

Inverse Gottron's papule: Is it a critical sign of rapidly progressive interstitial lung disease?

W K B A Fernando¹, G K De Zoysa Rajakaruna², A S Danthararayana², J Akarawita³

Sri Lanka Journal of Dermatology, 2022-2023, 23: 36-37

Introduction

Dermatomyositis (DM) is an autoimmune connective tissue disorder associated with painful proximal muscle weakness, elevated muscle enzymes, inflammatory changes in muscle biopsy, inflammatory myositis in electromyogram and classic cutaneous manifestations. Cutaneous manifestations of DM include heliotrope rash, poikiloderma involving "v" area of neck, shawl sign, holster sign, gottron papules, gottron sign, reverse gottron sign, mechanic's hands, ragged cuticles, vasculitic manifestations, panniculitis and calcinosis cutis.

Inverse Gottron sign/papules (erythematous, flat-to-raised lesions involving the palmar aspect of the fingers) are a rare but specific cutaneous finding in DM. Pathogenesis is vasculopathy, explaining their association with digital ulceration and anti-MDA5 antibodies. It tends to be associated with rapidly progressive interstitial lung disease¹. It's important to identify this sign early to prevent morbidity and mortality in DM.

Case presentation

A 42-year-old painter, with a background history of proximal muscle weakness for 5 months duration presented with rapidly progressive shortness of breath and hoarseness for 2 weeks. He denied a history of Raynaud's, diffuse alopecia, skin tightening, or oral ulcers.

Examination revealed painful proximal muscle weakness with shoulder girdle power 4/5 MRC grading, and pelvic girdle 4/5 MRC grading. He was dyspneic with fine end-inspiratory crepts in both lungs. Cutaneous examination revealed ulcerated papules over interphalangeal joints of the palmar aspect of the right thumb, right index finger and left middle finger (Figure 1,2). Few retiform purpura were noted adjacent to ulcerated papular lesions of palms and both plantar aspects of feet (Figure 3). No other cutaneous manifestations of DM were identified.

Laryngeal examination of vocal cords was normal. UGIE revealed-oesophageal candidiasis



Figure 1.



Figure 2.



Figure 3.

¹Senior Registrar in Dermatology, ²Registrar in Dermatology, ³Consultant Dermatologist, National Hospital of Sri Lanka.

Investigations showed ANA- 1/100, ESR-90 mm/1st hour, CPK-345 IU/l and HRCT was suggestive of interstitial lung disease. Skin biopsy from a retiform lesion showed features of vasculitis. EMG was not suggestive of myopathy or myositis. DsDNA, U1RNP, C3, C4, Hepatitis B surface antigen, Hepatitis C antibody, C-ANCA, P-ANCA, retroviral screening, rheumatoid factor, and serum electrolytes were normal. Sputum AFB and gene expert, sputum pyogenic culture, COVID PCR were negative MDA5 antibodies were not done due to unaffordability.

He had rapidly progressive interstitial lung disease which required intubation and ventilator support. He was treated with IV methylprednisolone, IV immunoglobulins and IV cyclophosphamide. Despite the above treatment his interstitial lung disease worsened and the patient succumbed within 10 days of hospital admission.

Discussion

Inverse gottron sign is commonly associated with Anti-MDA5 associated DM^{2,3}. It is often seen in DM associated with interstitial lung disease⁴ and there are some reported cases with rapidly progressive interstitial lung disease with high mortality¹.

Conclusion

Dermatomyositis can have different presentations initially and, in some patients, can progress rapidly to develop complications. Identification of inverse gottron sign will aid clinicians in anticipating rapidly progressive interstitial lung disease.

References

1. Lee HJ, Feng A, Huan NC. Inverse Gottron's Sign in A Patient with Anti-MDA5 Associated Dermatomyositis with Rapidly Progressive Interstitial Lung Disease. *Am J Med Sci*. 2021; **362**(4): e35.
2. Ward Ian MD, Hiles Paul MD, Arroyo Ramon MD, Downs Walter MD, Bell David MD. Digital Pulp Ulcerations and Inverse Gottron Papules in Melanoma Differentiation-Associated Gene 5-Related Dermatomyositis. *Journal of Clinical Rheumatology (JCR)* 2016; **22**(5): 274-5.
3. Jain S, Sharma A. Inverse Gottron's sign in anti-MDA5 antibody-associated dermatomyositis. *Rheumatology* 2020; **59**(10): 3013.
4. Irie K, Matsumura N, Hoshi M, Yamamoto T. Inverse Gottron's papules in patients with dermatomyositis: an underrecognized but important sign for interstitial lung disease. *Int J Dermatol*. 2021; **60**(2): e62-e65.