systemic disease can usually be treated symptomatically if necessary, and appropriately triaged to observation or for subspecialty referral.



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