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Amyopathic dermatomyositis presenting as severe alopecia and prominent eyelid edema: a case report and literature review

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Abstract

Amyopathic dermatomyositis (AD) is an autoimmune connective tissue disease of uncertain etiology that comprises 20% to 30% of dermatomyositis patients. Multiple studies suggest an association between malignancies and AD; moreover, some also report fulminant lung disease in patients with AD. We present a 62-year-old female patient with severe clinical presentation of AD, characterized by diffuse severe alopecia and pronounced symmetrical livid edema of the upper and lower eyelid, and positive anti-melanoma differentiation-associated gene 5 (anti-MDA-5) antibodies, who was successfully treated with concurrent therapy with hydroxychloroquine, methylprednisolone, and methotrexate as well as topical corticosteroids. Underlying malignancies or lung disease were excluded. Furthermore, we recommend vigilance in the key differential diagnosis of cutaneous lupus erythematosus because they can both present with similar clinical and histopathological features. The remission correlated with the complete absence of anti-MDA-5 antibodies.

Keywords: amyopathic dermatomyositis, alopecia, eyelid edema, anti-MDA-5 antibodies, methotrexate

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Introduction

Dermatomyositis is a rare autoimmune connective tissue disease of uncertain etiology. It most often affects both the muscles and the skin, but infrequently it may occur as amyopathic dermatomyositis (AD). Approximately 20% of dermatomyositis patients are estimated to have clinically amyopathic disease (1, 2). Dermatomyositis is classified as clinically AD, or *dermatomyositis sine myositis*, when muscular involvement does not develop within 6 months after the onset of cutaneous symptoms. However, muscle involvement has been known to occur at a later stage (1, 3, 4).

Case report

A 62-year-old female patient, a former smoker with a known history of arterial hypertension and chronic gastritis, had previously been evaluated by a regional dermatologist, resulting in a suspected clinical and histopathological diagnosis of discoid lupus erythematosus. She was treated with topical corticosteroids, topical immunomodulatory therapy, and systemic hydroxychloroquine for 5 months. Despite this therapy, her condition gradually progressed.

She reported that symptoms first appeared 3 years prior to presentation, beginning with a scaly rash of the scalp and progressive hair loss. Within 1 year, the disease worsened, leading to erythema of the face and scalp, diffuse frontal alopecia, and mild eyelid swelling. Due to the deterioration of her cutaneous manifestations, she was referred to the tertiary dermatology center.

At the tertiary dermatology center, she presented with severe heliotrope rash—symmetrical livid edema of the upper and lower eyelid—that obscured her vision. She had diffuse scaly erythema on the scalp and face, diffuse severe alopecia with a Severity of Alopecia Tool (SALT) score of 56.4, with involvement of the eyebrows, livid erythematous plaques on the anterior neck and upper chest (V sign), photodistributed poikiloderma involving the upper back (shawl sign), symmetrical erythematous livid plaques and papules on the elbows (Gottron sign), extensor surfaces of the metacarpophalangeal joints, and both the proximal and distal interphalangeal joints, which aligned with the clinical picture of Gottron papules (Fig. 1a–b), as well as mild fissuring with hyperkeratotic papules and scaling along the sides of the fingers and palm (mechanic's hands). Nailfold changes, including cuticular dystrophy, were observed. Dermoscopy revealed nailfold telangiectasias with dilated capillary loops, which alternated with vessel dropouts (Fig. 1c). In addition, dermoscopy of photoexposed lesions showed telangiectasias, and scalp dermoscopy indicated non-scarring diffuse alopecia with telangiectasias, empty follicles, perifollicular erythema, and diffuse hair shaft thinning (Fig. 1d). Dermatomyositis was suspected. Differential diagnoses included other connective tissue disorders, severe lupus erythematosus, severe contact dermatitis, and other photo-induced dermatosis. Upon admission to the inpatient department, thorough investigations were conducted. Histopathology of a skin biopsy revealed superficial vacuolar interface dermatitis with an increased production of mucin in the superficial part of dermis, which coincided with the diagnosis of dermatomyositis; nevertheless, it is not diagnostically specific. The result of direct immunofluorescence microscopy was minimal and not significant. Furthermore, a scleroderma-like pattern observed during nailfold capillaroscopy suggested the presence of a systemic autoimmune disease. The patient denied proximal muscle weakness, which was further assessed through targeted clinical examination, including manual muscle testing (MMT8 test 148/150 points). Electromyography (EMG) was normal, and there were no laboratory signs of muscle inflammation (normal creatine kinase, myoglobulin, aldolase, lactate dehydrogenase, aspartate aminotransferase, and alanine aminotransferase levels).

The patient's immunoserology tests were negative for anti-

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nuclear antibodies, extractable nuclear antigen antibodies, levels of C3 and C4 complement, CH50 test, antiphospholipid antibodies, and anti-double-stranded DNA antibodies, but low positive for anti-melanoma differentiation-associated gene 5 (anti-MDA-5) antibodies (62 AU). There was polyclonal hypergammaglobulinemia (18 g/l, reference value 5.9–15.1 g/l), but other laboratory tests were insignificant (blood count, C-reactive protein, erythrocyte sedimentation rate, electrolytes, blood urea nitrogen, creatinine, liver function panel, blood lipid profile, thyroid hormones, protein electrophoresis, tumor markers, chitotriosidase, endomysial antibody IgA, tissue transglutaminase antibody IgA, tryptase, NT-proBNP, troponin, hemoglobin A1c, magnesium, calcium, QuantiFERON-TB test, serology tests for HIV and hepatitis B and C, and urinalysis).

Based on the characteristic chronic cutaneous features, dermoscopic and capillaroscopic findings, supportive histopathology, anti-MDA5 seropositivity, and absence of clinical, laboratory, and electrophysiological evidence of muscle involvement, the final diagnosis of AD was established.

Paraneoplastic etiology was excluded with extensive laboratory tests, endoscopic examinations (colonoscopy, esophagogastroduodenoscopy, and bronchoscopy), radiologic imaging (chest X-ray, chest computed tomography (CT), abdominal ultrasound, and transvaginal ultrasound), and specific cytopathological investigations (Papanicolaou smear, fine needle aspiration of a suspicious axillar lymph node found on chest CT, and urine cytology).

Specific lung investigations, including bronchoscopy, chest CT, and pulmonary function tests, revealed signs of chronic bronchitis, which could be consistent with the patient's history of smoking.

Due to the severe chronic cutaneous clinical picture and insufficient regression with hydroxychloroquine and intensive topical corticosteroids, concurrent therapy with methylprednisolone in a decreasing dose (initiating at a dose of 32 mg [0.5 mg/kg] daily for 4 weeks) and methotrexate (10 mg once weekly, followed by an increase to 15 mg per week) was prescribed on top of hydroxychloroquine (200 mg daily) and topical therapy with corticosteroid ointment. At the follow-up appointment 1 month later, significant clinical regression was already evident, with persistent amelioration over the subsequent months. By 4 months, there was marked reduction in eyelid edema and livid erythematous plaques on the neck, upper chest, elbows, and hands. In addition, clinical and dermoscopic improvement in alopecia with noticeable hair regrowth and less prominent telangiectasias was observed.

After 6 months of methotrexate treatment, although substantial improvement had been achieved, residual cutaneous disease persisted. Therefore, to enhance long-term disease control and prevent relapse, the methotrexate dose was increased to 20 mg per week. The patient continued to tolerate treatment well. The therapy consisted of methotrexate (20 mg weekly), hydroxychloroquine (200 mg daily), and low-dose methylprednisolone (alternating 4 mg and 2 mg daily).

Potential drug interactions and safety considerations were

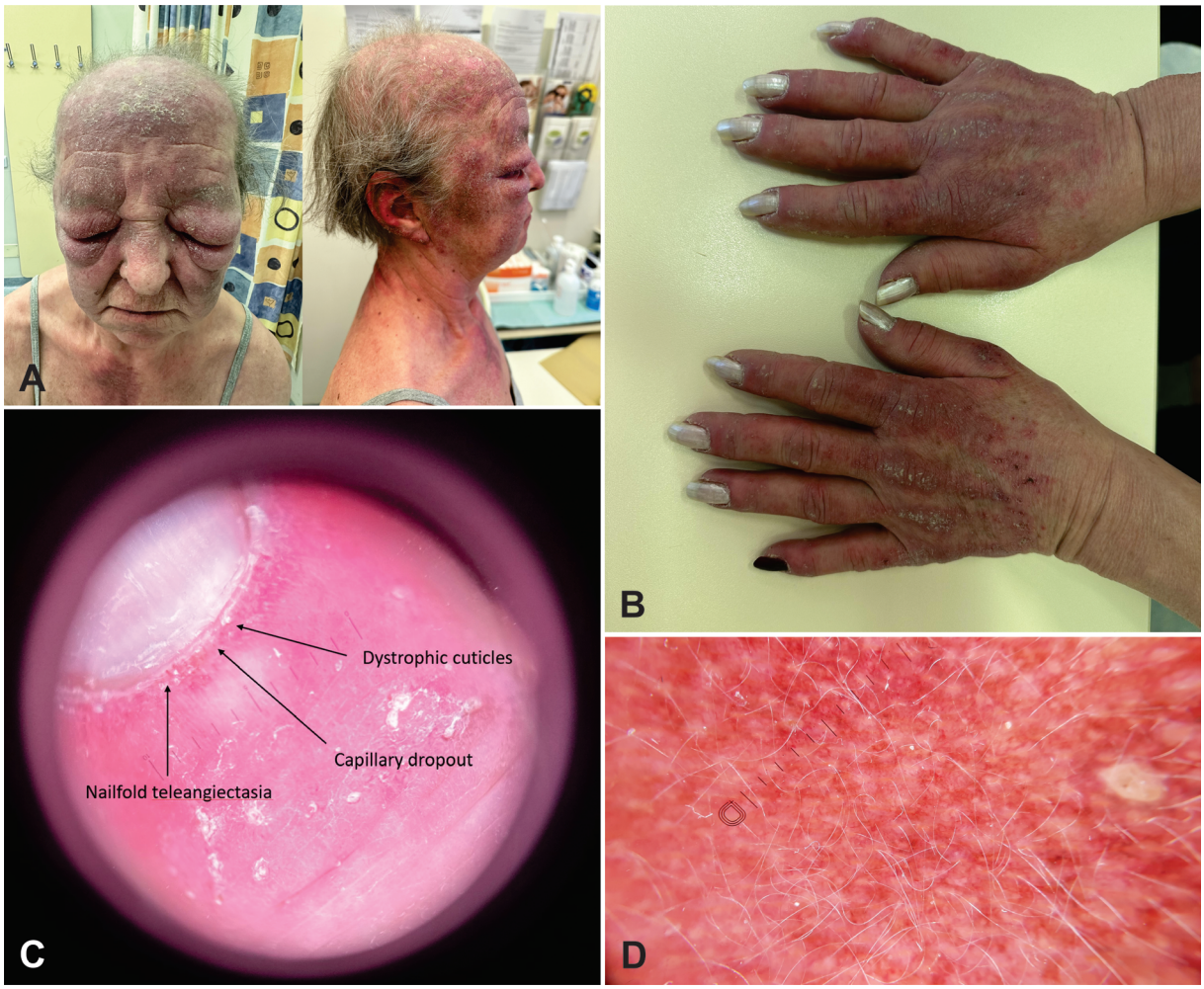


Figure 1 | a) Clinical picture of a severe symmetrical livid edema of the upper and lower eyelid, diffuse scaly erythema on the scalp and face, diffuse severe alopecia, livid erythematous plaques on the neck and upper chest (V sign), photodistributed poikiloderma (shawl sign); b) clinical picture of mechanic's hands with the presence of Gottron papules; c) dermoscopy of nailfold changes: nailfold telangiectasias with dilated capillary loops, which alternated with vessel dropouts; d) scalp dermoscopy: non-scarring diffuse alopecia with telangiectasias, empty follicles, perifollicular erythema, and diffuse hair shaft thinning.

continuously assessed. The combination of methotrexate and hydroxychloroquine is commonly used in dermatomyositis; however, potential additive hepatotoxicity and increased infection risk require regular laboratory monitoring (5, 6). Therefore, the routine follow-up included complete blood count, liver and renal function tests, and clinical surveillance for infectious complications. Ophthalmologic examination was performed to exclude retinopathy due to the long-term use of hydroxychloroquine. Folic acid supplementation was provided alongside methotrexate therapy.

At the 1-year follow-up following the initial diagnosis, the patient's skin changes showed continued improvement and a SALT score of 0 (Fig. 2a–b), there were no clinical or laboratory signs of malignancy, and the previously performed malignancy screening and diagnostic investigations were repeated, again showing no evidence of neoplastic disease. There remained no evidence of muscle involvement; in addition, magnetic resonance imaging (MRI) and ultrasound of the muscles of the upper and lower extremities were performed and revealed no signs of myositis. Evaluation by a pulmonology specialist revealed no evidence of lung involvement attributable to dermatomyositis. Moreover, repeat testing at the 1-year mark demonstrated seroconversion, with previously positive anti-MDA5 antibodies becoming negative.



Figure 2 | a) Clinical improvement at the 1-year follow-up: reduction in eyelid edema, noticeable hair regrowth and less prominent teleangiectasias; b) clinical improvement at the 1-year follow-up: regression of Gottron papules and clinical presentation of mechanic's hands.

Discussion

Clinically dermatomyositis is characterized by photodistributed eruption. Cutaneous manifestations may include Gottron sign and Gottron papules, mechanic's hands, nailfold changes, scalp erythema and scale, non-scarring alopecia, heliotrope rash, eyelid edema, and characteristic patterns of poikiloderma (7–10). Photodistributed poikiloderma is a distinctive feature of dermatomyositis, frequently affecting the upper chest (V sign) and upper back (shawl sign) (10). Anti-MDA5 antibody-associated AD is characterized by distinctive cutaneous manifestations, including mucocutaneous ulcerations, palmar papules, nonscarring alopecia, and panniculitis, which are likely attributed to severe underlying vasculopathy (11), some of which were seen in our patient.

Scalp involvement is increasingly recognized as a common manifestation of dermatomyositis, reported in up to 63% to 82% of patients. Studies have demonstrated associations between specific myositis-related antibodies and scalp disease, including alopecia and erythema, particularly in transcriptional intermediary factor 1-gamma (TIF1)-, nuclear matrix protein (NXP2)-, and MDA5-positive dermatomyositis, with approximately 13% of alopecia cases reported in association with MDA5 positivity (12). Consistent with these observations, our anti-MDA5–positive patient presented with marked scalp erythema and non-scarring alopecia, further highlighting the relevance of scalp disease in this subtype.

The diagnosis of dermatomyositis is based on clinical examination supported by various investigations. These may include assessment of muscle enzymes, detection of autoantibodies, electromyographic studies, imaging techniques, histopathological analysis of skin and muscle, and evaluation for underlying malignancy (13, 14). In patients with AD, tests for muscle involvement, including muscle enzyme levels, EMG, and MRI, per definition show normal results (13). Nonetheless, muscle biopsy can still have a diagnostic role in selected cases, particularly when there is diagnostic uncertainty, atypical clinical presentation, discordant test results, or suspicion of subclinical muscle involvement (5, 15, 16). In our patient, proximal muscle weakness was absent, laboratory evaluation demonstrated normal muscle enzyme levels, and EMG findings were normal; therefore, muscle biopsy was not performed because the likelihood of clinically significant myositis was considered very low. Nevertheless, individuals presenting with normal muscle enzyme levels should be monitored at regular intervals because they may harbor subclinical myopathy, progress to classic dermatomyositis, or persist with amyopathic dermatomyositis (4). In the absence of muscular symptoms, dermatomyositis can be a very difficult diagnosis (8). The only validated criteria for AD from the European League Against Rheumatism (EULAR) / American College of Rheumatology (ACR) require the presence of two out of three pathognomonic cutaneous features

(Gottron sign, Gottron papules, heliotrope rash) and the absence of muscle involvement for diagnosis. However, these criteria may fail to identify up to 25% of AD cases, leading to misdiagnosis, often with lupus erythematosus (16), due to the similarity in histologic findings with dermatomyositis (17). There is a need for re-evaluation and refinement of the current criteria to enhance sensitivity and specificity in diagnosing AD (16).

The detection of myositis-specific autoantibodies plays a critical role in informing the diagnosis, prognosis, and therapeutic approach to dermatomyositis because several of these antibodies are strongly associated with particular clinical subtypes. Numerous studies have identified a correlation between anti-MDA-5 antibodies and AD (8, 9, 11, 18–21). MDA5 is a recently identified autoantigen target in a subset of patients with dermatomyositis (9). Moreover, studies indicate that approximately 40% to 60% of patients with clinical AD are positive for anti-MDA5 antibodies (22, 23). This prevalence appears to be even higher in East Asian populations, with some cohorts reporting rates as high as 70% to 80%, particularly among patients that develop rapidly progressive interstitial lung disease (RP-ILD) (24–26). Nevertheless, a novel MDA5 antibody-associated dermato-pulmonary syndrome has also been reported in non-Asian populations (27). Given that interstitial lung disease associated with clinically AD is often resistant to intensive immunosuppressive treatment and may exhibit a rapidly progressive and potentially life-threatening course (11, 18, 19, 28), evaluation and re-evaluation for pulmonary involvement is mandatory (8, 11).

Multiple studies have demonstrated that anti-MDA5 antibody titers tend to decline gradually in response to treatment (9, 18–21, 29). Some reports indicate a reduction within the initial months of therapy (29); however, normalization is generally observed following sustained and effective treatment, often reflecting a slower rate of decline (9, 29). These findings support the potential utility of anti-MDA5 antibodies as biomarkers for monitoring disease activity, assessing therapeutic response, and predicting relapse (9, 18–21, 29). Conversely, a study by Yoshiyuki Abe et al. reported an early decline in anti-MDA5 titers in most patients, including those with fatal outcomes, raising concerns about the reliability of this marker for guiding treatment decisions in dermatomyositis (30). Although a decline in antibody titers may be associated with clinical improvement, the relationship between seroconversion and disease activity remains incompletely understood (9, 18–21, 29). Therefore, frequent and serial monitoring of anti-MDA5 antibody levels, in conjunction with comprehensive clinical evaluation, is essential for understanding their correlation with disease activity and for guiding therapeutic decisions and assessing prognosis.

Inflammatory myopathy patients are at higher risk for cancer, particularly within the 3 years preceding diagnosis and the 3 years following diagnosis (31). Multiple studies suggest an association between malignancies and AD; the reported frequency varies from 15% to 25%. Genitourinary malignancies, particularly ovarian carcinoma, and colorectal cancer appear to be more prevalent (32, 33). In addition, in certain Southeast Asian populations, there is a higher prevalence of nasopharyngeal carcinoma (34). Other commonly identified malignancies include breast, pulmonary, gastric, and pancreatic cancers, as well as lymphomas, including non-Hodgkin lymphoma (33). Paraneoplastic etiology should be therefore evaluated on a regular basis further in high-risk patients, ideally once a year, with particular emphasis during the initial 3 years after diagnosis (14).

Although clear guidelines for the management of patients with AD are lacking, some recommendations exist; accordingly, a selection of investigations that may be clinically appropriate for these patients is presented below (Table 1) (4, 5, 15, 25, 31, 35–36).

It is well recognized that the cutaneous manifestation of dermatomyositis is challenging to treat and often requires a multifaceted and multidisciplinary approach. The management of AD differs significantly from that of classic dermatomyositis because systemic corticosteroids are not typically used. Treatment options include topical corticosteroids, topical tacrolimus, antimalarial therapy (used as monotherapy or in combination), low-dose weekly methotrexate, mycophenolate mofetil, intravenous immunoglobulin (IVIg), dapsone, tofacitinib, and thalidomide (6, 8, 37–43). Hydroxychloroquine is commonly used as the initial treatment, followed by methotrexate or mycophenolate mofetil, with IVIg reserved for refractory cutaneous dermatomyositis (6).

Moreover, individuals with dermatomyositis are at elevated risk for hydroxychloroquine-induced cutaneous adverse reactions compared to those with lupus erythematosus (5, 44). Nonetheless, hydroxychloroquine continues to be frequently utilized as a first-line treatment, and it is advisable to provide patients with appropriate pre-treatment counseling (44).

However, clinical evidence supporting a definitive treatment algorithm remains limited (6, 11). In addition, using high-protection sunscreen, wearing protective clothing, and minimizing sun

Table 1 | Recommendations on additional investigations to evaluate a potential paraneoplastic etiology (5, 31, 35, 36) and muscle (4, 5, 15) and lung involvement (5, 25).

Paraneoplastic etiology	Muscle involvement	Lung involvement
Extensive laboratory tests: complete blood count, serum liver function tests, serum erythrocyte sedimentation rate / plasma viscosity, serum C-reactive protein, tumor markers: PSA (men), CA 125 (women), CA 19-9 (men and women); protein electrophoresis, lactate dehydrogenase, urinalysis, fecal occult blood tests	Neurological and sequential visits by muscle group assessment	Pulmonary function tests with CO diffusion
Radiologic investigations: chest X-ray, chest CT, abdominal ultrasound, transvaginal ultrasound, abdominal and pelvic CT, mammogram	Muscle enzyme levels: serum creatine kinase, aldolase, (myoglobulin, lactate dehydrogenase, aspartate aminotransferase, alanine aminotransferase)	Chest CT/HRCT
Endoscopic investigations: colonoscopy, esophagogastroduodenoscopy, (bronchoscopy)	EMG	(Bronchoscopy, depending on chest CT result)
Specific cytopathological investigations: Pap smear, urine cytology, fine needle aspiration of axillar/other lymph node, depending on results of previous investigations	MRI; muscle biopsy, depending on results of previous investigations	

CO = carbon monoxide, CT = computed tomography, EMG = electromyography, HRCT = high-resolution computed tomography, MRI = magnetic resonance imaging, PSA = prostate specific antigen, CA = cancer antigen.

exposure are crucial for managing cutaneous manifestations (8).

The therapeutic strategy in our case was determined in collaboration with a rheumatologist. Because the initial topical corticosteroid therapy and antimalarial were not effective, oral corticosteroids and methotrexate were added with good clinical response. Low-dose methylprednisolone was added due to the severity of the cutaneous involvement, including marked eyelid edema, extensive poikiloderma, and severe alopecia, and was used as bridging therapy to achieve earlier clinical stabilization while awaiting the full therapeutic effect of methotrexate. Because there was no systemic anti-inflammatory activity or definite lung involvement and the anti-MDA5 antibody concentration was rather low, we continued with the selected therapy, which proved effective and resulted in disease remission and seroconversion to a negative anti-MDA5 antibody status.

Conclusions

Diagnosis of AD may be tricky at times. The clinician must remain vigilant in the key differential diagnosis of cutaneous lupus erythematosus because both diseases can present with similar clinical features, clinical course, and histopathology, and without myopathy. Severe alopecia of the scalp with prominent eyelid edema can be one of the key presentations in AD, which aid in the vast differential diagnosis and may encourage the clinician to search for other key features of dermatomyositis. When the diagnosis is established, extensive tests must be performed to exclude possible paraneoplastic etiology and lung involvement. Achieving regression in AD is difficult, and both intensive topical and concurrent three-track systemic therapy may be indicated in refractory cases to achieve remission.

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