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CASE REPORT

A Silent Muscle Story: Clinical Insights from Amyopathic Dermatomyositis

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ABSTRACT

Amyopathic dermatomyositis (ADM) is a rare subset of dermatomyositis accounting for ~20% of dermatomyositis cases. The prevalence is higher in females and peaks in middle age. Diagnosis relies on hallmark skin manifestations such as heliotrope rash and Gottron's papules, supported by skin biopsy, serologic markers (anti-MDA5, anti-TIF1γ), and imaging.

Unlike classic dermatomyositis, muscle enzymes and electromyography findings are typically normal. A 15-year-old female with no significant medical history presented with a one-month history of lowgrade fever and a progressive rash, along with oral ulcers for 15-days. The rash initially began as ill-defined macular to maculopapular lesions with fine scaling, predominantly involving sun-exposed areas, including the face (sparing the eyelids), ears, chin, anterior neck, and extensor surfaces of the arms, which extended later).

There is currently no specific randomized controlled trial that primarily involves ADM only. Further, treatment is often guided by the extent of cutaneous and systemic involvement. A better understanding may improve prognostication and therapeutic strategies, reducing morbidity and malignancy-associated mortality.

Keywords: Amyopathic dermatomyositis, Malignancy-associated mortality, Rash
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INTRODUCTION

Amyopathic dermatomyositis (ADM) is a rare subset of dermatomyositis characterized by classic cutaneous findings without clinically significant muscle involvement. It accounts for approximately, 20% of dermatomyositis cases. The prevalence is higher in females and peaks in middle age. Diagnosis relies on hallmark skin manifestations such as heliotrope rash and Gottron's papules, supported by skin biopsy, serologic markers (anti-MDA5, anti-TIF1γ), and imaging. Unlike classic dermatomyositis, muscle enzymes and electromyography findings are typically normal.[1]

Management includes corticosteroids, immunosuppressants (methotrexate, mycophenolate mofetil), and biologics (rituximab, JAK inhibitors). Cancer screening is crucial due to associated malignancy risk.[1,2]

CASE

A 15-year-old female with no significant medical history presented with a one-month history of low-grade fever and a progressive rash, along with oral ulcers for 15-days. The rash initially began as ill-defined macular to maculopapular lesions with fine scaling, predominantly involving sun-exposed areas, including the face (sparing the eyelids), ears, chin, anterior neck, and extensor surfaces of the arms (Figures 1, 2 & 3). Over time, it extended to the upper back and evolved into a more erythematous appearance. The patient reported notable photosensitivity, with exacerbation of the rash following sun exposure.

Systemic review revealed no muscle weakness or arthralgia. Laboratory investigations demonstrated a positive antinuclear antibody (ANA) with a homogeneous pattern and anti-Mi-2 antibody positivity.

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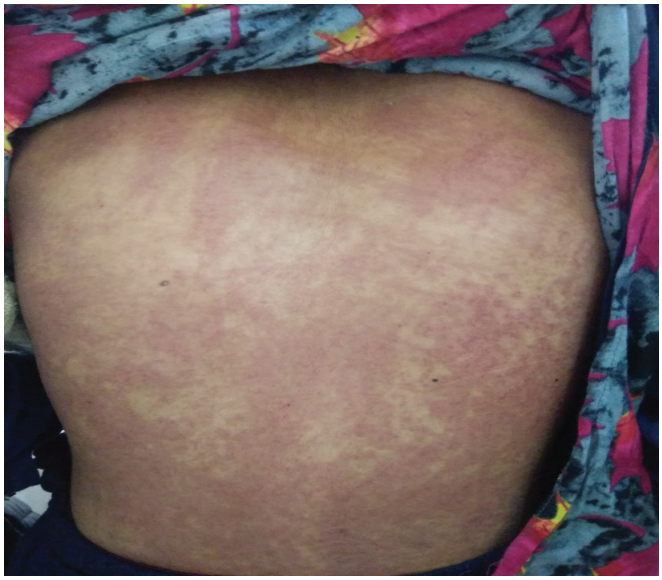

Figure 1: Extended rash on upper back.

Serum creatine phosphokinase (CPK) levels were within normal range (94 U/L), and electromyography was not indicative of myositis. These findings, coupled with the absence of clinical muscle involvement, supported a diagnosis of amyopathic dermatomyositis.

Further evaluation revealed proteinuria and bicytopenia. A direct Coombs test was positive, indicating immune-mediated cytopenia. Despite the overlap of features with SLE, the combination of characteristic cutaneous findings and anti-Mi-2 positivity favored ADM. Imaging studies, including a CT scan of the thorax, abdomen, and pelvis (CT TAP), were unremarkable,

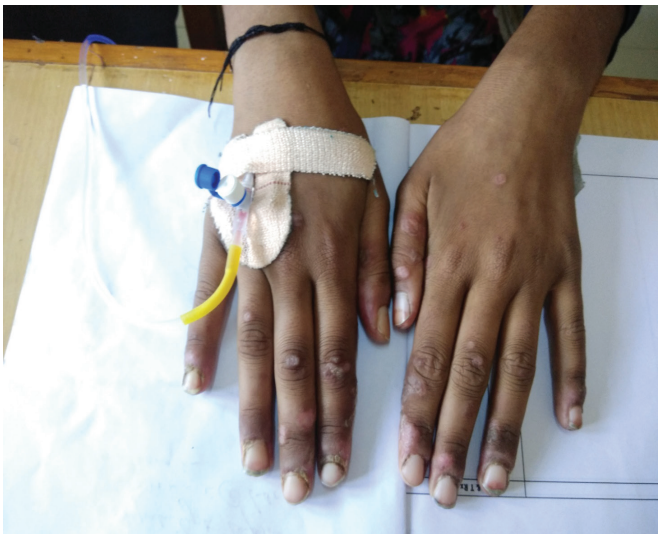

Figure 2: Rash on extensor surface on arms.

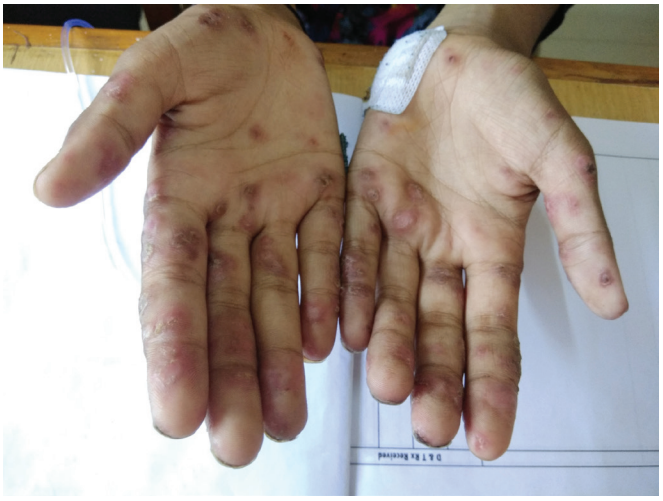

Figure 3: Rash on hands.

effectively ruling out malignancy. There was no evidence of interstitial lung disease on chest imaging.

DISCUSSION

Amyopathic dermatomyositis is an extremely rare, mostly idiopathic, multisystem connective tissue disease that is characterized by dermatologic lesions of classic dermatomyositis without myopathy or muscle weakness. Hallmark cutaneous manifestations include Gottron's sign, eyelid or periorbital heliotrope rash, and less commonly poikiloderma. There remains a substantial risk for development of interstitial lung disease or malignancy in diagnosed patients.

Amyopathic dermatomyositis (ADM), also commonly or more aptly referred to as clinically amyopathic dermatomyositis (CADM), is a distinct subtype of dermatomyositis (DM) known by the presence of pathognomonic cutaneous manifestations in the absence of clinically evident muscle weakness for a minimum duration of six months. ADM accounts for approximately 10–20% of DM cases and is increasingly recognized due to improved awareness and diagnostic techniques.[2,3] ADM falls within the idiopathic inflammatory myopathies (IIMs) spectrum. It is subclassified into 3 types usually:

1. Pure Amyopathic Dermatomyositis: Persistently present cutaneous disease without any evidence of muscle involvement, even on laboratory or imaging studies.
2. Hypo myopathic Dermatomyositis: Patients exhibit subtle or subclinical evidence of muscle inflammation (e.g., elevated muscle enzymes, abnormal MRI or electromyography) without overt weakness.

3. Evolving Dermatomyositis: Patients often present as amyopathic but subsequently develop classic myopathic features, meeting criteria for dermatomyositis with muscle involvement.[3,4]

Diagnostic Criteria

The diagnosis of ADM is primarily clinical and supported by laboratory and imaging findings. Criteria include:

1. The presence of hallmark cutaneous findings of DM (e.g., Gottron's papules, heliotrope rash, V-sign, or shawl sign).
2. Absence of clinical muscle weakness for at least six months following symptom onset.
3. Normal or mildly elevated serum muscle enzymes (creatine kinase, aldolase).
4. Normal muscle MRI, EMG, or biopsy, or findings insufficient to establish a diagnosis of inflammatory myopathy.
5. Detection of myositis-specific autoantibodies (e.g., anti-MDA5, anti-TIF1γ) may assist in classification and prognostication.

Clinical Manifestations

Characteristic cutaneous signs are central to the presentation of ADM and are often indistinguishable from classic DM. These manifestations include heliotrope rash, Gottron's papules, poikiloderma, shawl sign and photosensitive rashes. Despite the absence of muscle weakness, systemic involvement often complicates the picture with Interstitial lung disease (ILD) being a significant and potentially life-threatening complication (particularly in individuals with anti-MDA5 antibodies). Additional clinical features include arthritis, lipodystrophy, and gastrointestinal vasculopathy, especially seen and reported in paediatric populations.[5]

Management

There is currently no specific randomized controlled trial which primarily involves ADM only, and treatment is often guided by the extent of cutaneous and systemic involvement. First-line therapy dedicated for cutaneous disease includes antimalarial agents such as hydroxychloroquine which has been long used in rheumatological diseases. Topical modalities include corticosteroids and calcineurin inhibitors (e.g., tacrolimus), which are commonly used. In refractory or relatively severe cases, systemic immunosuppressive agents including antimetabolites like methotrexate, mycophenolate mofetil, and intravenous immunoglobulin (IVIG) have shown good results. In patients with ILD, particularly those with anti-MDA5 antibodies, apart from aggressive immunosuppression with corticosteroids, calcineurin inhibitors (e.g., tacrolimus or cyclosporine), rituximab has been pivotal for symptom control.[5,6]

RECENT ADVANCES

Autoantibody profiling has significantly improved the understanding and identification of ADM phenotypes. Anti-MDA5 is believed to be strongly associated with rapidly progressive ILD, while anti-TIF1γ is thought to correlate with severe cutaneous disease and confers an increased malignancy risk in adults. Radiological modalities like high-resolution chest CT and muscle specific MRI have proved valuable in assessing subtle and subclinical involvement. Therapies targeting interferon pathways and Janus kinase (JAK) inhibitors have shown promise in otherwise refractory disease.[6,7]

Prognosis

ADM usually has a favourable prognosis, particularly in those patients who do not have systemic involvement. Patients with anti-MDA5 positivity although face increased morbidity and mortality due to ILD. Nearly 20–30% of ADM cases may evolve into classic dermatomyositis lately. Vigilant monitoring is essential to detect any changes disease progression and to manage the complications promptly.[7]

CONCLUSION

Publishing on ADM is valuable due to its under-recognition, evolving diagnostic markers, and emerging therapies. Newer avenues include targeted biologics, interferon inhibition, and advanced imaging techniques. A better understanding may improve prognostication and therapeutic strategies, reducing morbidity and malignancy-associated mortality.

REFERENCES

1. Ledenko T, Sorić Hosman I, Ćorić M, Gagro A. Case Report: Simultaneously Developed Amyopathic Dermatomyositis and Autoimmune Sclerosing Cholangitis - a Coincidence or a Shared Immunopathogenesis? Front Immunol. 2022 Feb 23;13:825799. doi: 10.3389/fimmu.2022.825799. PMID: 35281002; PMCID: PMC8906471.
2. Javadi Parvaneh V, Yasaei M, Rahmani K, Nilipour Y. Juvenile Clinically Amyopathic Dermatomyositis:

A Case Report and Review of Literature. Iran J Child Neurol. 2019 Summer;13(3):113-120. PMID: 31327976; PMCID: PMC6586448.

3. Miyake Z, Ishii A, Okiyama N*, et al* Amyopathic dermatomyositis combined with peripheral neuropathy*BMJ Case Reports CP* 2020;**13:**e237250.
4. S. Plamondon, P.B. Dent ,Juvenile amyopathic dermatomyositis: Results of a case finding descriptive survey,J Rheumatol, 27 (8) (2000), pp. 2031-2034
5. R.D. Sontheimer,J Am Acad Dermatol, 46 (4) (2002), pp. 626-636
6. P.R. Chickermane, D. Mankad, R.P. Khubchandani,Disease patterns of juvenile dermatomyositis from western India ,Indian Pediatr, 50 (10) (2013), pp. 961-963
7. P. Gerami, H.W. Walling, J. Lewis, L. Doughty, R.D. Sontheimer ,A systematic review of juvenile-onset clinically amyopathic dermatomyositis ,Br J Dermatol, 157 (4) (2007), pp. 637-644