

# Microscopic polyangiitis with dermatomyositis

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## Abstract

Dermatomyositis is a rare autoimmune disease with a heterogeneous presentation that often has multiple extramuscular manifestations, although it does not typically involve the renal function. A 62-year-old female presented with proximal muscle weakness and rashes, which are classic symptoms of dermatomyositis without creatine kinase (CK) elevation. Initial serologic evaluation revealed a positive p-ANCA, although she did not develop renal failure for several months, at which point renal biopsy findings were consistent with microscopic polyangiitis. The patient was initially treated with cyclophosphamide, maintained with rituximab, and has been in remission for more than 2 years. Dermatomyositis and microscopic polyangiitis are both uncommon diseases, but are concomitantly present in this patient. A positive p-ANCA and development of renal insufficiency should be promptly evaluated in dermatomyositis patients.

**Keywords:** Dermatomyositis, microscopic polyangiitis, ANCA associated vasculitis, renal failure

## Introduction

Dermatomyositis is a rare autoimmune disease with a heterogeneous presentation that often has multiple extra muscular manifestations, although it does not typically involve the renal function. Here we report a 62-year-old female with proximal muscle weakness and rashes, which were classic symptoms for dermatomyositis without CK elevation. Initial serologic evaluation revealed a positive p-ANCA, although she did not develop renal failure for several months, at which point renal biopsy findings were consistent with microscopic polyangiitis (MPA).

## Case Presentation

In February, 2015, a 62-year-old Caucasian female noted reddish-purple lesions over her eyelids, metacarpal-phalangeal joints, and posterior neck, followed by progressive weakness, numbness, finger color changes with cold exposure, and extreme fatigue. In June, 2015, a rheumatologist diagnosed her with dermatomyositis (laboratory data in Table 1). She was started on prednisone 20 mg and mycophenolate mofetil 500 mg twice daily.

In August, 2015, she presented with progressive weakness and persistent rash. Examination revealed erythematous plaques over extensor surfaces, abnormal nail fold capillaroscopy, and mild bilateral lower extremity proximal weakness. Laboratory test data are shown in Table 1. A myositis panel was indeterminate for Mi-2, suggestive of p155/140 antibodies and several unidentified autoantibodies. Jo-1, double stranded DNA, Smith/RNP, SCL-70, and glomerular basement membrane antibodies were absent. C3 and C4 were normal. Urinalysis showed 2+ protein and 3+ blood with many red blood cells. Results of infectious disease evaluation were negative.

Magnetic resonance imaging of the thigh did not show focal muscle inflammation. CT of the chest, abdomen, and pelvis was negative for malignancy or acute pulmonary process. A muscle biopsy showed mild selective type II fiber atrophy, but no overt inflammation. A subsequent kidney biopsy revealed pauci-immune crescentic glomerulonephritis. Of eight glomeruli in the sample, five had cellular to early fibrocellular crescents. One arteriole demonstrated fibrin accumulation in the lumen and wall, suggestive of fibrinoid necrosis.

The patient was diagnosed with dermatomyositis and concomitant MPA and was treated with 3 days of sol-medrol 1 g, followed by prednisone 60 mg daily and one-time intravenous cyclophosphamide 700 mg, followed by daily oral cyclophosphamide 150 mg. She achieved near normalization of renal function by November, 2015. In January, 2016, she developed persistent neutropenia and cyclophosphamide was changed to rituximab 1000 mg every 2 weeks for two doses. Prednisone was gradually tapered and renal function completely normalized by March, 2016. She has been maintained on rituximab and remains in remission



**Cite this article as:** Bauer EM, Brahn E. Microscopic polyangiitis with dermatomyositis. *Eur J Rheumatol* 2017; 4: DOI: 10.5152/eurjrheum.2017.17008.

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Submitted: 10 January 2017

Accepted: 16 March 2017

Available Online Date: 29 August 2017

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**Table 1.** Progression of relevant laboratory values throughout current patient's course

Laboratory Testing (reference range)	June 2015	August 2015	May 2016
White Blood Cell (4.5-15.5×10 <sup>3</sup> cells/uL)	7.5	9.9	2.34
Hemoglobin (12-16 mg/dL)	13.1	9.6	13.0
Hematocrit (36%-46%)	40.5	28.9	39.4
Blood Urea Nitrogen (6-22 mg/dL)	23	58	21
Creatinine (0.5-1.6 mg/dL)	0.9	2.8	1.2
Urine Protein/Creatinine (<0.40)	n/a	1.21	0.8
Creatinine Kinase (38-282 U/L)	78	25	86
Aldolase (2.3-10.3 U/L)	4.3	4.3	12.3
P-ANCA	>8.00 (<1.00)	1:640 (<1:20)	<1:20
Myeloperoxidase antibody	n/a	79 (<20)	<20
C-ANCA	<0.20 (<1.00)	<1:20 (<1:20)	<1:20
Erythrocyte Sedimentation Rate (0-20 mm/h)	1	31	8
C-Reactive Protein	4.8 (0.0-2.5 mg/L)	1.4 (<0.8 mg/dL)	<0.3 (<0.8 mg/dL)

P-ANCA: perinuclear anti-neutrophil cytoplasmic antibodies; C-ANCA: cytoplasmic anti-neutrophil cytoplasmic antibodies

**Table 2.** Comparison of known cases of p-ANCA-positive renal vasculitis in patients with dermatomyositis

	Kawai et al. (1)	Yuste et al. (2)	Current Case
Demographics	60-year-old Japanese Male	47-year-old Female	62-year-old Caucasian Female
Duration of dermatomyositis diagnosis	10 years	3 months	7 months
Features of dermatomyositis	Heliotrope rash Gottron's sign Amyopathic	Heliotrope rash Proximal weakness Dysphonia Amyopathic	Heliotrope rash Gottron's sign Proximal weakness Amyopathic
Known associated antibodies	Jo-1-negative ANA-positive	ANA-negative	Jo-1-negative ANA-positive Mi-2 indeterminate P155/140-positive
Initial management of dermatomyositis	Prednisone 20 mg daily	Prednisone 40 mg daily Azathioprine 50 mg daily	Prednisone 20 mg daily CellCept 500 mg BID
Signs of RPGN	Fever, proteinuria, hematuria, elevated creatinine, renal biopsy confirmative	Proteinuria, hematuria, renal biopsy confirmative	Fever, proteinuria, hematuria, renal biopsy confirmative
p-ANCA/MPO	Positive	Positive	Positive
Initial management of ANCA vasculitis	Methylprednisolone 500 mg IV×3 days Plasma exchange	Prednisone 60 mg daily Cyclophosphamide	Methylprednisolone 1000 mg IV×3 days Cyclophosphamide
Maintenance therapy	Oral prednisone taper	Mycophenolate 500 mg BID Oral Prednisone taper	Rituximab Oral prednisone taper
Clinical outcome	Hemodialysis dependent	Progressive renal recovery	Renal recovery

RPGN: rapidly progressive glomerulonephritis; p-ANCA: perinuclear anti-neutrophil cytoplasmic antibodies/ myeloperoxidase; ANCA: anti-neutrophil cytoplasmic antibodies; ANA: antinuclear antibodies; BID: bis in diem

more than 2.5 years after onset, with a normal p-ANCA and myeloperoxidase antibody.

Written informed consent was obtained from the patient.

## Discussion

To our knowledge, there have been only two other reported cases of patients with dermatomyositis who subsequently developed an ANCA-associated vasculitis (AAV) (1, 2). Both prior patients also initially presented with dermatomyositis and minimal, if any, overt inflammatory muscle pathology. The patient described by Kawai et al. (1), in 2011, had a 10-year history of dermatomyositis, while the patient described by Yuste et al. (2), in 2014, developed both diseases within months of each other. As in the case presented here, both patients were p-ANCA/MPO-positive with confirmatory renal biopsies and without documentation of pulmonary involvement. Table 2 summarizes disease courses and management.

Dermatomyositis is a rare (incidence, 1.16-19 per million) and frequently heterogeneous disease characterized by proximal greater than distal muscle weakness, violaceous poikiloderma, typically involving the eyelids and malar area, anterior upper chest and back, extensor surfaces, and sun exposed areas (3). Many, but not all patients, will have elevation of muscle enzymes. Common extramuscular manifestations include polyarthritis, neuropathy, Raynaud's phenomenon, and interstitial lung disease (4). While AAV appears to be extremely rare in dermatomyositis patients, vasculitis of other organs is a known complication, especially in juvenile dermatomyositis patients. Vasculitis of the skin, lungs, gastrointestinal tract, testicles, muscle, cardiac, and central nervous systems has been reported (5-7).

Microscopic polyangiitis is a necrotizing, pauci-immune, small-vessel vasculitis, commonly resulting in glomerulonephritis and pulmonary capillaritis in the absence of granulomatous inflammation (8). The incidence is estimated at 5.9 per million individuals (9). The majority of MPA patients are positive for ANCA (95%) and predominantly directed to MPO (70%) (10). Evidence increasingly suggests that ANCA/MPO may directly contribute to the pathogenesis of MPA (11, 12).

ANCA-associated vasculitis has been reported in other connective tissue diseases, most commonly in scleroderma, followed by systemic lupus erythematosus, antiphospholipid antibody syndrome, rheumatoid arthritis, and mixed connective tissue disease (13, 14).

Dermatomyositis and MPA are rare diseases, and concomitant disease has been reported only twice previously. Non-ANCA-associated vasculitis, however, is not an uncommon complication of dermatomyositis. Our patient responded well to cyclophosphamide induction and rituximab maintenance. After more than 2.5 years, she remains in remission.

**Ethics Committee Approval:** N/A.

**Informed Consent:** Written informed consent was obtained from the patient who participated in this study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Data Collection and/or Processing - E.M.B., E.B.; Analysis and/or Interpretation - E.M.B., E.B.; Literature Search - E.M.B.; Writing Manuscript - E.M.B., E.B.; Critical Review - E.B.

**Conflict of Interest:** No conflict of interest was declared by the authors.

**Financial Disclosure:** The authors declared that this study has received no financial support.

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