

# Pyoderma Gangrenosum-like ulceration as a presenting feature of pediatric Granulomatosis with Polyangiitis

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## Case Report

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## Abstract

**Background:** Granulomatosis with polyangiitis (GPA) is an anti-neutrophilic cytoplasmic antibody-associated vasculitis affecting small to medium-sized vessels and involves most commonly the kidneys and the respiratory tract. Skin involvement can be seen in up to 50% of children with GPA and is the initial presenting symptom in 7.7%. Pyoderma gangrenosum (PG)-like ulcers are rarely described as a skin manifestation in GPA and very few cases have been reported previously in children.

**Case presentation:** We describe 3 new pediatric cases of GPA with PG-like ulcerations. The mean age at first symptom was 15 years. Two patients had PG-like ulceration as their initial presentation; additional symptoms eventually led to the diagnosis of GPA 2-24 months later. In 1 case, proteinase 3 (PR3) was negative when first tested, but converted to positive when systemic symptoms emerged, in the other 2 cases PR3 was positive at presentation. All 3 patients had prominent facial lesions. None of the patients responded to treatment with antibiotics or medications commonly used to manage PG, including corticosteroids and cyclosporine. All patients had excellent responses to rituximab. An electronic database literature review was performed and 4 previously reported cases were identified. We assessed the clinical characteristics, serology, and response to treatment of 4 previously reported and newly diagnosed cases.

**Conclusion:** PG-like ulceration is a rare presentation of pediatric GPA which may precede classic systemic GPA symptoms. The predominance of facial ulcer, granulomatous and neutrophilic inflammation on skin biopsy and lack of response to PG treatments are characteristic of GPA-associated PG-like ulcers. Our review suggests that treatment with rituximab may be needed to improve the skin lesions. Recognizing that PG-like ulcerations can occur in pediatric GPA may result in timely diagnosis, appropriate treatment and improved prognosis.

## Background

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a form of anti-neutrophil cytoplasmic antibodies (ANCA) associated vasculitis. GPA affects small to medium-sized vessels and involves most commonly the kidneys and respiratory tract. Skin involvement has been reported in 36% – 50% of children with GPA and is the initial presenting symptom in 7.7%. Palpable purpura and acneiform papules are most commonly seen. Rarely, patients with GPA develop pyoderma gangrenosum (PG)-like ulcerations. Very few cases of this dermatologic manifestation have been reported among GPA patients and, more specifically, in pediatric GPA patients. Early recognition of this rare entity and appropriate diagnostic work-up for GPA are essential in order to reduce morbidity and mortality by facilitating disease-specific treatment.

We describe three new pediatric cases initially diagnosed with PG who were later diagnosed with childhood-onset GPA. We also review four previously reported cases in the literature.

## Case 1

A 16-year old, previously healthy male initially presented to his primary care physician with acne-like lesions on his face, chest, upper back and arms. He was treated with topical antibiotic and retinoids in addition to oral antibiotics for 2 months; however, the lesions continued to worsen and developed into ulcerations (Fig. 1a). Skin biopsies were performed by dermatology and initially interpreted as perifollicular inflammation with giant cell reaction and abscess formation consistent with acne. Treatment with oral prednisone, 80 mg daily for suspected PG was initiated with some improvement. A few weeks later, he developed systemic symptoms of cough and sinusitis. The rheumatology team was consulted and the diagnosis of GPA was made based on sinus, pulmonary, and renal involvement in addition to elevated inflammatory markers and a positive PR3. Upon further review, the initial skin biopsy was interpreted as palisaded neutrophilic and granulomatous inflammation with multinucleated giant cells and erythrocyte extravasation (Fig. 2). He was treated with intravenous (IV) methylprednisolone, plasmapheresis, 2 doses of IV cyclophosphamide and a course of rituximab. Following this treatment, the skin ulcers improved significantly (Fig. 1b), as did his systemic disease.

## Case 2

A 15-year old female presented with left arm and facial ulcerations (Fig. 3a). An extensive infectious and immunologic work-up was negative, including ANCA serology. Chest x-ray and colonoscopy were normal. She received more than one year of various topical

and systemic treatments including antibiotics, mycophenolate mofetil, prednisone (maximum 40 mg per day), colchicine and cyclosporine for presumed PG without significant improvement in the lesions. One year after the initial presentation, she developed chronic sinusitis resistant to antibiotic therapy, with destruction of the medial wall of the maxillary sinus. Rheumatology evaluation revealed elevated inflammatory markers and a positive PR3 for the first time. Sinus biopsy showed necrotizing granulomatous inflammation. Skin biopsy was interpreted as granulomatous and neutrophilic inflammation within the deep dermis (Fig. 4). She was treated with pulse methylprednisolone and a course of rituximab, followed by maintenance therapy with rituximab every six months. Since commencement of this treatment regimen, no new skin lesions have appeared. The facial ulcers healed with poor cosmetic result (Fig. 3b).

### Case 3

A 14-year old female presented with fever and a solitary, painful, nodule located above the left scapula. The fever persisted and she developed a dry cough, hemoptysis and chest pain. Inflammatory markers were increased (ESR 130 mm/hour, CRP 195 mg/L). A homogenous infiltration predominantly of the left lung parenchyma, granulomas, and a stenosis of the left main bronchus were found on chest CT. Her poor clinical condition did not allow for a lung biopsy. Empiric antibiotic treatment for suspected pneumonia was ineffective. She developed additional skin lesions on her left forehead, left cheek, back of the neck, back and in the pubic area that all progressed into extremely painful, large (up to 7 cm), deep, sharp-edged ulcers with an erythematous and fibrinous base (Fig. 5a, 5b). A skin biopsy, reviewed by our institution, displayed granulomatous inflammation comprised predominantly of histiocytes, neutrophils, few plasma-cells and multinucleated giant cells (Fig. 6). Given these pathology findings in addition to lung and sinus involvement and a positive PR3, the diagnosis of GPA was given. Treatment with intravenous methylprednisolone pulses, oral prednisone and monthly IV cyclophosphamide (cumulative dose 5.5 g) did not induce remission of her cutaneous or systemic features. Treatment with rituximab dramatically improved her PG-like lesions and clinical condition, however, she developed a 50% subglottic stenosis and required local treatment with corticosteroid injections and dilation. The currently 20-year old patient's GPA is in remission on azathioprine and IVIG replacement for hypogammaglobulinemia resulting from persistent B-cell depletion.

### Results Of Literature Review

A search through two electronic databases (PubMed and Medline), using the following keywords: GPA, PG, PG-like lesions/ulceration, Wegener's granulomatosis, pediatric, identified 4 pediatric cases of GPA with PG-like lesion. Clinical characteristics of these cases in addition to our three new cases reported above are presented in Table 1. The mean age at first symptom was 15 years with M:F ratio of 5:2. All patients had a positive PR3 (except patient 5 for which ANCA status was not available). In 2 cases, PR3 was negative when first tested, but converted to positive on repeat evaluation when systemic symptoms emerged. Five children had PG-like ulceration as their initial presentation (patients 1, 2, 5, 6, 7), Additional symptoms eventually led to GPA diagnosis two to twenty-four months later. The ulcerative lesions initially manifested as purpura, nodule or acne in 4 patients. All patients had mainly facial lesions. Additional locations were chest and back. Four of seven patients had renal involvement and 6/7 had upper respiratory tract involvement.

Table 1  
Demographic and clinical characteristics of patients with GPA and PG-like lesions

Age (years)	Sex	Description of skin lesion	Anatomical location	Histology	Serology	Organ involved	Time to diagnosis(months)	Treatment	Reference
1 16	M	Acne  ulceration	Face, chest, back	Palisaded neutrophilic and granulomatous inflammation with multinucleated giant cells and erythrocyte extravasation	PR3 ab positive	Sinus, pulmonary, renal	4	ABX, CS, CYC, PLEX, rituximab	
2 15	F	Ulceration	Face and arm	Granulomatous and neutrophilic inflammation within the deep dermis	Negative  positive PR3 ab	Sinusitis	24	ABX, CS, cyclosporine, MMF, rituximab	
3 14	F	Painful ulceration	Face, back, pubic	Granulomatous inflammation comprised predominantly of histiocytes, neutrophils, few plasma-cells and multinucleated giant cells	PR3 ab positive	Sinus, subglottic stenosis, pulmonary	2	CS, CYC, rituximab, azathioprine	
4 18	M	Ulceration	Post auricular	Granulomatous, perifollicular, and supportive inflammation	PR3 ab positive	URT	N/A	CS, CYC, TMP-SMX	#6 Confere et al
5 16	M	Purpura, pustule, ulcer	Peri-auricular, shin	Perivascular infiltrates of lymphocytes, histiocytes, plasma cells and neutrophils	N/A	URT  Renal  Pulmonary	18	ABX, CS, CYC	#7 Chyu et al
6 12	M	Nodule, ulcer	Shin, leg, chest	Intense neutrophilic dermal infiltrate with giant cells and small vessel vasculitis	Negative  positive PR3 ab	Renal	N/A	ABX, CS, cyclosporine, CYC, rituximab	#8 Kass et al
7 Mid-teens	M	Acne  ulceration	Face and neck	Geographic necrosis	PR3 ab positive	URT  Renal	> 2 months	ABX,  CS, Infiximab  rituximab	#9 Moen et al

Abbreviation: M: male; F: female; PR3 ab: proteinase 3 antibody; N/A: not available; ABX: antibiotic; CS: corticosteroids; CYC: cyclophosphamide; MMF: mycophenolate mofetil; PLEX: plasma exchange; TMP-SMX: trimethoprim/sulfamethoxazole; URT: upper respiratory tract.

## Discussion

Although cutaneous manifestations are common in GPA, PG-like lesions are rarely seen. Frumholtz et al summarized the cutaneous manifestations of 1553 adult patients with ANCA-associated vasculitis. While 36.7% (273/743) of patients with GPA had cutaneous involvement, PG was observed in only 1.1% of patients. Very few cases of GPA with PG-like lesions have been reported previously, especially in children. We hereby present 3 new cases of pediatric GPA with PG-like lesions, and review these together with the 4 previous pediatric cases found in the literature.

PG is a rare, rapidly progressive, sterile ulcerating neutrophilic dermatosis which usually presents as painful pustules or nodules. It may evolve into necrotic plaques with raised edges and ultimately into deep violaceous ulcers. PG lesions most frequently affect the lower extremities and tend to be multifocal and recurrent. Young adults aged 20–50 are most commonly affected, while pediatric cases account approximately for only 4% of cases. The diagnosis is based on the typical clinical appearance and accompanying histological findings which can be variable and often non-specific. These include a predominantly neutrophilic inflammatory infiltrate with evidence of necrosis and hemorrhage at the base of the lesion. Approximately 50–70% of PG cases are

associated with systemic conditions, particularly autoimmune and auto-inflammatory diseases and malignancies . Amongst the pediatric population, inflammatory bowel disease, juvenile idiopathic arthritis (JIA), Takayasu arteritis and immunodeficiency conditions have been associated with PG .

Weenig et al reviewed medical charts of 240 patients who received the diagnosis of PG and found that 49 patients (20%) had a different final diagnosis including vascular occlusive disease, vasculitis, cancer and infection. The mean lag time to the final diagnosis was 10 months. Ulcerative skin involvement mimicking PG is often termed “pyoderma gangrenosum-like ulceration”. There may be a lack of widely used diagnostic criteria, but PG-like ulceration and PG are two separate entities with different histopathologic findings . The histopathologic features of PG-like ulcers in GPA include palisaded neutrophilic and granulomatous dermatitis, necrotizing vasculitis and basophilic collagen degeneration. These histopathologic features are not typically seen in classic PG. The term “PG-like ulceration” is preferred in the setting of systemic findings that are consistent with GPA.

The clinical characteristics of the 7 pediatric patients summarized in Table 1 were slightly different from known demography of pediatric GPA patients. The median age at diagnosis was 15 years, which is older than the median pediatric age at GPA diagnosis (12.5–14.5 years) found in previous studies. There was a male predominance 5:2, while previous studies show a female predominance in pediatric GPA patients .

Interestingly, all 7 patients had facial involvement. In contrast, the most common anatomical location of “classic” PG is the lower extremities, found in 50% of adults with primary PG . Systematic review of 170 pediatric cases with PG found 46% of patients had disseminated skin disease, 30% had disease limited to the lower extremities and only 3.9% had facial involvement <sup>11</sup>. The atypical anatomical distribution with predilection for facial lesions should increase suspicion of an underlying disease such as GPA.

The common treatment for PG includes topical therapy in milder disease (corticosteroids, dapsone, tacrolimus etc.) and systemic treatment with oral corticosteroids as first line and cyclosporine as a second line treatment for more resistant disease<sup>18</sup> . Our three cases presented above had only minimal improvement on antibiotic and multiple immunosuppressive medications, including cyclophosphamide. However, treatment with rituximab led to resolution of active skin disease in all cases. A similar response to rituximab was reported in adults with GPA and PG-like ulceration <sup>22 23</sup> . Significant scarring remained as a result of the depth of the ulcerating lesions. The initial treatment given to the patients described above not only had a minimal effect on the PG-like lesions, but also may have masked and delayed the systemic presentation of GPA, leading to delayed referral for rheumatology evaluation. Delay in diagnosis and initiation of appropriate treatment can lead to more extensive skin disease and scarring, as well as increased morbidity and mortality related to the underlying GPA diagnosis.

## Conclusion

PG-like ulceration is a unique and rare presentation of pediatric GPA which sometimes can precede the classic systemic GPA symptoms. The predominance of facial ulceration, granulomatous and neutrophilic inflammation on skin biopsy and lack of response to PG treatment are much more common in GPA-associated skin ulceration than in PG. Treatment with rituximab resulted in improvement of both the PG-like ulcerations and other GPA disease manifestations in most of the patients in this review. Recognizing that PG-like lesions can occur in pediatric GPA may result in a timely diagnosis and appropriate treatment, and thus can help improve prognosis.

## Abbreviations

ANCA - anti-neutrophil cytoplasmic antibodies

CT - *computed tomography*

GPA - Granulomatosis with polyangiitis

IV – intravenous

JIA – Juvenile Idiopathic Arthritis

PG – Pyoderma Gangrenosum

PR3 – Proteinase 3

## Declarations

**Ethics approval** - IRB does not require the submission of a protocol in order to publish the case report.

<https://biologicalsciences.uchicago.edu/sites/biologicalsciences/files/2019-06/policiesandproceduresmanual.pdf>

**Consent forms** – were signed by the patient / patient's guardian.

**Availability of data and materials** - *The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.*

**Competing interests** – *The authors declare that they have no competing interests.*

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### Authors' contributions:

RSO – collected and analyzed the data and co-wrote the article.

LWW – analyzed the data, co-wrote and revised the article.

OO, AR contributed the dermatology and derm-pathology aspect and revised the article

LH, RT, TD – contributes cases, revised the article

LP – contributed pathology aspect and insight, revised the manuscript.

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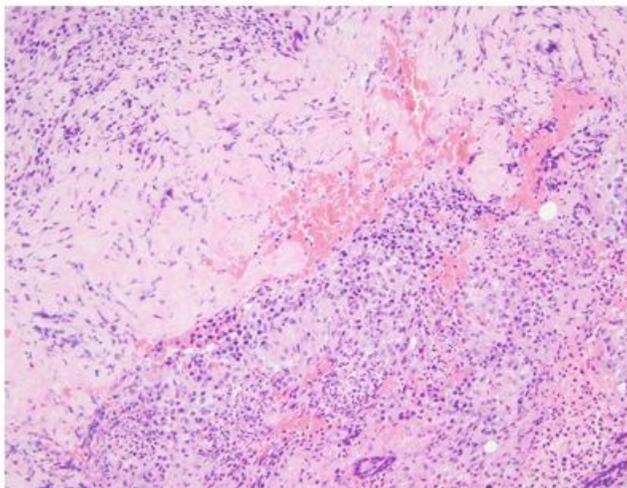
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## Figures



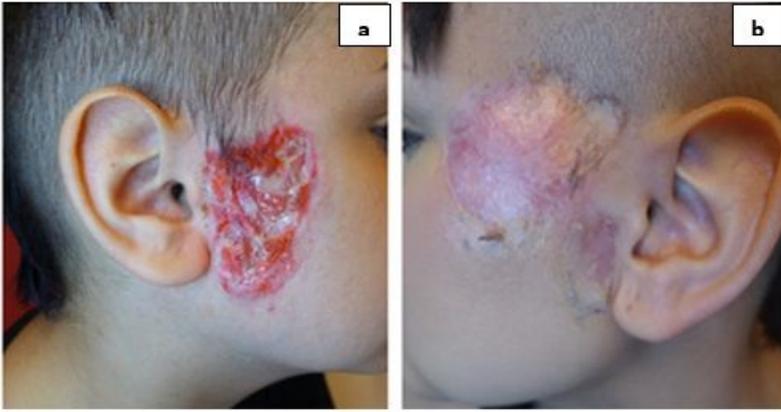
**Figure 1**

Pre (a) and post (b) treatment forehead lesions images of a 16-year-old male (patient 1)



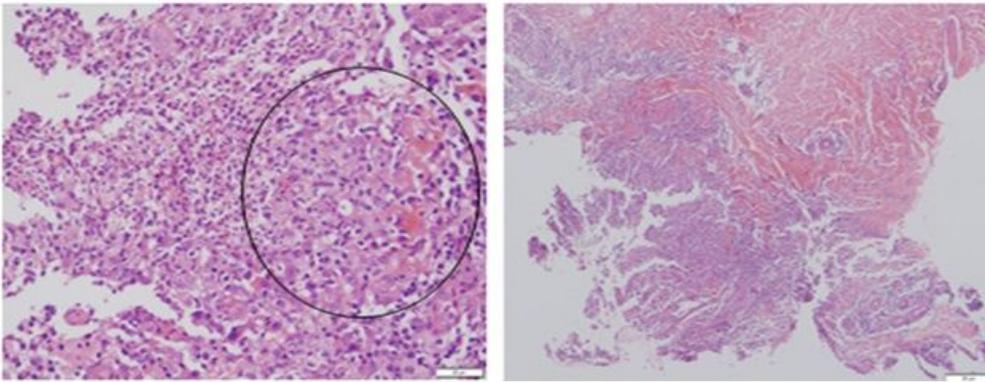
**Figure 2**

Case 1 Skin biopsy. High power microscopic examination demonstrating palisaded neutrophilic and granulomatous inflammation with multinucleated giant cells and erythrocyte extravasation



**Figure 3**

Pre (a) and post treatment images of (b) facial lesions of a 15 year old female (patient 2) showing poor cosmetic outcome.



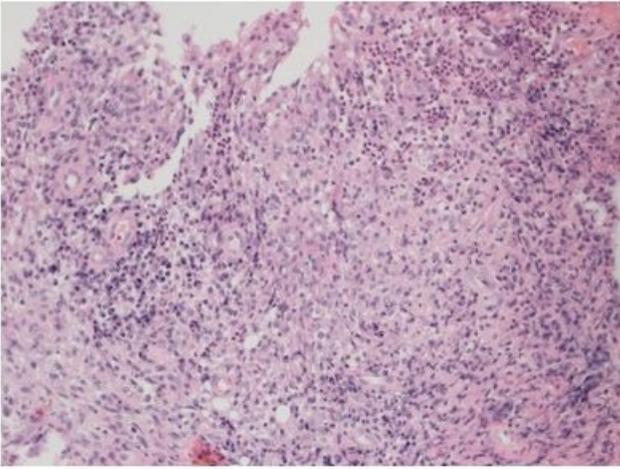
**Figure 4**

patient 2 skin biopsy. High and low power microscopic examination demonstrating granulomatous and neutrophilic inflammation within the deep dermis



**Figure 5**

Facial (a) and pubic (b) ulcerative lesions of a 14-year old female (patient 3).



**Figure 6**

Facial (a) and pubic (b) ulcerative lesions of a 14-year old female (patient 3).