Seminars in Arthritis and Rheumatism ▮ (2017) ▮▮▮–▮▮▮

Contents lists available at ScienceDirect

Seminars in Arthritis and Rheumatism

journal homepage: www.elsevier.com/locate/semarthrit

State of the art on nailfold capillaroscopy in dermatomyositis and polymyositis

Chiara Bertolazzi, MD[a], Maurizio Cutolo, MD, PhD[b], Vanessa Smith, MD, PhD[c], Marwin Gutierrez, MD, PhD[d,*]

[a] Division of Neurosciences, Instituto Nacional de Rehabilitación, Mexico City, Mexico
[b] Research Laboratories and Academic Division of Clinical Rheumatology, Department of Internal Medicine, University of Genova, IRCCS San Martino, Genova, Italy
[c] Department of Rheumatology, Ghent University Hospital, Ghent, Belgium
[d] Division of Musculoskeletal and Rheumatic Disorders, Instituto Nacional de Rehabilitación, Mexico City, Mexico

ARTICLE INFO

Keywords:
Capillaroscopy
Dermatomyositis
Polymyositis
Juvenile dermatomyositis
Inflammatory myositis
Disease activity

ABSTRACT

Objectives: To provide an overview of the main nailfold capillaroscopy (NFC) changes described in dermatomyositis (DM) and polymyositis (PM) and to discuss the current evidence supporting its clinical relevance and applications in daily practice.
Methods: All relevant literature in the field of NFC and DM and PM published in the last 30 years has been systematically reviewed. A systematic research was performed in the electronic databases PubMed and EMBASE.
Results: A total of 540 publications were identified according to the proposed filters and 27 were included for the review. The articles have been critically analyzed with a focus on technical aspects, examined anatomical areas, main pathological capillaroscopy findings ,and the relationship between NFC alterations and critical parameters of DM and PM.
Conclusions: The overview confirms that NFC is a safe and noninvasive tool able to help the clinician in the diagnosis of DM and PM and to better characterize the phase of disease activity of these patients.

Introduction

Inflammatory myopathies (IMs) are a group of acquired muscle diseases with heterogeneous features occurring both in children and adults [1].

Dermatomyositis (DM), polymyositis (PM), inclusion body myositis, overlap myositis, and myositis associated with cancer are the main nosological entities comprised in IMs definition, with difference in clinic, immunopathology, histology, and prognosis [2–4].

Nailfold capillaroscopy (NFC) is an in vivo, noninvasive and inexpensive imaging technique that allows direct observation of the capillary network in living tissue throughout the intact skin [5–7]. It has gained remarkable interest among rheumatologists because of its utility in both clinical practice and research activity [8–10].

To date, several studies have demonstrated the diagnostic and prognostic role of NFC in rheumatic disorders, especially in systemic sclerosis (SSc) and Raynaud phenomenon (RP) [11–15].

IMs, especially DM, offer an ideal scenario for the use of NFC due to its active involvement of vascular endothelium that induces a complement-mediated microangiopathy [2–4]. In fact, the characteristic NFC pattern named scleroderma (SD) pattern—that consists in a well-defined complex of alterations in distribution, shape, number, and dimension of capillaries—that has been typically described in SSc, has also been reported in other "scleroderma spectrum disorders" such as DM and mixed connective tissue disease [8,15–17]. Otherwise, the role of NFC in PM is currently not completely defined.

This may induce to consider that NFC may play a key role in providing essential findings for the management of PD and PM, and become a reference tool for the clinical decision making of these patients. On this light, the aim of this review is to provide an overview of the main NFC changes described in DM and PM (including adults and juvenile patients) and determine their

Abbreviations: NFC, nailfold capillaroscopy; DM, dermatomyositis; JDM, juvenile dermatomyositis; PM, polymiositis; IMs, inflammatory myopathies; SSc, systemic sclerosis; SD, scleroderma; RP, Raynaud phenomenon; GP, Gottron's papules; ILD, interstitial lung disease; NCD, nailfold capillary density; ERL, end row loops.

* Correspondence to: Marwin Gutierrez, MD, Instituto Nacional de Rehabilitación, Calzada Mexico-Xochimilco 289, Colonia Arenal de Guadalupe, CP 143898, Mexico City, Mexico.
E-mail address: dr.gmarwin@gmail.com (M. Gutierrez).

http://dx.doi.org/10.1016/j.semarthrit.2017.06.001
0049-0172/

potential implications in clinical setting (with particular focus on disease activity), in order to discuss the current evidence supporting its clinical relevance and applications in daily practice and to provide future research fields.

Methods

Research strategy

To identify all available studies, a detailed research pertaining to the topic of review was conducted according to PRISMA (Preferred Reporting Items for Systematic reviews and Meta-Analyses) guidelines [18]. A systematic research was performed in the electronic databases (PubMed and EMBASE), using the following search terms in all possible combinations: *capillaroscopy*, *videocapillaroscopy*, *nailfold capillary*, *inflammatory myopathies*, *myositis, polymyositis*, *juvenile polymyositis*, *dermatomyositis*, and *juvenile dermatomyositis*. The Boolean operators "AND" and "OR" were used in combining more key words to increase the specificity and reduce the sensitivity of our search. In addition, the reference lists of all retrieved articles were manually reviewed. In case of missing data, study authors were contacted by e-mail to try to retrieve original data. Two independent authors (C.B. and M.G.) analyzed each article and performed the data extraction independently. In case of disagreement, a third investigator (M.C.) was consulted. Discrepancies were resolved by consensus.

Inclusion and exclusion criteria

All relevant literature in the field of NFC and DM and PM published in the last 30 years has been systematically reviewed (a large period of review was considered since there are not previous reviews in this topic). We included original articles concerning studies in humans, published between January 1986 and June 2016. We excluded from this review the following types of publications: articles not published in English, case reports, letters to the editor and/or nonhuman studies. Research results were screened to avoid duplicates. Titles, abstracts, and full reports of articles identified were systematically screened with regard to inclusion and exclusion criteria.

Given the characteristics of the included studies, the evaluation of methodological quality of each study was performed with the Newcastle–Ottawa Scale (NOS), which is specifically developed to assess quality of nonrandomised observational studies [19].

Results

Approximately 540 publications were identified in PUBMED and EMBASE databases. The results of the research strategy are illustrated in Figure 1. Demographic data, number of patients enrolled and type of diseases included in the review studies are illustrated in Table 1 [20–46].

Furthermore, Table 2 resumes the technical aspects of NFC examinations adopted, characteristics of the device employed, and examined areas. Finally, Table 3 shows the main pathological capillaroscopy findings whereas Table 4 reports the relationship between NFC alterations and selected clinical aspects.

Juvenile dermatomyositis

Juvenile dermatomyositis (JDM) and adult DM share the disease characteristics of classical rash and muscle weakness. JDM presents some differences with respect to the adult variant, as the progression, that may be unicyclic (or transient) with resolution within 2–3 years, or chronic (polycyclic or progressive). Additionally JDM shows more frequently cutaneous calcifications (30% of patients), cutaneous ulceration (10% of patients), and intestinal vasculopathy that may lead to ulceration/perforation. On the contrary, important clinical features of adult DM such as interstitial lung disease (ILD) and malignancy are less common in younger adults and rarely seen in children [2–4].

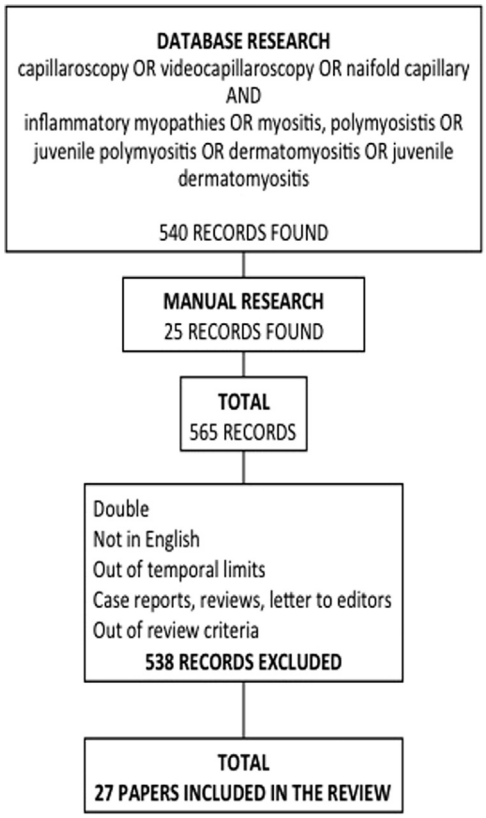

Fig. 1. Graphic of research strategy.

Main NFC changes in JDM

The NFC changes of the studies reviewed are described in Table 3.

The presence of a capillaroscopy SD pattern (adopted definitions of SD pattern are reported in Table 3) was described in 63–74% of JDM patients independently of disease activity whereas it was reported in 84–92% of patients with high activity state versus 0–38% in those in remission [31,32,43].

Ingegnoli et al. [31], in a comparative study, described a significant increase of major abnormalities ($p < 0.0001$) such as elonged, enlarged, giant, and bushy capillaries in JDM patients with respect to healthy children.

Dolezalova et al. [27] described mainly a reduced nailfold capillary density (NCD) (mean = 4.5 capillary/mm), wide capillaries and massive bushy and giant capillaries in 62 % patients with JDM. Scheja et al. [24] confirmed in their study a decreased NCD (median = 2.5 capillary/mm) associated with an increased capillary area (median = 2.5×10^{-3} mm^2) in all JDM patients with respect to healthy children (Table 3).

Schmeling et al. [40] in a cohort of 92 JDM patients showed a decreased NCD (mean 5.4 capillary/mm). Interestingly, the reduction of capillary density was greater in the subgroup of patients with new-onset JDM (mean 3.4 capillary/mm).

Piotto et al. [43] observed that the abnormal findings varied between the active and inactive phase of disease. In active disease,