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Case Report

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Optimal Treatment Strategies for Chronic Cutaneous Vasculitis with Concurrent IGA Monoclonal Gammopathy: A Case Report

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Abstract

This case report investigates the complex presentation and management of a 61-year-old female patient with bilateral leg ulcerations, ultimately diagnosed with cutaneous vasculitis and concurrent IgA monoclonal gammopathy, classified as monoclonal gammopathy of clinical significance (MGCS). MGCS refers to nonmalignant monoclonal gammopathies that cause significant disease, often manifesting with symptoms affecting the nerves, kidneys, and skin. Our patient exhibited chronic ulcerations and a challenging clinical course over four years, requiring extensive investigation and multiple therapeutic interventions. Initial treatments, including high-dose corticosteroids and rituximab, provided minimal to limited improvement. However, significant clinical response was observed following the administration of hydroxychloroquine and dapsone. This case underscores the importance of recognizing MGCS in patients presenting with monoclonal gammopathy and unexplained symptoms, the critical role of skin biopsies in diagnosis, and the necessity of a multidisciplinary approach in managing these rare and complex conditions. The report highlights the complexities of diagnosing and treating cutaneous vasculitis associated with IgA monoclonal gammopathy and discusses the potential benefits of targeted therapies such as hydroxychloroquine and dapsone in achieving optimal outcomes.

1. Introduction

MGCS represents a spectrum of conditions where the monoclonal protein causes morbidity despite the absence of malignancy. This case report describes a 61-year-old female patient who presented with bilateral leg ulcerations persisting over the course of four years. She was ultimately diagnosed with cutaneous vasculitis in the context of an IgA monoclonal gammopathy, classified as MGCS. This case underscores the importance of considering MGCS in patients with unexplained symptoms and monoclonal gammopathy. The patient's clinical course was refractory to initial treatments. High-dose corticosteroids, commonly used to manage vasculitis, and rituximab, an anti-CD20 monoclonal antibody that depletes B-cells, provided only minimal improvement. This lack of response necessitated a reevaluation of the therapeutic strategy. A significant clinical response was observed following the administration of hydroxychloroquine and dapsone, highlighting their potential role in managing similar cases. Hydroxychloroquine, an antimalarial with immunomodulatory properties, inhibits antigen presentation and reduces the production of pro-inflammatory cytokines [1]. Dapsone, an antibiotic with anti-inflammatory effects, suppresses neutrophil chemotaxis and inhibits myeloperoxidase, mechanisms relevant to the pathology of vasculitis [2]. The patient's response to these treatments underscores their potential efficacy in refractory cases of cutaneous vasculitis associated with monoclonal gammopathy. The diagnostic process in this case relied heavily on histopathological examination through skin biopsies, revealing vasculitis involving small and medium-sized vessels with occasional thrombi. This finding, coupled with protein electrophoresis showing a monoclonal IgA kappa confirmed by immunofixation, was crucial in establishing the diagnosis of MGCS. Management of cutaneous vasculitis in the context of MGCS requires a multidisciplinary approach, integrating the expertise of dermatologists, rheumatologists, and hematologists. This case report illustrates the complexities involved in diagnosing and treating such conditions and underscores the importance of a comprehensive and collaborative approach to achieve optimal outcomes. The case highlights the necessity of considering MGCS in the differential diagnosis of chronic ulcerations and the potential benefits of targeted therapies such as hydroxychloroquine and dapsone in achieving disease control.

2. Case Presentation

A 61-year-old female presented with complaints of bilateral leg ulcerations persisting for over four years. The patient is overweight with a significant history of hypertension and pre-diabetes. There is no significant family history of similar conditions. She was initially seen by dermatology and rheumatology for her chronic leg ulcers, suspected to be due to vasculitis. Her serologic tests were

negative for ANCA, rheumatoid factor (RF), and Hepatitis B and C. However, antinuclear antibody (ANA) was positive with a titer of 1:40 with nucleolar pattern. Initial Diagnosis and Treatment: A punch biopsy, 3 years prior, revealed vasculitis involving small and medium-sized vessels with occasional thrombi.

PROTEIN electrophoresis showed a monoclonal IgA kappa confirmed by immunofixation. Initial treatments included high-dose corticosteroids up to 60mg daily, which provided no significant improvement, and rituximab (RTX) administered a year after the initial diagnosis. Despite these treatments, the patient continued to experience significant ulcerations, especially on the lower extremities. Subsequent Evaluations and Adjustments: Due to the patient's persistent cutaneous vasculitis in the context of an IgA monoclonal gammopathy, she was diagnosed with MGCS. The patient's CD19 counts were monitored, showing low levels post-RTX. She was prescribed colchicine, and later, hydroxychloroquine (HCQ) and dapsone were introduced, which marked a significant turning point in her management. A marked reduction of ulcer size was noted on both lower extremities Follow-Up and Current Status: The patient continued with HCQ 200 mg twice daily, dapsone 75 mg daily, and clobetasol to the wound edges, showing significant improvement. The leg ulcerations showed marked improvement with the current regimen (Figures 1-2).

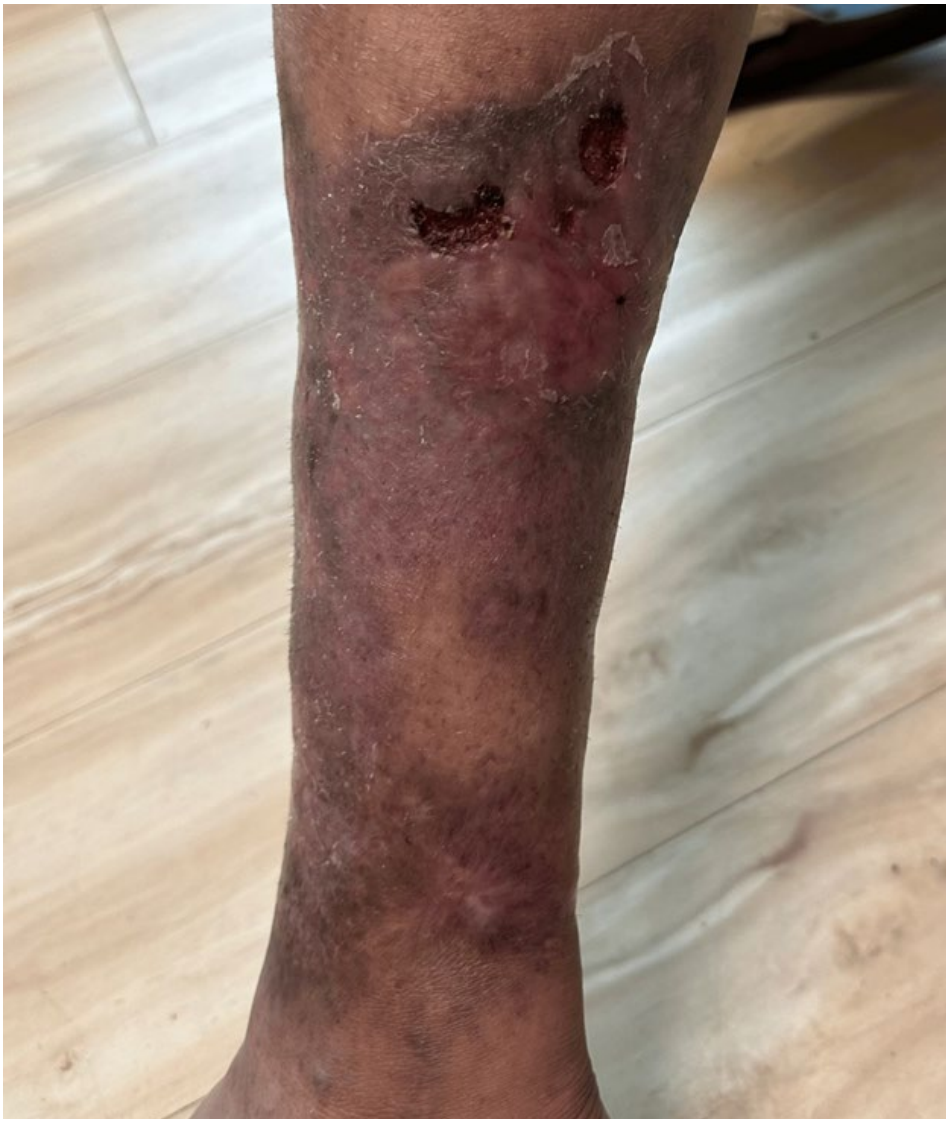

Figure 1: Left lower Extremity Ulceration: *Extensive dusky brownish purple plaque on the left lower extremity with focal partial thickness punched out ulcerations along the posterior and lateral aspects. The ulcerations exhibit minimal granulation tissue and cribriform scarring, indicative of chronic vasculitic changes.*

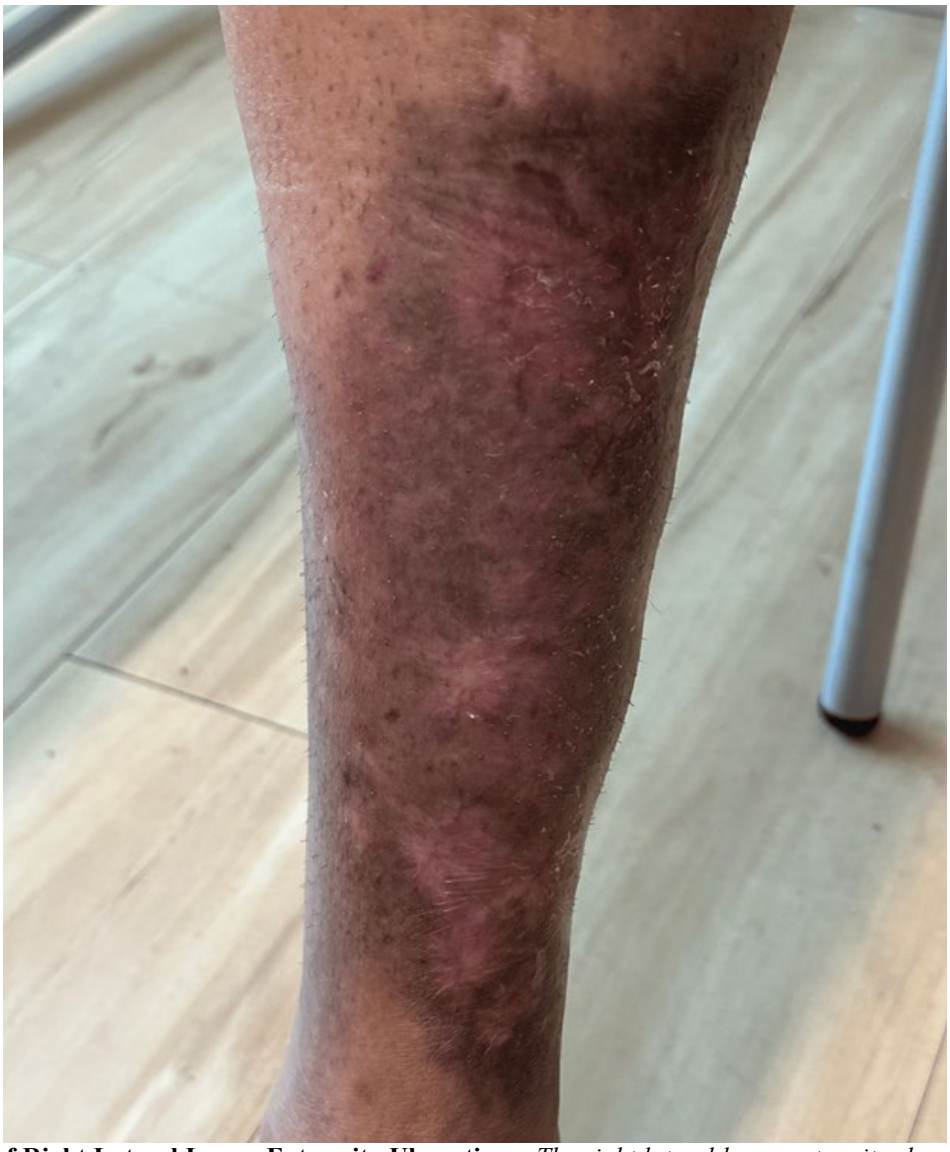

Figure 2: Improvement of Right Lateral Lower Extremity Ulcerations: *The right lateral lower extremity showing dusky brownish purple plaque with significant improvement following treatment with dapsone and hydroxychloroquine. The plaque appears less extensive with reduced inflammation and improved healing compared to initial presentation.*

2.1 Medications and Management Plan

The patient's medications included aspirin, clobetasol, gabapentin, dapsone, HCQ, losartan, and vitamin D3. Her treatment plan focused on continuing the current regimen of dapsone and HCQ, with regular follow-ups for monitoring and adjustments as needed (Table 1). Annual ophthalmologic exams were recommended due to the long-term use of HCQ.

Medication	Dosage	Frequency/Delivery
Clobetasol	0.05%	Topical ointment, twice a day
Dapsone	75 mg	25 mg in the morning, 50 mg in the evening
Hydroxychloroquine (HCQ)	200 mg	Orally twice a day

Table 1: Current Medication Regimen for MGCS-related Chronic Vasculitis

3. Discussion

The management of cutaneous vasculitis, particularly in the context of monoclonal gammopathy, presents unique challenges due to its complex pathophysiology and varied clinical manifestations. In this case, the patient's prolonged and refractory bilateral leg ulcerations were ultimately attributed to IgA monoclonal gammopathy. This diagnosis falls under the category of Monoclonal Gammopathy of Clinical Significance (MGCS), which encompasses nonmalignant monoclonal gammopathies causing significant clinical disease. The initial diagnostic process was hindered by negative serologic tests for ANCA, RF, and Hepatitis B and C, which are commonly associated with vasculitic syndromes [3]. The positive ANA at a titer of 1:40 with a nucleolar pattern suggested an autoimmune component, yet did not point to a specific diagnosis. A crucial step in confirming the diagnosis was the skin biopsy, which revealed vasculitis involving small and medium-sized blood vessels, thus guiding the subsequent management plan. The patient's clinical course exemplifies the importance of considering MGCS in patients with unexplained ulcerations and monoclonal gammopathy. Her treatment history underscores the necessity of individualized therapy in achieving disease control. Initial high-dose corticosteroids and rituximab (RTX) provided limited improvement. RTX, while effective in depleting B-cells and reducing immunoglobulin production, did not fully address the underlying pathology in this case. The persistent ulcerations, particularly in the left leg, prompted the consideration of alternative therapies. The introduction of hydroxychloroquine (HCQ) and dapsone marked a significant turning point in the patient's treatment. HCQ, an antimalarial with immunomodulatory properties, has been shown to be effective in treating various autoimmune conditions, including cutaneous lupus and rheumatoid arthritis. Its efficacy in this case may be attributed to its ability to inhibit antigen presentation and reduce the production of pro-inflammatory cytokines [4]. Dapsone, an antibiotic with anti-inflammatory properties, is commonly used in the treatment of dermatitis herpetiformis and other IgA-related dermatopathologies has shown benefit in various neutrophilic dermatoses. Its mechanism, primarily through inhibition of myeloperoxidase and suppression of neutrophil chemotaxis, likely contributed to the observed clinical improvement [5]. The positive therapeutic response to HCQ and dapsone highlights the importance of considering these agents in the treatment of refractory cutaneous vasculitis, particularly in cases associated with monoclonal gammopathy. This case stresses the necessity of a multidisciplinary approach in managing complex dermatologic conditions associated with systemic diseases. Collaboration between dermatology, rheumatology, and hematology specialists was crucial in achieving an accurate diagnosis and effective treatment plan. Additionally, the importance of regular monitoring and follow-up cannot be overstated, particularly in patients receiving immunosuppressive therapy and those at risk of hematologic complications. Future directions for research focusing on the pathophysiology of MGCS and its dermatologic manifestations, as well as the development of standardized treatment protocols would give better insight on the nature and treatment of similar cases. Additionally, larger studies are needed to evaluate the long-term efficacy and safety of therapies such as hydroxychloroquine and dapsone in managing cutaneous vasculitis associated with monoclonal gammopathy

4. Conclusions

This case highlights the diagnostic and therapeutic challenges in managing cutaneous vasculitis associated with IgA monoclonal gammopathy. The limited response to conventional therapies underscores the need for a high index of suspicion for MGCS in patients with unexplained ulcerations and monoclonal gammopathy. The significant clinical improvement observed with hydroxychloroquine and dapsone suggests these agents as viable therapeutic options for achieving disease control in similar cases. Key takeaways from this case include the critical role of skin biopsies in establishing a definitive diagnosis, the importance of considering MGCS in the differential diagnosis of chronic ulcerations, and the potential benefits of targeted immunomodulatory therapies. The multidisciplinary approach, involving dermatology, rheumatology, and hematology specialists, was essential in optimizing patient outcomes. This case report aims to contribute to the growing body of knowledge on the dermatologic manifestation of MGCS and its clinical management, providing valuable insights for clinicians encountering similar complex cases. The successful management of the patient's condition highlights the potential for individualized, targeted therapy in achieving optimal patient outcomes in the face of challenging and rare dermatologic diseases.

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