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Cardiac Involvement in Dermatomyositis: Successful Management using a Novel IVIg Preparation

Simon Waigand, Julia K. Winkler, Alexander H Enk*

Abstract

Dermatomyositis (DM) is a rare idiopathic inflammatory myopathy characterized by skin, muscle and various systemic involvement, with cardiac manifestations serving as critical prognostic factors. The management of cardiac involvement in DM remains challenging and subject to ongoing debate. We present a 72-year-old female with DM complicated by cardiac involvement, refractory to corticosteroids and mycophenolate mofetil. Treatment with a novel intravenous immunoglobulin preparation led to a marked reduction in cardiac enzyme levels, as well as improvements in strength, dyspnea, and dysphagia. This IVIg preparation differs regarding manufacturing process and shows a favorable safety profile. This case highlights the potential of IVIg in managing cardiac involvement in DM.

Keywords: Dermatomyositis; Cardiac involvement; High-dose intravenous immunoglobulins (IVIg); Yimmugo®

Introduction

Dermatomyositis (DM) is a subtype of idiopathic inflammatory myopathies (IIMs) characterized by an inflammatory response that primarily affects skeletal muscles and the skin [1]. Adult DM prevalence is estimated at 6-7 cases per 100,000 annually, with women affected about twice as often as men [2]. Pathognomonic signs include Gottron's papules or sign, heliotrope rash, the shawl or V-sign, nailfold abnormalities, and scaly dermatoses on the head and scalp. Besides skin manifestations, systemic involvement is common in DM, frequently affecting multiple organ systems, including muscles, joints, the gastrointestinal tract, lungs, and cardiovascular system [3].

Among the various systemic complications, cardiac involvement warrants special attention. With more sensitive, non-invasive detection methods, awareness of cardiac manifestations in polymyositis (PM) and DM has significantly increased, with an incidence ranging from 9% to 72% [4] Literature indicates that subclinical cardiac manifestations are considerably more common than clinical manifestations. Among these, rhythm disturbances are the most frequently observed subclinical issue. When clinical cardiac complications occur, congestive heart failure is the most common and can arise at any disease stage [5]. Cardiac involvement is now acknowledged as a crucial prognostic factor, with some experts considering it a primary determinant of mortality in PM and DM [6]. This underscores the importance of recognizing both subclinical and clinical cardiac involvement to improve patient management and outcomes.

Treatment of DM is tailored based on the severity of the disease and the extent of organ involvement. Immunosuppressive therapy with

Affiliation:

Department of Dermatology, University Hospital Heidelberg, Heidelberg, Germany

***Corresponding Author**

Alexander H Enk, Department of Dermatology, University Hospital Heidelberg, Heidelberg, Germany.

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corticosteroids (GC) remains the primary treatment option. GC are often combined with steroid-sparing agents, such as azathioprine or mycophenolate mofetil (MMF). For more severe cases, additional treatments may be considered including intravenous immunoglobulin (IVIg) [2]. However, management of cardiac involvement in DM remains complex and is subject to ongoing debate due to the variability in cardiac manifestations and therapeutic responses.

This case report discusses a patient with DM complicated by cardiac involvement, offering a detailed account of the clinical presentation and therapeutic approach, including treatment with a novel IVIg.

Case Report

A 72-year-old female patient presented to our dermatology department in June 2024 with a six- to eight-week history of fatigue, difficulty climbing stairs, and weakness in her arms, along with symptoms of facial swelling, redness, and increased hair loss.

Dermatological examination revealed periorbital edema with a heliotrope rash on the face, head, and décolleté, along with a shawl sign on the neck (Figure 1). A holster sign on both hips, Gottron's sign on the fingers and hands, as well as nailfold erythema and hyperkeratosis, were observed. The scalp exhibited erythematous scaling with noticeable hair thinning.

During the initial hospitalization, a biopsy and an MRI scan were conducted based on the suspected diagnosis of DM. Imaging revealed myositis affecting all examined striated muscles, with a focus on the left shoulder and upper arm muscles. The skin biopsy was also compatible with DM (Figure 2). Laboratory results showed an elevated ANA-titer (1:20000), ENA negative, with TIF1 gamma antibodies detected. A computed tomography (CT) scan of thorax and abdomen, along with a colonoscopy, esophagogastroduodenoscopy (OGD) and manometry, showed no abnormalities.

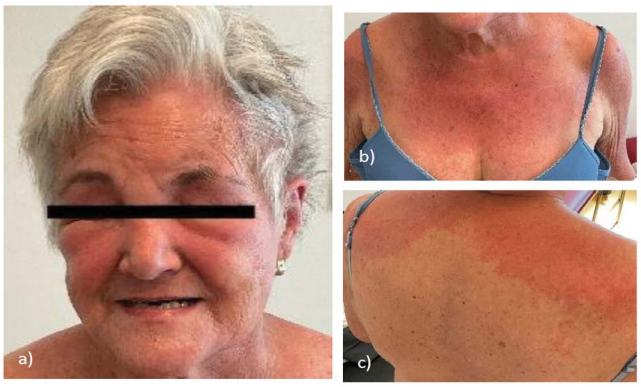

Figure 1: Clinical manifestation with periorbital edema with a heliotrope rash on the face (a), décolleté (b) and shawl sign on the neck (c).

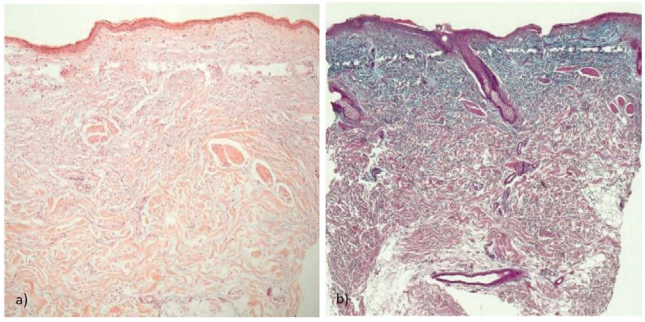

Figure 2: Histopathology reveals an atrophic epidermis with regular stratification, compact orthokeratosis and mild parakeratosis (a, HEx50). In the dermis pigment-laden macrophages and significant connective tissue elastosis are found. Massive mucin deposits in the dermis is characteristic of dermatomyositis (b, Alcian PAS).

Ten days post-discharge, the patient returned to our outpatient clinic, reporting significant deterioration. She experienced severe upper extremity weakness, could not dress independently, exertional dyspnea limiting mobility to a few steps and difficulty swallowing, which limited fluid intake.

Oral prednisolone therapy 80 mg once daily (1 mg/kg bodyweight) was initiated. Clinical examination confirmed severe dyspnea (NYHA class IV). Elevated troponin T (TnT, 99.5 pg/mL) and D-dimer levels (>35.20 mg/L) were detected. The electrocardiogram (ECG) showed low voltage in the precordial leads and discrete S-wave persistence up to V6. CT angiography revealed a peripheral pulmonary embolism without right heart involvement, and anticoagulation with rivaroxaban 15 mg twice daily was initiated. No deep vein thrombosis could be detected.

The initially elevated TnT level, attributed to pulmonary embolism, persisted despite anticoagulant therapy. Cardiac MRI with contrast showed a normal size, mildly hypertrophic left ventricle with a good systolic function (LV-EF 59%). Localized inferolateral basal edema was observed, accompanied by increased T1 relaxation time and late gadolinium enhancement, with no focal edema on T2-weighted images. Cardiac involvement related to DM was assumed, and mycophenolate mofetil (MMF) 1000 mg three times daily was added.

To rule out functional heart impairment, regular echocardiographic exams were performed, showing no cardiac dysfunction. A long-term ECG revealed frequent supraventricular extrasystoles, some repetitive and occurring as atrial runs, along with isolated polymorphic ventricular extrasystoles (Lown class IIIa).

Under continuous therapy, the patient demonstrated an objective increase in muscular strength and subjective improvement in dyspnea and dysphagia, but cardiac enzyme levels stayed elevated. Consequently, we initiated IVIg

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therapy with Yimmugo® at a dosage of 2g/kg bodyweight over four days, repeated every for weeks. GC was reduced to 20 mg/day over four months, and MMF lowered to 2 g/day three months ago due to lymphopenia, with lymphocyte levels starting to normalize. No side effects from IVIg therapy were observed and cardiac enzyme levels rapidly reduced, with the patient reporting improved strength, dyspnea, and dysphagia.

Discussion

High-dose IVIg is well-established for treating autoimmune and systemic inflammatory diseases. Regarding DM, numerous case reports, small case series, and a double-blind, placebo-controlled study support its efficacy [7-9]. IVIg is particularly preferred for severe or refractory DM when GC and conventional immunosuppressants are ineffective and may be considered first-line in life-threatening cases [10].

Based on the results of a clinical trial in 2021, IVIg have received approval in the US and EU for DM treatment. Despite its high safety profile, adverse events may occur, ranging from mild side effects such as headache, fever and nausea to rare but serious complications such as thromboembolic and cerebrovascular events [11]. There are currently no defined treatment recommendations for cardiac involvement in IIM, due to the relative rarity of the condition. Current recommendations are based solely on case reports and case series.

In a study of 15 PM/DM patients with myocarditis, corticosteroid monotherapy resulted in remission of skeletal muscle inflammation, but myocarditis persisted in 83% of cases [12]. Supporting this view, Danieli et al. [8] presented a case series of seven PM and DM patients, including one DM patient with cardiac arrhythmia and dyspnea, who did not respond to prior immunosuppressive treatments. A combined regimen of MMF and IVIg led to clinical improvement in all cases [8]. Yoshimatsu et al. [13] further demonstrated the efficacy of high-dose IVIg in treating cardiomyopathy in severe DM, where cardiomyopathy developed despite intensive immunosuppressive therapy [13]. This suggests that IVIg may play a pivotal role in the management of cardiac complications in DM. In contrast, He et al. [14] reported a DM case with initial clinical improvement on GC therapy but persistent cardiomyopathy unresponsive to high-dose steroids and IVIg. The patient exhibited worsening cardiac markers and reduced urine output, leading to acute heart failure and death [14].

In our case the patient exhibited elevated cardiac enzyme levels, persistent dyspnea and dysphagia, despite treatment with high-dose steroid therapy and MMF. We initiated treatment with a novel IVIg preparation whose manufacturing process differs from other IVIg preparations and involves the reduction of shear stress by vibromixing and the removal of the complement system activator properdin. Thrombogenic factors are eliminated through caprylic acid and low pH treatment. In vitro assays, including clotting tests, chromogenic assays, non-activated partial thromboplastin time, and the TGA method, confirmed the absence of thrombogenic factors and measurable coagulation activity in this preparation [15].

Given the patient's history of thromboembolic events and the increased risk of venous thromboembolic complications during IVIg treatment, Yimmugo® was selected and administered over four days due to the patient's prior pulmonary embolism. A rapid decrease in persistently elevated cardiac enzymes was observed, along with an objective increase in strength, reduced dyspnea, and a subjective improvement in dysphagia. No adverse events were reported, suggesting that the new IVIg may be an optimal choice for long-term treatment of DM.

Notably, this is the first documented case of cardiac involvement alongside positive TIF-1-gamma detection, highlighting the potential of Yimmugo® in managing severe DM. Given the association of TIF-1-gamma with paraneoplastic dermatomyositis, we thoroughly investigated a possible paraneoplastic origin but found no evidence of malignancy. However, ongoing surveillance will be essential to exclude emerging neoplastic processes.

Prospective comparative studies and further real-world data are essential to confirm the efficacy, safety, and tolerability of this new IVIg preparation in treating this rare, potentially life-threatening disorder and to support informed long-term management decisions.

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Conflict of interest

Alexander H. Enk received advisory board honoraria, consultancy fees and support for the documentation of patient cases from Biotest AG. Julia K. Winkler received travel expenses and honoraria from Biotest AG. Simon Waigand declares to have no competing interests.

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