Images in Rheumatology

Raynaud’s Phenomenon of the Tongue

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An elderly patient with multiple autoimmune conditions was found to have intermittent unilateral and bilateral tongue cyanosis consistent with Raynaud’s phenomenon in the setting of severe systemic vasculitis. Her presentation, laboratory findings, and clinical course are reviewed.

A 71-year-old woman with a history of multiple autoimmune conditions including Sjögren’s syndrome, mixed connective tissue disease, and autoimmune hepatitis was admitted to hospital for complications during treatment for marginal cell lymphoma. She developed multiple deep venous thromboses, followed by a spinal hematoma after initiation of heparin. Additionally, she started having intermittent cyanosis of her toes, fluctuating tender swelling of her hands, and a superficial vascular rash on her upper arm. Magnetic resonance angiography of the affected regions showed no structural abnormalities and echocardiography ruled out endocarditis.

Laboratory evaluation revealed positivity for antinuclear antibody, SSA, SSB, rheumatoid factor, and cryoglobulins. She had low complement levels, slightly elevated prothrombin time, elevated D-dimer, and normal fibrinogen, erythrocyte sedimentation rate, and C-reactive protein. Further investigations showed absence of anticardiolipin, anti-β2-glycoprotein, and antipherin antibodies. Lupus anticoagulant panel was negative and protein S was normal.

Based on the clinical and laboratory findings, she was diagnosed with severe systemic vasculitis and started on high-dose steroids, cyclophosphamide, and calcium channel blockers. Within days her tongue exhibited features consistent with Raynaud’s phenomenon. Several times per day, and often associated with ipsilateral headache, her tongue showed both unilateral and bilateral cyanosis, likely attributable to lingual artery spasm. While experiencing no pain, she had significant dysarthria. Blue discoloration resolved temporarily with the application of warm water.

Lingual artery thrombosis was felt to be less likely given the fluctuating symptoms and occasional bilateral involvement. Acrocyanosis could be considered, but usually leads to persistent cyanosis of acral body parts. Unfortunately, the patient died from a bowel perforation in the setting of high-dose steroids.

Figure 1. A 71-year-old patient’s tongue showed features consistent with Raynaud’s phenomenon. Several times per day, her tongue showed both unilateral and bilateral cyanosis that resolved temporarily with the application of warm water. A shows bilateral cyanosis; B shows unilateral cyanosis on right with normal left side. Color photographs available from the author upon request.