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Capillaroscopic examination in the early diagnosis of systemic sclerosis

ABSTRACT

Systemic sclerosis (SSc) is a connective tissue disease characterised by impairment of vascular function. One of the examinations particularly useful in the early diagnosis of SSc is nail fold capillaroscopy. In SSc, the capillaroscopic findings show characteristic microcirculatory disorders defined as SSc-associated microangiopathy. The term microangiopathy includes the following pathological changes: decreased numbers of vessels with areas of avascularisation, widened vascular loops leading to the formation of megacapillaries, the presence of ramified loops and cap-shaped microextravasations. The important pathological changes occurring already in the early forms of SSc are megacapillaries Apart from early diagnosis, capillaroscopy is of prognostic importance, as the type of capillaroscopic lesions may correlate with disease activity and the degree of involvement of internal organs. In addition, capillaroscopy is used to monitor the course of SSc and the response to treatment.

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KEY WORDS: capillaroscopy; systemic sclerosis; Raynaud phenomenon

INTRODUCTION

Systemic sclerosis (SSc) is a multi-organ connective tissue disease characterised by impairment of vascular function and morphology with non-specific inflammation and progressive fibrosis. The vascular abnormalities in SSc can affect any internal organ, yet the gastrointestinal tract, lungs, heart and kidneys are most frequently involved [1]. The first clinical symptom of SSc is Raynaud's phenomenon, which occurs in 99% of SSc patients and often precedes the skin or organ changes for many years. Therefore, in the early period, SSc is difficult to diagnose [2]. One of the tests particularly useful in the early diagnosis of SSc is nail fold capillaroscopy applied in the differential diagnosis of primary Raynaud's symptom and microcirculatory disorders accompanying systemic connective tissue diseases, especially SSc [3, 4].

USEFULNESS OF CAPILLAROSCOPY

The examination consists of imaging the capillaries of the papillary layer within the nail fold of the second to the fifth finger of each hand under a stereoscopic microscope or video capillaroscope at 50–200-fold magnification [5]. The capillary density, vascular dimensions, vascular morphology and the presence of additional elements, such as micro-haemorrhages, are evaluated [4, 5]. The normal capillaroscopic image is characterized by a regular arrangement of capillaries and more than 7 capillaries per 1 linear mm; moreover, the shape of the capillaries should resemble a hairpin or an inverted letter U with a diameter of about 20 μm. The morphology of vascular loops may vary in different healthy individuals, but the abnormalities are not typical of SSc-associated microangiopathy. Changes in vascular morphology can include slightly dilated, tortuous or crossed vessels [5]. In SSc, the capillaroscopic findings show characteristic microcirculatory disorders defined as SSc-associated microangiopathy. The term microangiopathy includes the following pathological changes: decreased numbers of vessels with areas of avascularisation, widened vascular loops leading to the formation of megacapillaries, the

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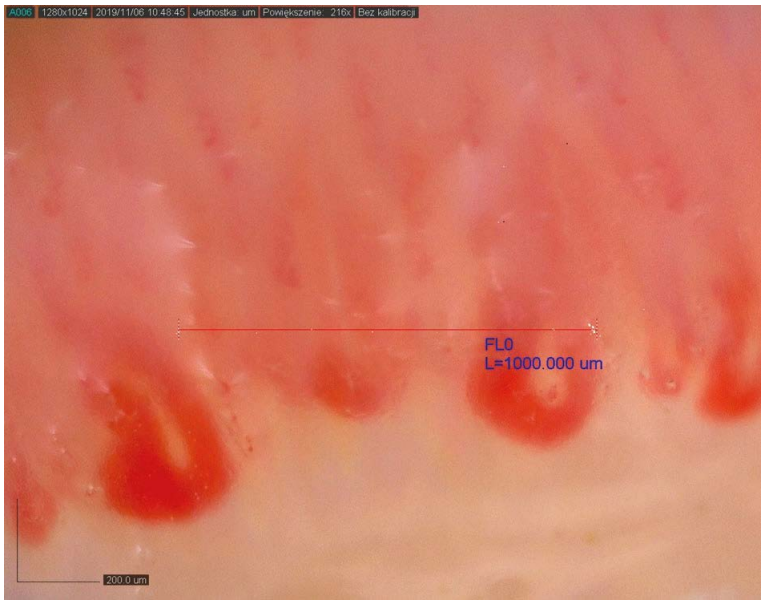

Figure 1. Megacapillaries

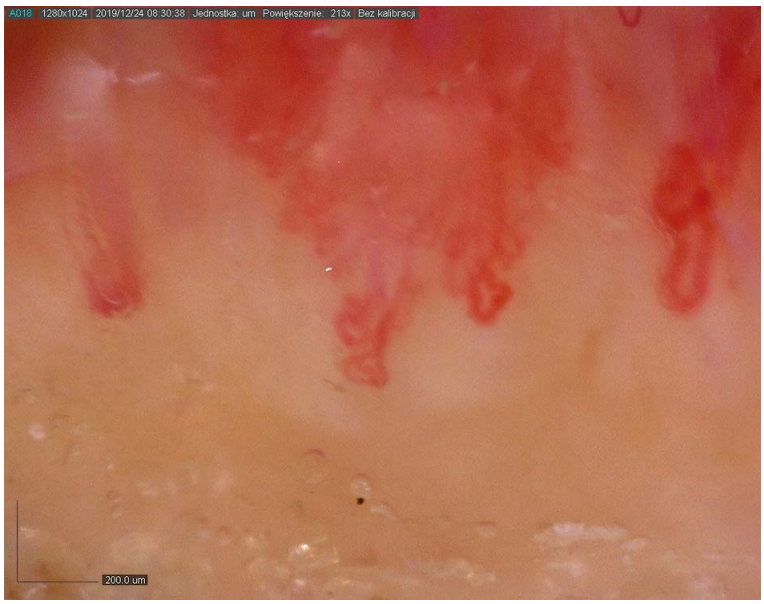

Figure 2. Neovascularisation

presence of ramified loops and cap-shaped microextravasations. The important pathological changes occurring already in the early forms of SSc are megacapillaries [4]. A megacapillary is defined as a homogenously dilated capillary with a diameter exceeding 50 μm (Fig. 1). When one megacapillary is observed in the patient, early SSc microangiopathy can be suspected; in such cases, observation of patients and thorough diagnostic procedures are recommended [6]. The second characteristic feature of SSc-related microangiopathy is the loss of capillaries and the presence of avascular areas [5]. Still another feature is neovascularisation, which is characterised by the presence of bushy, ramified or dendriform vascular loops (Fig. 2). These changes lead to derangement of the vascular array and often occur on the outer edges of avascularisation areas. The final feature of SSc-related microangiopathy, which is of lesser importance than the previously described ones, is the presence of cap-like extravasations, formed above the apical part of the capillaries and indicating damage to their wall [5]. In addition to its usefulness in differentiating primary and secondary Raynaud's phenomenon, nail fold capillaroscopy often allows one to determine the duration of

SSc or its activity [6]. Cutolo et al. have classified the capillaroscopic lesions into early, active and late patterns [7]. According to their classification, megacapillaries are typical of early SSc-related microangiopathy and are not found in the late period of the disease. As the disease progresses, the number of vessels tends to decrease; avascular areas and neovascularisation are observed, which is typical of active lesions. In the late stage of SSc, avascular areas and dendriform vessels predominate [5, 7]. Considering that patients with Raynaud's phenomenon and anticentromere and a-Scl-70 antibodies are a huge diagnostic problem, nail fold capillaroscopy is diagnostically important [7]. In 2012, new classification criteria were developed under the auspices of the European Alliance of Associations for Rheumatology (EULAR/ACR) in which capillaroscopic changes were included [8]. Numerous reports have demonstrated an association of capillaroscopic lesions with internal organ involvement and disease activity [9, 10]. In patients with active lesions, capillaroscopy significantly more often demonstrated pulmonary arterial hypertension and finger ulcers. The rapid dynamics of changes, with a tendency to reduce the number of vessels, especially during the first 3 years, correlates with a more severe course of the disease and quick development of interstitial lung disease [11]. Isolated reports have suggested a possible role of capillaroscopy in monitoring the treatment of SSc patients. In one study, an increase in the number of vessels and a slight decrease in avascular areas were observed in SSc patients after 3 years of iloprost treatment [12].

CONCLUSIONS

To sum up, the capillaroscopic examination is an inexpensive and non-invasive method particularly useful in the early diagnosis of SSc when the disease is difficult to detect, especially before the onset of skin and organ lesions. Moreover, capillaroscopy is of prognostic importance, as the type of capillaroscopic lesions may correlate with disease activity.

CONFLICT OF INTEREST

None declared.

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