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CASE REPORT

Capillaroscopic pattern in paraneoplastic Raynaud's phenomenon

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Abstract The capillaroscopic pattern in paraneoplastic Raynaud's phenomenon (RP) has not been investigated systematically and is not well-defined. Here, we present three case reports of patients with paraneoplastic rheumatic conditions, manifested with severe secondary RP with emphasis upon capillaroscopic findings. The first patient is a 58-year-old male with known psoriasis and psoriatic arthritis, severe RP and scleroderma-like syndrome, resulting in a paraneoplastic syndrome in the context of a lung cancer. At capillaroscopic examination classic "scleroderma" pattern, an "early" type was found. The second patient is a 48-year-old woman with an abrupt onset of paraneoplastic dermatomyositis, severe RP, and a lung cancer. The capillaroscopic examination revealed frequent dilated and giant capillaries, hemorrhages and severe microvascular disarrangement—the so-called "scleroderma-like" pattern typical of the idiopathic forms of the disease. The third patient is a 56-year-old woman with paraneoplastic dermatomyositis, secondary RP, and thyroid cancer. The capillaroscopic examination showed dilated and giant capillaries, elongated capillaries, decreased mean capillary density with avascular areas, severe disarrangement, single hemorrhages, and clear evidence of neoangiogenesis. These capillaroscopic features characteristic of the "scleroderma-like" pattern are indistinguishable from those in idiopathic dermatomyositis like in the second case. Taken together, the cases illustrate the problem that capillaroscopic patterns in paraneoplastic RP in the context of scleroderma-like syndrome and dermatomyositis appear to be indistinguishable from the microvascular changes in the respective idiopathic rheumatic diseases.

Keywords Capillaroscopy · Paraneoplastic · Raynaud's phenomenon

Introduction

The association of cancer with autoimmune phenomena is well-known in different rheumatic diseases such as polymyositis/dermatomyositis, Raynaud's phenomenon (RP), scleroderma-like syndromes, Sjögren's syndrome, inflammatory arthritis etc. [1–4]. Rheumatic disorders associated with cancer include different manifestations, most of which have no features distinguishing them from idiopathic rheumatic diseases. They may precede the clinical manifestation of the neoplasm, occur concomitantly or after its diagnosis [2, 4]. The association between malignancies and peripheral blood vessel diseases was described initially by Raynaud (1862) and Trousseau (1865). Ischemia of the fingers was reported in patients with carcinomas of the breast, stomach, and the esophagus, in leukemia patients and in patients with other malignancies. This phenomenon is caused by the secretion of vasoactive substances by the tumor cells and the respective immune response of the organism. A newly appeared RP after the age of 60 may be a paraneoplastic manifestation and may serve as a key hint

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for an underlying malignancy. In addition, RP may be a part of the manifestations of the paraneoplastic rheumatic condition [5, 6]. Nailfold capillaroscopy is an instrumental method with great value for the evaluation of the microcirculation in RP patients. However, the capillaroscopic pattern in paraneoplastic RP has not been defined yet. The most specific capillaroscopic pattern in rheumatology is those in systemic sclerosis. It is characterized by the presence of dilated and giant capillaries, hemorrhages, loss of capillaries, and neoangiogenesis. It was described for the first time by Maricq et al. [7], who termed these findings "scleroderma-type" capillaroscopic pattern. The same authors found that some of the parameters of this pattern can also be observed in other connective tissue diseases such as dermatomyositis, mixed connective tissue disease and termed these findings "scleroderma-like" pattern [7–9]. Cutolo et al. [8] described three phases of capillaroscopic changes in systemic sclerosis: "an early phase" with the appearance of few dilated and/or giant capillaries, few hemorrhages, relatively preserved distribution; "an active phase" with higher number of giant capillaries and hemorrhages; and "a late phase" with severe loss of capillaries and presence of ramified capillaries [9]. As the capillaroscopic pattern in paraneoplastic RP is not studied specifically and is not well defined, here we present three case reports of patients with paraneoplastic rheumatic conditions, who manifested with severe secondary RP (one man with scleroderma-like syndrome and two women with dermatomyositis) with a distinct emphasis on capillaroscopic findings.

Case report 1

A 58-year-old male patient presented with history for psoriasis and psoriatic arthritis with 15 years' duration. The patient has been treated with methotrexate 15 mg weekly. In the last 5 years the patient reported about increasing severe digital ischemia with pallor, cyanosis, reactive hyperemia, pain, and numbness during cold exposure. Skin thickening of the fingers appeared during the last year. At clinical examination, acroosteolysis of the distal phalanx of the 2nd and 3rd right fingers and sclerodactyly was found, without skin involvement proximal of the metacarpophalangeal joints, the lower extremities, face or trunk. Computed tomography revealed a tumor in the middle lobe of the right lung with a diameter of 3 cm with metastases in the lymph nodes of the right hilus. At the capillaroscopic examination, the classic "scleroderma-type" pattern was observed with the characteristic dilated and giant capillaries, single hemorrhages, slightly decreased mean capillary density, relatively preserved capillary distribution—all features that are specific for the "early" capillaroscopic pattern (Figs. 1, 2).

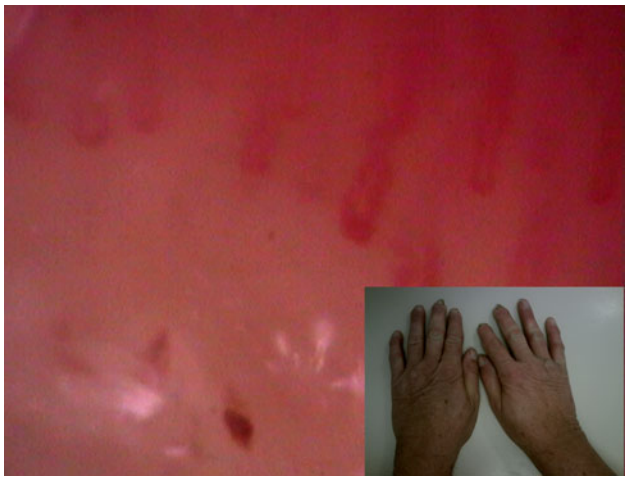

Fig. 1 "Early" scleroderma capillaroscopic pattern in a 58-year-old man with psoriasis, psoriatic arthritis, severe Raynaud's phenomenon, and scleroderma-like syndrome. Dilated and giant capillaries, hemorrhages, relatively preserved capillary distribution are demonstrated; magnification 200×, videocapillaroscope Videocap 3.0 (DS Medica, Italy). Sclerodactyly (skin edema and thickening distal of the metacarpophalangeal joints of the hands) and acroosteolysis of the distal phalanx of the 2nd and 3rd right fingers are evident at the picture of the lower right part

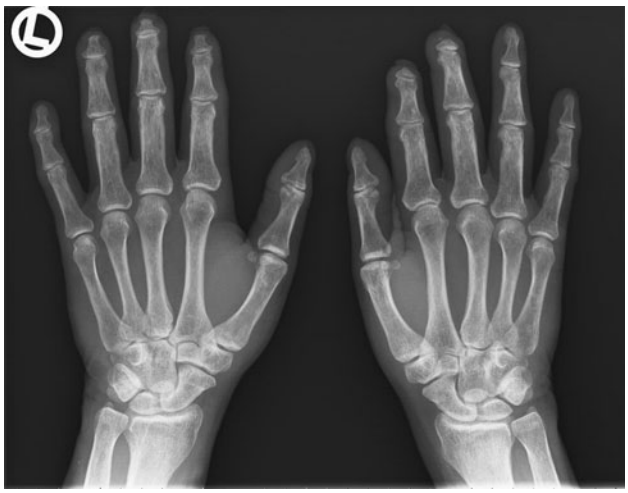

Fig. 2 X-ray of the hands of the same patient, presented at the fig. 1. Acroosteolysis of the distal phalanges is demonstrated

Case report 2

A 48-year-old woman with dermatomyositis, who referred to the Department of Rheumatology and Clinical Immunology, Kerckhoff Clinic, Bad Nauheim with a 5-month duration of proximal muscle weakness with abrupt onset that progressed rapidly and confined the patient to the bed. The muscle weakness had appeared simultaneously with classic skin rash and severe RP. Chest computed tomography detected a round formation in the right upper lobe and enlarged lymph nodes in the right hilus. The histological

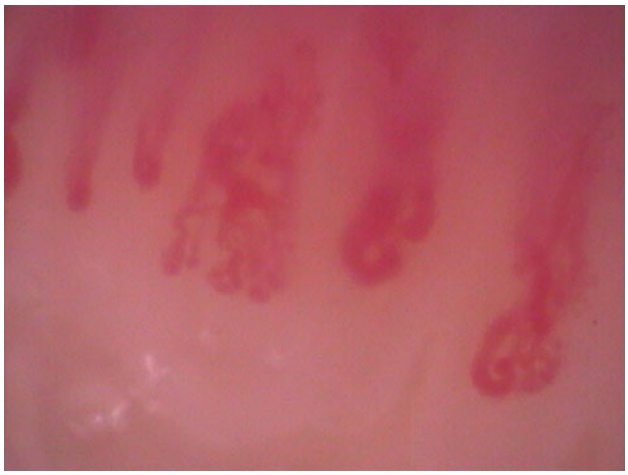

Fig. 3 Capillaroscopic pattern in a 56-year-old woman with paraneoplastic dermatomyositis associated with cancer of thyroid gland reveals dilated, giant capillaries, and neoangiogenesis; magnification 200×, videocapillaroscope Videocap 3.0 (DS Medica, Italy)

diagnosis of these lesions indicated differentiated papillar adenocarcinoma. The capillaroscopic examination showed frequent dilated and giant capillaries, hemorrhages and severe microvascular disarrangement—the so-called "scleroderma-like" pattern typical of the idiopathic forms of the disease.

Case report 3

A 56-year-old woman with a proven diagnosis dermatomyositis with secondary RP presented also in our outpatient clinic. One year later, a papillar carcinoma of the thyroid gland has been found and partial thyroidectomy has been performed. The capillaroscopic examination revealed dilated and giant capillaries, elongated capillaries, decreased mean capillary density with avascular areas, severe microvascular disarrangement, single hemorrhages, and a clear evidence for neoangiogenesis (Fig. 3).

These capillaroscopic features characteristic of the "scleroderma-like" pattern in dermatomyositis are indistinguishable from those in idiopathic dermatomyositis as in the second case.

Taken together, the cases illustrate the problem that capillaroscopic patterns in paraneoplastic RP in the context of scleroderma-like syndrome and dermatomyositis appear to be indistinguishable from the microvascular changes in the idiopathic rheumatic diseases.

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