Case Report

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Pyoderma Gangrenosum-like Ulcers Associated Granulomatosis with Polyangiitis

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Running Title PG-like Ulcers in GPA: Case Report

ABSTRACT

Granulomatosis with polyangiitis is an ANCA-positive vasculitis primarily affecting small to medium-sized vessels, typically involving the lungs and kidneys. This article presents a remarkable case of a patient with an unusual pyoderma gangrenosum-like ulcer on the face and granulomatosis with polyangiitis. A 28-year-old man, previously diagnosed with granulomatosis with polyangiitis, presented at a dermatology clinic with a 6-month-old ulcer in the right preauricular region following a traumatic incident. A spindle biopsy revealed suppurative granulomatous dermatitis with secondary vascular damage, while cultures showed no signs of infection. The patient was subsequently treated with rituximab, leading to a positive clinical response. This case underscores the challenge of diagnosing this rare manifestation of granulomatosis with polyangiitis and suggests that rituximab could be a valuable treatment option.

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Introduction

Granulomatosis with polyangiitis (GPA) is an ANCA (Anti-Neutrophil Cytoplasmic Antibody)-positive small-medium vessel vasculitis [1]. It is an uncommon disease with an incidence of about 12.8 cases per million person-years in working-age adults in the United States of America [2]. The physiopathology remains unknown, but infectious, toxic, and pharmacologic triggers in genetically predisposed individuals may influence disease onset [3]. Almost any organ can be affected, but the respiratory tract and kidneys are the most commonly damaged [1]. Skin involvement occurs in up to 34% of patients, either as an initial manifestation or more frequently during the disease [4]. Among the most frequent cutaneous manifestations of GPA are palpable purpura (38.1%), mucocutaneous ulcers (28.58%), and subcutaneous nodules (14.21%) [5].

We present an uncommon case of pyoderma gangrenosum (PG)-like ulcer on the face of a patient diagnosed with GPA.

Case Presentation

A 28-year-old man attended a dermatology clinic for a six-month history of an ulcer in the right preauricular area, which had progressed in size and was painful. He claimed to have suffered a lesion after a trauma occurring on pavement, for

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which he received treatment with trimethoprim/ sulfamethoxazole without improvement. He had a history of granulomatosis with polyangiitis since he was 14 years of age and was under treatment with azathioprine and prednisone with poor adherence(Figure1).

Given the suspicion of GPA activity vs. pyoderma gangrenosum vs. fungal/mycobacterial infection, it was decided to perform a spindle biopsy of the margin, where ulcerative and suppurative granulomatous dermatitis with secondary vascular damage was reported. Periodic acid-Schiff (PAS) and Ziehl-Neelsen stains were negative. A culture of the lesion was performed without isolates of microorganisms. It was decided to start treatment with 2 doses of rituximab (1 g each), resulting in epithelization of the ulcer six months after the first dose (Figure 2).

Discussion

PG is a painful, necrolytic, ulcerative neutrophilic dermatosis that is usually associated with autoimmune and inflammatory conditions [6]. The legs are affected most of the time, while the face is an uncommon location [7]. PG-like ulcers are a rare manifestation of GPA, with a frequency between 1.1%-9.5% in patients with GPA and skin disease [5]. Facial localization is more consistent with a PG-like ulcer associated with GPA [8], but the antecedent of trauma made us consider differential diagnoses such as PG or mycobacterial infection.

Differentiating PG and PG-like ulceration can be challenging. Histopathology alone is not enough to distinguish them because some PG cases with

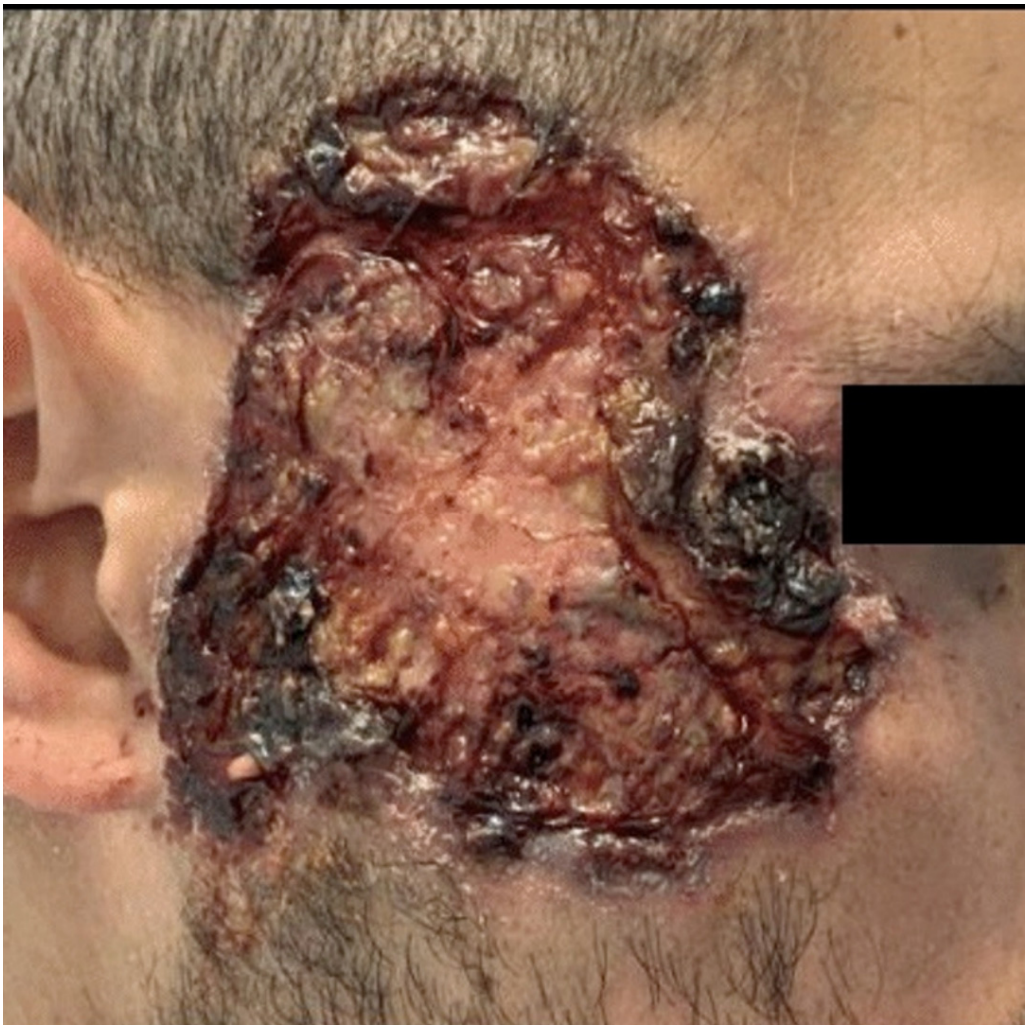

Fig. 1. Pyoderma gangrenosum-like ulcers associated granulomatosis with polyangiits**.** A) Dermatosis located on the head, affecting the preauricular, cheek, and temporal region on the right side; characterized by a 5 x 7 cm ulcer with irregular edges, covered almost entirely by fibrin and bloody crusts; the circumference of the lesion with a purple coloration.

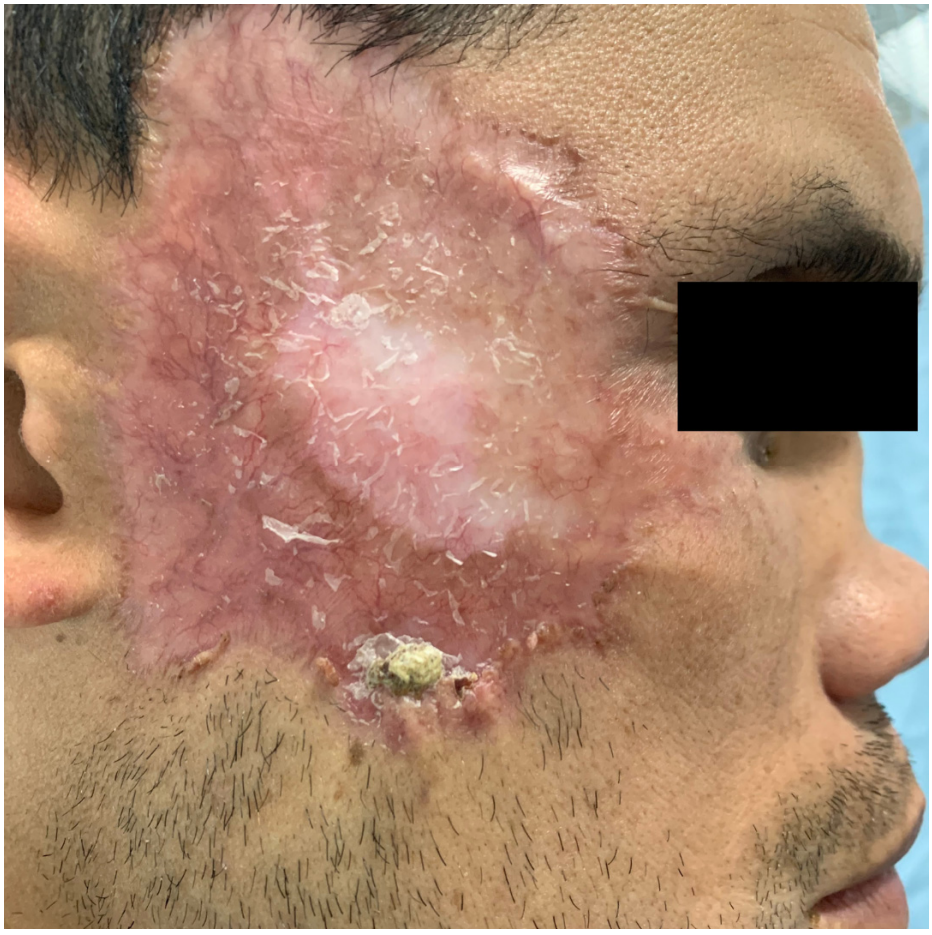

Fig. 2. Pyoderma gangrenosum-like ulcers associated granulomatosis with polyangiits**.** B) After treatment with rituximab, a normotrophic, erythematous scar in the right temporal region that causes alopecia on the hairline, lower eyelid retraction, and a scab on the lower end of the scar.

granulomatous inflammation have been described [6]. Genovesse et al. propose some terms for the difference between them, including a less raised, erythematous-violaceous edge in PG-like ulcers; different histology between both; c-ANCA positivity in PG-like ulcers associated with GPA; and systemic involvement that rarely occurs in PG [9].

In our case, the uncommon localization of the ulcer and histology consistent with vasculitis and granulomatous dermatitis in a poorly treated GPA patient were the criteria to classify it as a PG-like ulcer. Cultures must be obtained to rule out infections in Latin America, particularly for mycobacterial infections that can mimic PG [10].

There are no current guidelines for the treatment of PG-like ulcers associated with GPA. In the literature, there are some case reports with a favorable response to rituximab [11-13]. After six months, our patient received two doses of 1 g of rituximab with an adequate response. More studies are needed to validate its indication.

PG-like ulcers associated with GPA present a diagnostic challenge, requiring careful consideration of clinical presentation, histopathology, and other differentiating factors. We emphasize the importance of recognizing these rare skin manifestations of GPA. Rituximab shows potential as a therapeutic intervention, but further research and evidence are needed to standardize treatment guidelines for PG-like ulcers.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of Interests

The authors have no conflict of interest to declare.

Declarations

The patient authorized the use of their photo for publication purposes.

Data is available upon request to the corresponding author.

The study is in accordance with the ethical standards laid down in an appropriate version of the WMA Declaration of Helsinki-Ethical Principles for Medical Research Involving Human Subject.

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