Lightning-Storm Periungual Capillaries in a Patient with Complex Regional Pain Syndrome Type I

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Established Facts

- Dermatologic changes may be the only physical exam findings in patients with complex regional pain syndrome type 1.
- Vasomotor instability is a key feature of complex regional pain syndrome.

Novel Insights

- Onychoscopy of the proximal nail fold in patients with complex regional pain syndrome may reveal capillary changes such as a “lightning-storm” appearance.

Keywords

Periungual capillaries · Onychoscopy · Complex regional pain syndrome · Capillaroscopy

Abstract

Introduction: Complex regional pain syndrome (CRPS) is characterized by chronic pain out of proportion to injury following trauma. CRPS most commonly results after crush-type injury to the hand and may be associated with various cutaneous symptoms believed to be driven by vasomotor instability. Existing reports in the literature have employed a range of methodologies to describe and evaluate cutaneous and vascular changes in CRPS, though there exists no

current gold standard for diagnosis. **Case Presentation:** Here, we report a 71-year-old male with a 10-year history of CRPS who presented with abnormal capillaries on onychoscopy, demonstrating a “lightning-storm” pattern. **Conclusion:** Previous studies have shown increased blood flow to the cutis in the early stage of CRPS, yet diminished cutaneous flow during the later stages. However, one study showed increased nail capillary diameter in patients with later stage CRPS. Due to a variety of reported clinical characteristics and a nonspecific clinic presentation, both the recognition and diagnosis of CRPS is difficult. Nail fold capillaroscopy may be a useful and accessible tool for evaluating patients with CRPS.

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Introduction

Complex regional pain syndrome (CRPS) is a chronic aberrant response to injury characterized by chronic pain out of proportion to the degree of injury, swelling, movement abnormalities, bone resorption, and cutaneous changes [1, 2]. Changes in color, temperature, and sudomotor activity are commonly observed; these changes are also in flux, as CRPS is a triphasic disorder [1]. CRPS is divided into type I (CRPS-I), which has no definable nerve lesion and corresponds to what was originally known as reflex sympathetic dystrophy (RSD), and type II, which has an identifiable nerve lesion present [3]. CRPS most commonly develops after fractures, sprains, surgery, vascular events, or crush injuries, most frequently to the extremities [4, 5].

CRPS is important to recognize in a dermatologic setting as cutaneous findings may be the only signs of the disease entity present on exam. Skin abnormalities may also become more prominent in later stages of the disease [5]. Notably, several hair, nail, and skin changes have been reported in patients with CRPS [5–11]. Patients may suffer from chronic edema of the affected extremity, recurrent skin erosions or ulcerations, erythematous papules, folliculitis, atrophy, dermatitis, nonimmune bullous eruption, or even develop cellulitis of the affected area [6, 9]. Nail changes that have been reported include a “whitlow-like” presentation [6], leukonychia [8], trachyonychia [11], and Beau’s lines [7]. Most cutaneous changes in CRPS are of vascular origin, thus directly visualizing changes in blood vessels in initial evaluation of a patient may be key for diagnosis.

Case Report

A 71-year-old male with a 10-year history of CRPS which developed after trauma to the proximal left lateral forearm presented to the dermatology clinic. Physical exam revealed longitudinal erythronychia with normal proximal and lateral nail folds. Photographs of the nail folds were taken with a Fotofinder. Onychoscopy demonstrated irregular capillary loops defined by giant dilated vessels in a tortuous “lightning-storm” pattern as well as loss of the normal parallel motif (Fig. 1, 2). Pinpoint loops were also noted along with avascular areas affecting the proximal nail fold. All finger nail units were involved (Fig. 3).

Discussion

Vasomotor instability is a cardinal feature of CRPS [1]. Vascular changes in CRPS have been observed via various methods and with variable findings. Early reports dem-

Fig. 1. Onychoscopy of the proximal nail fold of a patient with complex regional pain syndrome demonstrating a “lightning-storm” appearance.

onstrated increased flow on affected extremities observed via rapid-sequence blood flow studies with bone scintigraphy, suggesting enhanced perfusion [12]. Then skin’s microcirculation was also studied early on in RSD with videophotometric capillaroscopy. This, in contrast, showed low nail fold-skin capillary blood cell velocity (CBV) and laser Doppler flux (LDF) values in patients versus controls, despite equal skin temperatures in both groups [13]. This difference was most likely associated to a later disease stage in these patients compared to previous studies as well as the fact that capillary blood cell velocity/LDF measure blood flow in the outermost layer of skin, whereas the increased temperature that is frequently observed in RSD reflects changes in blood vessels below this layer.

Cooke and ward used photoplethysmography (PPG) in addition to LDF, to evaluate the finger pulps of patients with CRPS-I. Photoplethysmography showed a large microcirculatory volume, while LDF showed low microcirculatory velocity, consistent with dilated vessels that no longer respond to rhythmic bursts of sympathetic activity [14].

As RSD is a disorder of different stages, this may partly explain the conflicting findings. Kurvers et al. [15] sought to further describe these stages, utilizing LDF and

Fig. 2. Two affected nail folds demonstrating tortuous capillaries on onychoscopy in a patient with complex regional pain syndrome.

Fig. 3. Longitudinal erythronychia in a patient with complex regional pain syndrome.

in vivo video capillary microscopy to detail differences in skin blood flow in the three stages of disease: stage I, defined by patients with a chronic warmth sensation; stage II, an intermittent warmth and cold sensation; and stage III, a chronic cold sensation. Total skin blood flow was increased in stage I and decreased in stages II and III, while nutritive skin blood flow was decreased in stages II and III compared to controls [15]. This may explain the trophic changes seen in CRPS-I.

Interestingly, both capillary diameter and capillary density measured by in vivo video capillary microscopy in the nail fold of the most symptomatic finger did not differ between controls and affected patients and did not change as a function of the duration of the syndrome. Capillary diameter was found to be larger in stage III than in stage II and possibly stage I [15]. This is in contrast to findings detailed by Tosti et al. [6], the first to describe cutaneous histopathologic findings of the first stage of CRPS-I. Nail biopsy in their case showed a vascular nodule expanding the proximal nail fold, consisting of proliferating capillary vessels embedded in a loose edematous stroma with a surrounding inflammatory infiltrate [6]. This supported the suggestion that vasoproliferation on

top of vasodilation causes the increased blood flow which characterizes the early inflammatory stage of CRPS-I.

Several diagnostic criteria for CRPS exist, and research has shown a lack of agreement between these different diagnostic sets as well as differences in clinical features in patients who meet these different criteria [16]. Likewise, due to the wide range of methodologies used to describe the vascular changes in RSD along with a frequent lack of stage description in these studies, there remains a need to establish a simple and accessible gold standard for CRPS diagnosis.

We did not identify any other cases of dilated capillary loops visualized on onychoscopy in a patient with CRPS. This may be due in part to the lack of use of this technique in previously defined cases or evaluation of patients in different stages of disease. CRPS continues to be a disease entity with low visibility and a lack of recognition among dermatologists [5]. More research is needed to highlight the cutaneous features commonly seen with CRPS to allow dermatologists to distinguish it from other similar entities. Further description of the cutaneous features of CRPS will also promote timely diagnosis in patients with undiagnosed CRPS presenting to dermatology with cuta-

neous complaints. We believe careful examination of the nail unit with onychoscopy, and specifically capillaroscopy, may serve as a useful diagnostic clue when evaluating patients with suspected CRPS.

Statement of Ethics

The subjects have given their written informed consent to publish their case and the image herein.

Conflict of Interest Statement

We have no conflicts of interest to disclose.

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Author Contributions

M.P. and T.V. wrote the manuscript. A.C. was responsible for the onychoscopy and the image. M.P., T.V., A.C., and M.Z. contributed to concept and design. M.P., T.V., A.C., and M.Z. critically reviewed the manuscript, approved the final submission, and agreed to be responsible for all aspects of the manuscript.