New Raynaud's: Nail Folds Predict Scleroderma

BY KATE JOHNSON
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SNOWMASS, COLO — The most significant predictor of progression to scleroderma in a patient with new onset Raynaud's phenomenon is the presence of capillary abnormalities at the proximal nail fold, according to David H. Collier, M.D.

Although scleroderma is primarily managed by rheumatologists, it is dermatologists who most commonly identify the early skin manifestations of the disorder, said Dr. Collier of the University of Colorado, Denver, and chief of rheumatology at Denver Health Medical Center.

In addition to Raynaud's, these manifestations include skin thickening, ulceration, telangiectases, calcinosis, and pigmentation changes.

Speaking at a clinical dermatology seminar sponsored by Medicis, Dr. Collier explained that Raynaud's phenomenon is an almost universal component of systemic sclerosis, and yet the vast majority of patients with Raynaud's never progress to scleroderma.

"Up to 10% of adult women can have Raynaud's, and less then 0.1% can go on to develop scleroderma," he said in an interview, adding that about 77% of Raynaud's patients are female.

By examining the periungual area of the finger, under gel with an opthalmoscope, the physician can easily assess capillary abnormalities at the proximal nail fold, he said.

"Instead of thin little loops of capillaries that you would see in a normal patient, you see capillary dilation and areas that are denuded or dropped out altogether," he said, explaining that capillary dilation occurs early in the disease, and after about 10 years, only denudation is typically visible.

"A patient with abnormal capilloscopy should be followed every 3-6 months for signs of progression to systemic sclerosis," he advised, adding that early identification of scleroderma and referral can allow for a prompt pulmonary evaluation and establishment of gastroesophageal reflux prevention/management.

Pitting or ulceration of the fingertips is another indication that a Raynaud's patient has scleroderma, said Dr. Collier.

"Primary Raynaud's disease does not give you pitting. So if you see pits—especially fingertip pits and ulceration—that's a red light [indicating] that you're dealing with an autoimmune disease. It's almost always Raynaud's secondary to scleroderma or mixed connective tissue disease, or occasionally lupus," he said.

In addition to the evaluation for capillary abnormalities, the scleroderma workup for patients presenting with Raynaud's should also include autoantibody testing, he said.

"If they also have the antibodies, that's the subgroup that I worry about the most for progressing to scleroderma, but it's not universal. I've certainly followed people with autoantibodies, and they didn't progress."

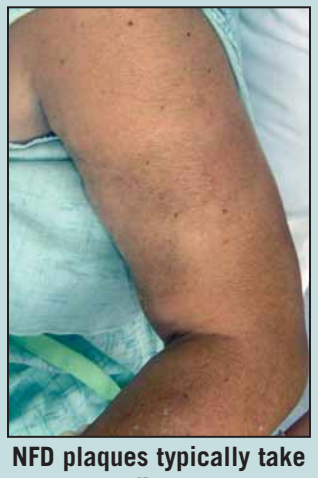
Anticentromere antibodies are seen in 20%-30% of scleroderma patients and are the most predictive of risk to progression to limited systemic sclerosis, although they are also commonly seen in primary biliary cirrhosis and, rarely, in other connective tissue diseases, such as rheumatoid arthritis, systemic lupus erythematosus, mixed connective tissue disease, and polymyositis, he said.

Anti–topoisomerase-1 antibodies (e.g., anti–scleroderma [Scl]-70) are present in 9%-20% of scleroderma patients. Anti–RNA polymerase 1-3 antibodies are seen in 20%. Antifibrillarin/anti-U3 ribonucleoprotein (anti-U3 RNP) antibodies are seen in 10%, and anti–neutrophilic cytoplasmic antibodies (ANCA) are seen in about 4%, though mostly in patients with diffuse systemic sclerosis. Finally, antipolymyositis /Scl (anti-PM Scl) and anti-Th/To (which recognizes certain RNA processing enzymes) antibodies are seen in about 2% of scleroderma patients. ■

A Rare Scleroderma Look-Alike: Nephrogenic Fibrosing Dermopathy

A recently described cutaneous fibrosing disorder could be mistaken for scleroderma, but there are some key differences, said Dr. Collier.

Worldwide, there have been only 170 cases of nephrogenic fibrosing dermopathy (NFD) reported since it was first described in 1997, he said. Yet "I think it's far more common than we're led to believe," he added.



COURTESY DR. DAVID H. COLLIER

NFD plaques typically take on a peau d'orange appearance.

The typical presentation of NFD consists of acute, lumpy, plaquelike indurations involving the lower limbs and occasionally the upper limbs and torso, he said.

Usually, scleroderma starts on the hands and face. But in NFD, these are almost always spared, he said.

The most common distribution of NFD skin presentation is between the ankles and the mid-thighs and between the wrists and mid-upper arms bilaterally, he said. Skin-colored to erythematous papules coalesce into brawny plaques with a peau d'orange appearance. There is a distinctive, irregular edge with amoeboid projections and islands of sparing within the indurated plaque. Eventually, the skin becomes markedly thickened and woody. Pruritis and causalgia are prominent features.

Unlike scleroderma, NFD often causes severe sharp pains in the affected areas, and renal insufficiency is necessary for the diagnosis.

The biopsy will show deposits of collagen and elastin—spindle cells, dendritic cells, and mucin deposits—"which is different from what we see in scleroderma."

Although NFD was initially thought to be only a cutaneous disease, there now appears to be a severe myopathic component. Joint contractures may develop within days or weeks of onset, likely resulting from facial and muscle fibrosis, Dr. Collier noted.

The abrupt emergence of this disease suggests that toxic exposures, infectious agents, or medical techniques may be involved.

—Kate Johnson