Reduced skin capillary density during attacks of erythromelalgia implies arteriovenous shunting as pathogenetic mechanism. By Drs. Mork C, Kvernebo K, Asker CL, Salerad EG.


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Erythromelalgia is characterized by burning pain, erythema, and increased temperature in acral skin. The pain is aggravated by warming and relieved by cooling. Increased microvascular arteriovenous shunting in deep dermal plexa has been hypothesized as the pathogenetic mechanism of pain in affected skin, inducing hypoxia during pain attacks. The aim of this study was to quantify skin capillary density in erythromelalgic patients before and after heat provocation, as increased skin temperature should increase the need for nutritive blood supply by the capillaries. Fourteen patients and 10 healthy control subjects were studied using an enhanced technique of computer-assisted analysis of capillary bed morphology and temperature measurements before and after central body heating. The increase in acral skin temperature was significantly higher (p < 0.05) in the eight patients where symptoms were induced after heat provocation, compared to asymptomatic patients and healthy control subjects. The number of visible capillaries in a field of view (1.7 mm2) decreased significantly (p = 0.01) in erythromelalgia patients from 105 (62-137) (median with total range) to 89 (49-118) after warming in areas with numerous arteriovenous anastomoses (nail bed region). In symptomatic patients, an even more significant reduction was observed (p = 0.01). The capillary size was also significantly reduced (p < 0.05) from 41.0 (31.5-50.5) (arbitrary units) to 37.3 (33.0-46.0) in symptomatic patients. The change in capillary density in the nail bed area was significantly larger in erythromelalgia patients (-17 (-49 to 39) compared to controls 0 (47 to 13) (p < 0.05), and in symptomatic patients -19 (-49 to -12) compared to asymptomatic patients -8 (-48 to 39) (p < 0.05) and controls (p < 0.01). The reduced skin capillary density after heating is compatible with increased microvascular arteriovenous shunting of blood and a corresponding relative deficit in nutritive perfusion (steal phenomenon) with skin hypoxia, causing the symptoms in erythromelalgia.
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