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Case Report

A 17-year-old Caribbean-American female presented with a 1-month history of violaceous papules, petechiae, and healing ulcerations on the distal digits of both hands (Figure 1) and feet. She reported that her fingers (Figure 2) had become cold and white intermittently during the winter months since she moved to the northeast from the Caribbean. She also described intermittent shortness of breath.

The patient did not have fever, chills, cough, photosensitivity, arthralgias, mucosal lesions, or any recent weight change. There was no history of new medications, frostbite, or repetitive stress injury and no family history of autoimmune disease. Dermatoscopic evaluation revealed dilated capillaries of the proximal nail folds.

Diagnosis

The history and physical examination findings are consistent with a diagnosis of Raynaud phenomenon (RP). Given the patient’s positive review of systems, healing digital ulcerations, and nail fold findings, laboratory testing was initiated.

Testing revealed positive antinuclear, Ro, and La antibodies and an elevated erythrocyte sedimentation rate (ESR). The complete blood cell count showed a white blood cell count of 3 K/uL (normal range, 4 to 10.5 K/uL), a hemoglobin level of 11 g/dL (normal range, 12 to 15 g/dL), and a hematocrit level of 34% (normal range, 35% to 45%). The patient’s mild leukopenia and anemia may be caused by her underlying condition, because both can be seen in patients with systemic lupus erythematosus (SLE). Complete metabolic panel, urinalysis, Smith, RNP, Scl-70, centromere, and histone antibodies findings were normal.

Based on the patient’s clinical presentation and the serological laboratory results, the most likely diagnosis is RP secondary to underlying autoimmune disease, such as SLE, systemic scleroderma, or mixed connective tissue disease.

The patient was advised to maintain her core body temperature, and extended-release nifedipine was prescribed. She was referred to a pediatric rheumatologist, who is concerned for underlying SLE, although the patient currently does not meet the clinical criteria.
Discussion

RP is an exaggerated vascular response to cold temperature or emotional stress. The vasoconstrictive response of the fingers and toes is characterized by the following 3 well-defined phases: (1) a vasospasm phase that causes pallor; (2) a cyanotic phase caused by deoxygenation of static venous blood; and (3) a hyperemic phase when blood flow is restored, resulting in erythema. All 3 phases are present in about 60% of patients.\(^1\)

RP is considered primary if there is no evidence of an underlying medical illness. Primary RP is more likely to have symmetrical involvement and affect younger females (median age, 14 years).\(^2\) Also suggesting primary RP are a lack of a history of potential secondary causes, including normal physical examination findings with the absence of digital ulceration, necrosis, or gangrene; normal nail fold capillaries; negative test results for antinuclear antibodies and other autoantibodies; and a normal ESR.\(^3\)

Secondary RP occurs in association with other conditions, such as SLE, scleroderma, and peripheral vascular disease. RP may be an early sign of systemic connective tissue disorders in about 13% of patients as demonstrated in a meta-analysis involving 639 patients.\(^4\)

Secondary RP is more prevalent in patients older than 30 years and in patients with severe prolonged attacks, asymmetrical involvement, and ischemic skin lesions. The presence of autoantibodies is also suggestive of secondary RP. However, antinuclear antibodies have a somewhat low predictive value of 30%. The presence of antibodies to a specific autoantigen is more indicative of underlying connective tissue disease.\(^4\)

The diagnosis can be made based on the clinical presentation and a history of symptoms consistent with RP. Diagnostic tools that may be useful include nail fold capillaroscopy, thermal imaging, and laser Doppler imaging.\(^5\) An abnormal nail fold capillary pattern has been shown to be the best predictor of possible connective tissue disease in patients with RP (positive predictive value, 47%).\(^4\)

Further autoimmune laboratory work-up to evaluate for secondary causes is warranted by the presence of sclerodactyly, digital necrosis, dilated capillary loops, and a positive review of systems. Treatment of patients with RP includes nonpharmacological therapy, such as maintaining core body temperature, and therapies that reduce emotional stress. Pharmacological treatments that have been shown to be beneficial include calcium channel blockers, angiotensin II-receptor inhibitors, selective serotonin reuptake inhibitors, and botulinum toxin type A injections. Calcium-channel blockers, such as nifedipine, are the first-line therapy. They have been shown to reduce attacks in patients with primary RP in a number of placebo-controlled trials.

Summary

In conclusion, this patient seemed to have characteristics that are seen more frequently in patients with primary RP, such as her young age and the symmetrical involvement of fingers. However, she had a positive review of systems, which warranted an autoimmune work-up. Further testing revealed the presence of autoantibodies. This case underlines the importance of proper assessment of a patient who presents with RP and investigation into possible secondary causes. Early diagnosis and management of any underlying disease may improve the symptoms of RP and prevent digital ulceration and necrosis.

References:

Links: