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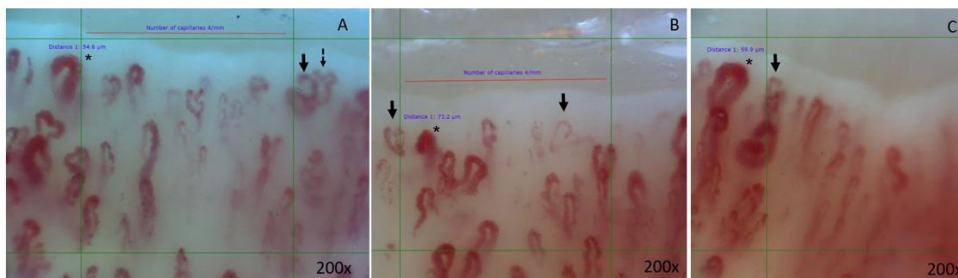
INTRODUCTION

A 59-year-old woman presented with a triphasic, bilateral Raynaud phenomenon (RP) on her follow up consultation for a known seropositive (RF+ and anti-CCP+), erosive, nodular, non-corticosteroid dependent rheumatoid arthritis (RA). Over 23 years, her disease was treated with corticosteroids, methotrexate, leflunomide, hydroxychloroquine, abatacept, alternative medicine, infliximab and lastly rituximab (RTX). Her RP has been stable for 30 years. Since the presence of secondary RP is described in RA patients, she never had a capillaroscopy in the past (1). Because she presented at the consultation with a prominent RP, the patient was re-evaluated and sent for a nailfold videocapillaroscopy (NVC).

RESULTS

NVC:

Giant capillaries (*) were found on several fingers together with a loss of capillary density and non-specific abnormalities including "bushy" capillaries (bold arrow) and concave tip (striped arrow) of single capillaries (Fig. A, B, C)



Clinical exam: rheumatoid nodules on both hands, skin exam completely normal
Serology: Over the last 10 years, she has had anti-nuclear antibodies at titers between 1:320 and 1:640 with 1x identification of anti-NOR90. RF+, anti-CCP+
Chest CT, lung function tests, echocardiogram: normal
RX both hands: band-like demineralization, bilateral metacarpophalangeal and interphalangeal impingement



DISCUSSION

In this patient with stable long-lasting secondary RP, NVC showed images compatible with those of a scleroderma spectrum disease (systemic sclerosis (SSc), dermatomyositis, mixed connective tissue disease, undifferentiated connective tissue disease). Although non-specific NVC abnormalities such as hemorrhages, visible subcapillary venous plexus, tortuous dilated capillaries and erythrocyte aggregations in the vessels have been described in RA, loss of capillary density and giant capillaries are indicative for a scleroderma pattern (2,3).

Although this patient did not meet the 2013 ACR/EULAR criteria for high probability of SSc diagnosis (score of 5), we concluded that in addition to her existing RA, she suffered from early SSc, or pre-scleroderma. Koenig *et al* (2008) and Valentini *et al* (2014) stipulate that a patient should suffer from RP associated with scleroderma marker autoantibody and/or typical capillaroscopy findings, without meeting either the 2013 ACR/EULAR criteria for SSc classification or the criteria for SSc sine scleroderma (she also had no internal organ involvement), to be labeled as affected by early SSc or pre-scleroderma (4,5). The progression to SSc in this patient was most likely limited due to several past RA therapies, particularly RTX. RTX improves skin fibrosis, stabilizes lung function and reduces the progression of microcirculation abnormalities in SSc patients (6,7).

CONCLUSION

- The presentation of (underlying) rheumatic diseases can be discrete in patients on immune modulating drugs
- Even in case of misleading clinical (or serologic) signs, NVC is a valuable tool to reveal (early) SSc diagnosis
- NVC should be performed in every suspected secondary RP
- RTX improves skin fibrosis, stabilizes lung function and reduces the progression of microcirculation abnormalities in SSc

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