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Article type: Clinical image

Received: June 18, 2019.

Accepted: June 25, 2019.

Published online: July 4, 2019.

ISSN: 1897-9483
Microvascular imaging of primary erythromelalgia

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Short title: Microcirculation in erythromelalgia

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Conflict of interest: none declared
Erythromelalgia is a rare microvascular disease characterized by recurrent severe burning pain and swelling accompanied by erythema of the extremities. It can be primary, or more frequently secondary. Primary erythromelalgia, also known as Mitchell’s disease, occurs spontaneously at any age. In contrast, secondary erythromelalgia is predominantly associated with myeloproliferative syndromes, connective tissue disorders or infectious diseases. Primary erythromelalgia is frequently caused by mutation of the voltage-gated sodium channel α-subunit gene SCN9A [1].

A 63-year-old woman presented with a 6-year history of paroxysmal attacks of burning pain accompanied by erythema affecting both hands and feet. She was physically active but in order to avoid pain, she routinely immersed her feet in cold water daily during the summer. She also simultaneously reported a 40-year history of Raynaud’s phenomenon with pallor involving all fingers on each hand.

Physical examination revealed erythema of both upper and lower extremities without evidence of skin disease. She did not exhibit any clinical or biological signs of connective tissue disease. Peripheral pulses and capillary refill time were normal as well as the nailfold capillaroscopic examination. Laboratory investigations, including full blood count, rheumatoid factor, CRP, antinuclear antibodies were normal. We made a diagnosis of primary erythromelalgia associated with primary Raynaud’s phenomenon, but the patient was not willing to take any drugs.

Laser Speckle Contrast Imaging (LSCI) is a technique that allows non-invasive and real-time monitoring of peripheral microcirculatory blood flow in a wide area of the body. LSCI gives very good reproducibility as well as excellent spatial and temporal resolutions [2]. In clinical studies, LSCI coupled with reactivity tests enables the estimation of endothelial and neurovascular function. LSCI is also an innovative tool to quantify microvascular response to pharmacological treatment [3].
**Figure 1** shows skin microvascular perfusion derived from speckle contrast analysis with colors ranging from blue (low perfusion) to red (high perfusion). LSCI provides a perfusion index proportional to skin blood flow and allows a record of up to 100 images per second [4]. At a temperature of 22°C, the patient demonstrated peripheral vasoconstriction. We induced a thermal hyperemia by raising room temperature from 22°C to 26°C. The patient showed a clinical paroxysm of erythromelalgia when the temperature exceeded a 25°C (photo) and it was preceded by a rapid increase in microvascular blood flux.

In the presented case we observed the coexistence of erythromelalgia and Raynaud’s phenomenon, which is very rarely reported in the literature as both conditions appear to be opposite in clinical symptomatology [1]. The measurements of skin blood flux coupled with reactivity tests enabled the observation of the microvascular dysfunction.

**References**


FIGURE 1 A - Measurement of the microvascular flux assessed by laser speckle contrast imaging on the right hand of a patient with primary erythromelalgia, at different room temperature. Please note that the middle finger had previously been treated with topical lidocaine/prilocaine cream and that the rise in skin blood flow was delayed. B - The left photo shows the hand of the patient together with the hand of a control subject on the right, at a room temperature of 26°C.