

Successful Immunosuppressive Treatment of Dermatomyositis: A Nailfold Capillaroscopy Survey

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Vascular involvement is often present in dermatomyositis (DM). Severe nailfold capillaroscopy (NC) findings have been described but their association with disease activity and/or response to treatment have rarely been investigated^{1–7}.

A 40-year-old man presented 6 months earlier for arthromyalgias, asthenia, fatigue, and muscle weakness, which was more evident at the lower limb level. He displayed erythematous-desquamative lesions on both hands, face, and neck, and Raynaud's phenomenon.

Laboratory investigations showed erythrocyte sedimentation rate 54/h, C-reactive protein 48 mg/l, creatine kinase 530 U/l, lactate dehydrogenase 485 U/l, and anti-Jo-1 positivity. Electromyography examination of the upper and lower limbs revealed a myositic impairment with augmented junctional activity and presence of polyphasic potentials, while a muscle biopsy showed perivascular lymphomononuclear infiltrates with degenerative zones and necrosis.

The patient started 40 mg daily of 6-methylprednisolone (6-MP) but, owing to a progressive dyspnea, he was hospitalized. A pulmonary high-resolution computed tomography scan showed severe interstitial lung disease (ILD), more evident at periilar and basal levels, with a bilateral pleural effusion. Severely restrictive ventilatory impairment (forced vital capacity 55%; diffusion capacity for carbon monoxide 36%) and respiratory alkalosis were also present.

To evaluate Raynaud's phenomenon we performed an NC, showing abnormal morphology, the distribution and number of the capillary loops with marked architectural derangement, frequent ramified and bizarre capillary loops, megacapillaries, and neoangiogenesis alternating with avascular areas. Frequent microhemorrhages were also found (Figure 1).

A 6-month therapy with 750 mg/month cyclophosphamide (intravenous) was started, along with 80 mg/day 6-MP, grad-

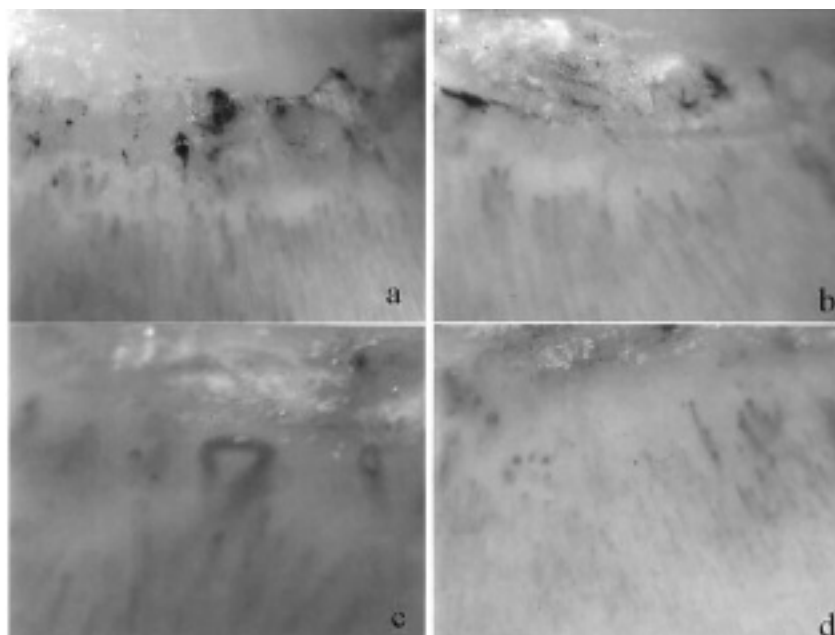


Figure 1. Nailfold capillaroscopy before immunosuppressive treatment, showing the index finger of the right hand (a), little finger, right hand (b), index finger, left hand (c), and ring finger, left hand (d). An abnormal morphology with irregular distribution and reduced number of capillary loops is shown, together with severe architectural derangement and presence of ramified and bizarre capillary loops in some fields. Megacapillaries, microhemorrhages, and neoangiogenic features are also visible associated with some avascular areas.

ually tapered to 8 mg/day. At the end of the treatment, the cutaneous lesions, strength, dyspnea, and muscle weakness improved. A mild basal interstitial involvement, with dyspnea on extended effort and restrictive lung impairment, was still present. Acute-phase reactants and serum muscle enzyme levels were within normal range. Table 1 shows the main clinical and laboratory changes during treatment.

The NC was repeated on the same section of the nailfold, by the same observer, with the same magnification, and under the same environmental conditions. It revealed marked improvement, with normalization in the number, morphology, and distribution of capillary loops and neovascularized

Table 1. Main clinical and laboratory data of the patient at the start and after 6-month immunosuppressive treatment.

	Before Treatment	After Treatment
Arthromyalgias	Present	Absent
Asthenia	Severe	Absent
Fatigue	Severe	Mild
Muscle weakness	Severe	Mild
Cutaneous lesions	Present	Improved
Raynaud's phenomenon	Present	Present
Dyspnea	Present at rest	On extended effort
ESR, mm	54	12
CRP, mg/l	48	Negative
CK, U/l	530	55
LDH, U/l	485	190

ESR: erythrocyte sedimentation rate; CRP: C-reactive protein; CK: creatine kinase; LDH: lactate dehydrogenase.

zones, and absence of microhemorrhages and giant capillaries (Figure 2). An immunosuppressive maintenance regimen was started with azathioprine 50 mg twice a day.

NC abnormalities are associated with skin involvement in untreated patients with DM³ and seem also to correlate with disease activity in inflammatory myopathies^{1,2}. DM-specific NC changes in a scleroderma-like pattern⁸ provide useful information that contributes to an early diagnosis and helps identify those cases with a poor prognosis⁴. Owing to the possible fatal outcome of severe ILD in DM, an aggressive immunosuppressive regimen is recommended^{5,9}.

Our case showed severe NC findings in a patient with DM and a clear improvement in these findings together with positive clinical and laboratory outcomes after immunosuppressive therapy, thus supporting the use of NC for diagnostic purposes and as a useful additional tool for followup and assessment of response to treatment in DM.

REFERENCES

1. Smith RL, Sundberg J, Shamiyah E, Dyer A, Pachman LM. Skin involvement in juvenile dermatomyositis is associated with loss of end row nailfold capillary loops. *J Rheumatol* 2004;31:1644-9.
2. Nascif AK, Terreri MT, Len CA, Andrade LE, Hilario MO. Inflammatory myopathies in childhood: correlation between nailfold capillaroscopy findings and clinical and laboratory data. *J Pediatr* 2006;82:40-5.
3. Christen-Zaech S, Seshadri R, Sundberg J, Paller AS, Pachman LM. Persistent association of nailfold capillaroscopy changes and skin involvement over thirty-six months with duration of untreated diseases in patients with juvenile dermatomyositis. *Arthritis Rheum* 2008;58:571-6.

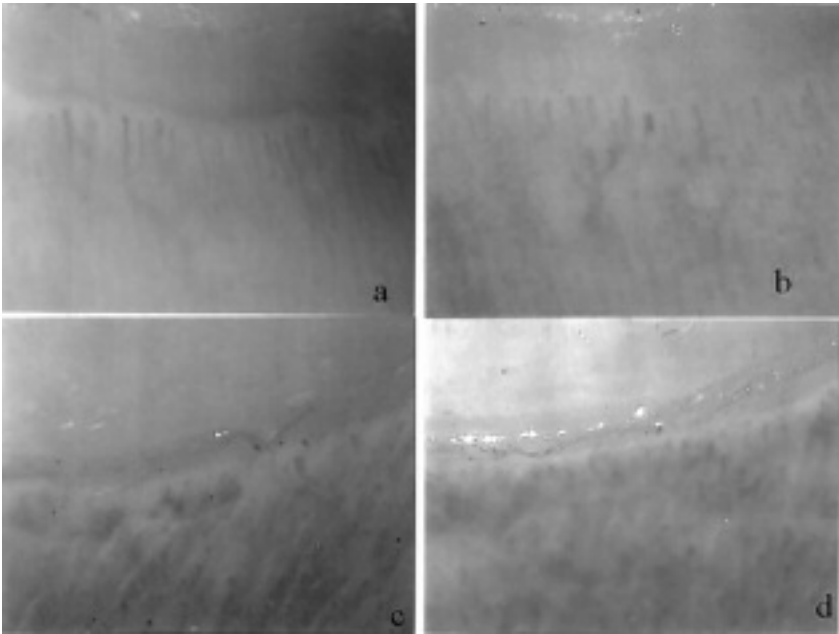


Figure 2. Nailfold capillaroscopy after 6 months of immunosuppressive treatment. (a) Index finger, right hand. (b) Little finger, right hand. (c) Index finger, left hand. (d) Ring finger, left hand. Marked improvement of capillaroscopic features is visible, with normal morphology, distribution and number of capillary loops, presence of revascularization, and absence of microhemorrhages and giant capillaries.

4. Selva-O'Callaghan A, Fonollosa-Pla V, Trallero-Araguas E, Martinez-Gomez X, Simeon-Aznar CP, Labrador-Horrillo M, et al. Nailfold capillary microscopy in adults with inflammatory myopathy. *Semin Arthritis Rheum* 2008 Nov 18. [E-pub ahead of print]
5. Fujisawa T, Suda T, Nakamura Y, Enomoto N, Ide K, Toyoshima M, et al. Differences in clinical features and prognosis of interstitial lung diseases between polymyositis and dermatomyositis. *J Rheumatol* 2005;32:58-64.
6. Aschwanden M, Daikeler T, Jaeger KA, Thalhammer C, Gratwohl A, Matucci-Cerinic M, et al. Rapid improvement of nailfold capillaroscopy after intense immunosuppression for systemic sclerosis and mixed connective tissue disease. *Ann Rheum Dis* 2008;67:1057-9.
7. Miniati I, Guiducci S, Conforti ML, Rogai V, Fiori G, Cinelli M, et al. Autologous stem cell transplantation improves microcirculation in systemic sclerosis. *Ann Rheum Dis* 2009;68:94-8.
8. Kameda H, Takeuchi T. Recent advances in the treatment of interstitial lung disease in patients with polymyositis/dermatomyositis. *Endocr Metab Immune Disord Drug Targets* 2006;6:409-15.
9. Cutolo M, Sulli A, Secchi ME, Olivieri M, Pizzorni C. The contribution of capillaroscopy to the differential diagnosis of connective autoimmune diseases. *Best Pract Res Clin Rheumatol* 2007;21:1093-108.