White and Blue Fingers: A Red Flag in Sickle Cell Disease

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J Gen Intern Med 28(3):477

DOI: 10.1007/s11606-012-2238-7

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Figure 1. Raynaud's phenomenon exhibiting acral cyanosis, pallor, and erythema.

A 34-year-old woman with hemoglobin SC disease had intermittently painful and swollen wrists, metacarpophalangeal (MCP) and proximal interphalangeal (PIP) joints. She photographed her right hand after going outdoors for 10 min (41 °F air temperature). The photograph demonstrated pallor, cyanosis, and erythema (Fig. 1),

Received February 20, 2012

Revised June 18, 2012

Accepted September 12, 2012

Published online October 5, 2012

which abated after 10 min and were unrelated to pain crisis. Based on history and exam, she was diagnosed with Raynaud's phenomenon. Evaluation revealed an ANA of 1:320 and negative Smith, RF, and CCP antibodies. Anti-RNP was elevated (25 EU/mL), suggesting possible mixed connective tissue disease and secondary Raynaud's phenomenon.

Hand pain and swelling in sickle cell disease can be from osteonecrosis, osteomyelitis, or hand-foot syndrome during vaso-occlusive crisis (up to 20 % incidence before age two, but rare in adults).^{1,2} However, Raynaud's phenomenon is not known to be associated with sickle cell disease, and warrants further investigation for other possible etiologies to avoid misdiagnosis and treatment delays.³ Secondary Raynaud's may result from vasculopathy, endocrinopathy, drugs, malignancy, or connective tissue diseases. Evaluation may include ESR, ANA, RF, and other disease-specific antibodies (Scleroderma, SLE, MCTD, etc.), as indicated by history. Close monitoring of patients with negative studies may be warranted, as 1 % per year may eventually develop a connective tissue disease.⁴

Acknowledgements:

Contributors: None

Funders: No internal or external funding sources

Prior presentations: None

Conflicts of Interest: The authors declare that they do not have a conflict of interest.

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