

Unilateral Gottron's Sign as a Diagnostic Challenge in Amyopathic Dermatomyositis: A Clinicopathological Study

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Abstract: Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by distinctive cutaneous manifestations and varying degrees of muscle involvement. **Gottron's papules and Gottron's sign** are considered pathognomonic and typically present symmetrically over extensor surfaces. Unilateral involvement is exceedingly rare and may delay diagnosis, particularly in cases of **amyopathic dermatomyositis (ADM)** where muscle enzymes remain normal. We report and analyze a rare presentation of unilateral Gottron's sign in a 70-year-old male subsequently diagnosed with anti-MDA5 positive ADM associated with interstitial lung disease (ILD). The patient presented with an 18-month history of asymptomatic violaceous plaques localized to the right hand, with additional lesions over elbows and knee, without objective muscle weakness. Laboratory evaluation revealed negative ANA and normal creatine kinase levels, consistent with ADM subsets (1, 1999). Extended myositis antibody profiling demonstrated positivity for **anti-MDA5 antibody**, a serological marker strongly associated with clinically amyopathic disease and ILD risk (2, 2011). Histopathology showed interface dermatitis with basement membrane thickening and lymphomononuclear infiltrate, supporting connective tissue disease pathology. High-resolution computed tomography confirmed bilateral basal ILD changes. The patient responded favorably to systemic corticosteroids and hydroxychloroquine. This case highlights the diagnostic complexity posed by unilateral cutaneous manifestations and underscores the importance of comprehensive serological and pulmonary evaluation even in the absence of classical symmetric lesions. Recognition of atypical presentations is crucial, as anti-MDA5 positive ADM carries significant systemic morbidity (3, 2021). Early diagnosis and immunomodulatory therapy remain essential to improve prognosis and prevent progressive lung involvement.

Keywords: *Dermatomyositis; Amyopathic dermatomyositis; Gottron's sign; Anti-MDA5; Interstitial lung disease; Unilateral rash*

Introduction: Dermatomyositis (DM) is a systemic autoimmune inflammatory disorder characterized by pathognomonic cutaneous manifestations and proximal muscle weakness. It belongs to the spectrum of idiopathic inflammatory myopathies and may involve multiple organ systems including lungs, gastrointestinal tract, and cardiovascular system (11, 2015). Classic dermatological signs such as Gottron's papules, heliotrope rash, shawl sign, and periungual telangiectasia often precede muscle involvement. Gottron's sign refers to violaceous erythema over extensor surfaces of joints and is considered highly specific for DM (1, 1999). Typically, these lesions are symmetrical; unilateral distribution is extremely uncommon and may mimic other papulosquamous disorders including lichen planus, psoriasis, contact dermatitis, or lupus erythematosus. Amyopathic dermatomyositis (ADM) represents a subset wherein characteristic cutaneous findings persist for at least six months without clinical or laboratory evidence of muscle involvement (13, 2006). ADM accounts for approximately 20% of DM cases and poses diagnostic difficulty due to absence of elevated muscle enzymes. Anti-MDA5 antibody-associated ADM is increasingly recognized as a distinct phenotype linked to rapidly progressive interstitial lung disease (ILD) and vasculopathic cutaneous ulcerations (2, 2011). Recognition of this subset is critical because pulmonary involvement significantly influences morbidity and mortality (9, 2020). Several reports have described unilateral heliotrope rash in anti-MDA5 positive dermatomyositis (3, 2021; 4, 2022), yet unilateral Gottron's sign remains rare. Proposed mechanisms for asymmetric distribution include localized trauma (Koebner phenomenon), ultraviolet radiation exposure, and altered regional immune response (5, 2016). Understanding such atypical presentations is essential for early diagnosis and prevention of systemic complications. This study aims to present a detailed clinicopathological evaluation of unilateral Gottron's sign in ADM and discuss its diagnostic implications.

Legends to figures:

Figure 1: Violaceous striate plaques with adherent white fine scales over right hand.



Materials and Methods:

This study was conducted as a descriptive case-based clinicopathological investigation in the Department of Dermatology at a tertiary care teaching hospital. Ethical approval was obtained from the Institutional Ethics Committee, and written informed consent was secured from the patient for participation and publication. A comprehensive clinical evaluation was performed including detailed dermatological examination, systemic review, occupational exposure assessment, and documentation of lesion morphology and distribution. Differential diagnoses considered were hyperkeratotic eczema, allergic contact dermatitis, psoriasis, lichen planus, lupus erythematosus, and connective tissue disorders. Laboratory investigations included complete blood counts, liver and renal function tests, serum lactate dehydrogenase, creatine kinase (CK-NAC), erythrocyte sedimentation rate, and antinuclear antibody (ANA) testing. A myositis-specific antibody panel was performed including anti-Jo1, anti-Mi2, anti-SRP, anti-PL-7, anti-PL-12, anti-PM-Scl, anti-Ku, anti-TIF1- γ , anti-U1 RNP, and anti-MDA5 antibodies using enzyme-linked immunosorbent assay. Electromyography was conducted to evaluate subclinical myopathy. A malignancy workup including chest radiography, abdominal ultrasonography, and tumor markers was undertaken due to known malignancy association in dermatomyositis (14, 2017). High-resolution computed tomography (HRCT) of the chest was performed to assess interstitial lung involvement. A punch biopsy from the hand lesion was processed for histopathological examination using hematoxylin and eosin staining. Diagnostic criteria for ADM were applied based on persistent cutaneous findings without muscle weakness for more than six months (1, 1999). Treatment protocol included systemic corticosteroids at 0.5–1 mg/kg/day along with

hydroxychloroquine 300 mg daily and topical corticosteroids. Clinical response was evaluated at four-week intervals through photographic documentation and symptom assessment. Data were analyzed descriptively.

Results:

A 70-year-old male farmer presented with unilateral violaceous plaques over the right hand of 18 months duration. Additional lesions were noted over elbows and knee. No objective muscle weakness was detected, and serum CK levels were normal. ANA was negative. Myositis antibody panel showed isolated **anti-MDA5 positivity**. Histopathology revealed orthokeratotic hyperkeratosis, irregular acanthosis, interface dermatitis, and lymphomononuclear infiltrate without mucin deposition. Electromyography and malignancy screening were normal. HRCT chest demonstrated bilateral basal interstitial changes consistent with ILD. Based on clinical, serological, and radiological findings, a diagnosis of **Amyopathic Dermatomyositis with ILD** was established. The patient showed marked improvement after initiation of oral prednisolone and hydroxychloroquine within one month, with reduction in erythema and plaque thickness.

Discussion:

Unilateral Gottron's sign is an uncommon presentation of ADM and may delay diagnosis. Anti-MDA5 positivity strongly correlates with ILD and warrants aggressive evaluation (2, 2011). Environmental triggers such as ultraviolet exposure and trauma may explain localized presentation (5, 2016). Early recognition of atypical cutaneous signs is critical to prevent pulmonary complications.

Summary: This case highlights the diagnostic challenge posed by unilateral Gottron's sign in **amyopathic dermatomyositis**. Despite absence of muscle involvement, anti-MDA5 positivity and ILD risk necessitate thorough systemic evaluation. Prompt immunosuppressive therapy can significantly improve outcomes. Awareness of atypical unilateral manifestations is essential for dermatologists and rheumatologists.

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Figure 2A: 10x magnification hematoxylin and eosin (H&E) staining histopathology image consistent with interface dermatitis with moderate lymphomononuclear lichenoid infiltrate.

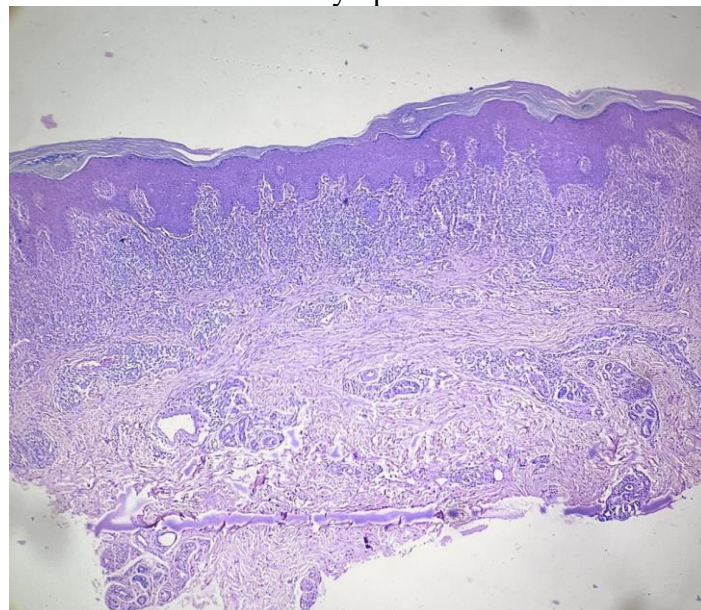
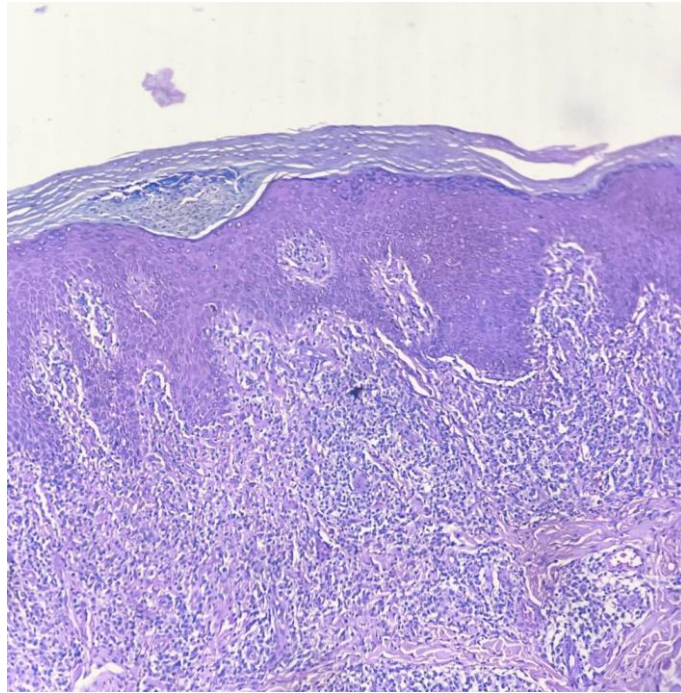


Figure 2B: 40x magnification hematoxylin and eosin (H&E) staining histopathology image displaying interface dermatitis with no mucin.



Tables if any