Pyoderma gangrenosum faciale in a patient with Crohn’s disease

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Key words: anti-Saccharomyces cerevisiae antibody; aspiration; Crohn’s disease; cutaneous; extracutaneous; face; inflammatory bowel disease; oropharyngeal dysphagia; perianal; pyoderma gangrenosum; skin.

INTRODUCTION

Pyoderma gangrenosum (PG) is commonly co-
morbid with an apparent or occult underlying con-
dition.1 PG normally occurs on the lower extremities, and facial manifestations are rare.1,2 We present a
case of severe ulcerative PG faciale, complicated by
cranial nerve dysfunction possibly leading to chem-
ical aspiration and death.

CASE REPORT

A woman in her 40s with a history of recurrent
cutaneous and oropharyngeal abscesses and aph-
thaes of indeterminate etiology, treated with antimi-
crobials and prednisone, 20 mg/d, was admitted to
the hospital for a recurrent maxillofacial abscess. A 2-
cm, purulent, subperiosteal fluid collection within
the left maxillary vestibule was noted on examina-
tion and computerized tomography (CT) maxillofa-
cial protocol. Empiric treatment was initiated for a
presumed infectious left vestibular maxillary abscess
and osteomyelitis. The patient underwent intraoral
incision and drainage and received empiric intrave-
nous vancomycin and piperacillin-tazobactam.
Abscess cultures grew normal oropharyngeal flora.
The patient was discharged on a 6-week course of
ertapenem and doxycycline with dose-reduced
prednisone, 10 mg/d.

A week later, the patient was re-admitted for
rapidly progressing edema, pain, and purulent left
cheek drainage, despite compliance with antibiotics.
Physical examination found an edematous, ecchy-
motic plaque with central eschar extending from the
left medial canthus to the inferior buccal cheek. After
several days, the eschar sloughed off spontaneously,
revealing an exudative ulceration to the fat layer and
connecting into the maxillary oral cavity (Fig 1, A
and B). Ulcer edge histopathology found a dense,
diffuse, mixed inflammatory infiltrate, and scattered
vascular channels in the mid dermis showed marked
fibrinoid degeneration and focal fibrin thrombi. No
vasculitis was seen, and tissue cultures remained
without growth. An undermined perianal ulcer was
also noted (Fig 2).

Laboratory findings were consistent with iron
deficiency anemia. Further investigations failed to
find paraproteinemia, anti-CCP antibodies, rheuma-
toid factor, anti–neutrophil-associated cytoplasmic
titers, or anti–proteinase 3 and antimeyeloperoxidase
antibodies. The patient reported a recent normal
colonoscopy at an outside hospital.

Ulcerative PG faciale was suspected. Iron defi-
cency without peptic ulcer disease symptoms or
menorrhagia made occult inflammatory bowel
disease (IBD) a compelling comorbid association.
Antibiotics were discontinued, and the patient
was started systemic corticosteroids, colchicine,
dapsone, and cyclosporine. The patient’s wound
showed marked improvement over her 24-day hos-
pitalization, and she was discharged home.

Further outpatient workup included elevated
anti-Saccharomyces cerevisiae IgG at 50.9 units and
IgA at 41.0 units (reference range, 0.0-24.9 units).
Colonoscopy and pill camera endoscopy while on

Abbreviations used:
CT: computerized tomography
CD: Crohn’s disease
IBD: inflammatory bowel disease
PG: pyoderma gangrenosum

JAAD Case Reports 2020;6:766-8.
2352-5126
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https://doi.org/10.1016/j.jdcr.2020.06.026
immunosuppressive therapy were nondiagnostic. Infliximab and azathioprine were necessary to effect ulcer re-epithelialization (Fig 3). The patient experienced a PG flare requiring intensified pharmacotherapy and a suspected aspiration pneumonia from benzodiazepine overdose. Modified barium swallow found aspiration past true vocal cords when swallowing thin liquids consistent with moderate oropharyngeal dysfunction.

The patient was later hospitalized with a productive cough progressing to worsening hoarseness and hemoptysis. A CT scan of the chest found multifocal centrilobular nodular airspace opacities predominantly in the right upper lobe, concerning for multifocal pneumonia, chemical pneumonitis, or extracutaneous PG. She was given antimicrobial treatment, and azathioprine and infliximab were held during hospitalization. After discharge, her facial PG and constitutional symptoms worsened. The patient chose to resume her immunomodulatory therapies with close monitoring rather than be re-admitted to rule out infection. Her condition stabilized and improved. However, after a trip to New York City in February 2020, the patient and her parents suffered from new-onset fevers, diarrhea, and cough. Although her parents recovered, the patient complained of severe shortness of breath and died at home presumably of coronavirus disease 2019 (COVID-19).

**DISCUSSION**

PG is difficult to diagnose because of variable presentation, overlap with other conditions, and absence of pathognomonic laboratory and histologic findings. Our patient met 1 major criterion and 4 minor criteria for ulcerative PG (biopsy showing neutrophilic infiltrate; infection exclusion; peripheral erythema, undermined border, and tenderness at ulceration site; cribriform scar at healed ulcer sites; decreasing ulcer size within 1 month of taking immunosuppressants), yielding a specificity of 90% for PG.

Facial and perianal PG are exceedingly rare. Only 4.5% of 356 patients or 7.8% of 103 patients present with facial involvement. We found 2 other cases of perianal PG after conducting a PubMed review using the terms “Pyoderma gangrenosum AND perianal.”

Because PG is associated with an underlying disease in 50% to 78% of patients, most commonly IBD, investigation into a comorbid condition is warranted. Endoscopic and histologic confirmation of IBD may be difficult in patients presenting with acute PG due to intolerance of or inadequate bowel preparation, timing of immunosuppressive therapies that blunt diagnostic sensitivity of gastrointestinal

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**Fig 1.** PG faciale. A. Initial presentation shows an edematous, bruise-like plaque with central eschar and purulent borders. B. Several days after presentation, the eschar spontaneously separated and revealed an ulcer to the fat layer with connection into the oral cavity.

**Fig 2.** Perianal PG. Near circumferential undermined anal ulcer.

**Fig 3.** Healing PG faciale. Pink, depressed, re-epithelialized scar is without eschar. An oral cavity opening is still present.
histology, and gastroenterologist willingness to pursue invasive tests in the absence of symptoms.

Our patient’s history of aphthous ulcers, iron deficiency anemia, and perianal disease commonly present in patients with CD. A meta-analysis of more than 1300 patients with and without suspected CD indicate a pooled anti-Saccharomyces cerevisiae antibody sensitivity of 55% (95% confidence interval, 52-59) and pooled specificity of 93% (95% confidence interval, 91-95) for CD. Diagnosis of underlying IBD may afford insurance coverage of US Food and Drug Administration-approved immunomodulatory therapies that may not otherwise be supported for PG alone.

PG most commonly presents extracutaneously in the lungs and typically affects 30- to 60-year-old women. Symptoms are variable, and most commonly include fever and cough, whereas other signs of respiratory involvement, such as dyspnea, crackles, and stridor, are reported less frequently. Chest radiograph and CT scan can reveal noncavitating or cavitating infiltrates or pulmonary nodules. Given the rarity of pulmonary PG and its variable presentation, no clinical guidelines exist for diagnosis and treatment. A differential diagnosis of granulomatosis with polyangiitis, infection, and malignancy is typically considered. Our patient’s delayed swallowing trigger and incomplete laryngohyoid elevation from PG-induced cranial nerve dysfunction increase her aspiration risk. Therefore, respiratory symptoms and pulmonary infiltrates, as well as reasonable rule out of granulomatosis with polyangiitis, suggest a differential diagnosis of infectious pneumonia, chemical pneumonitis, or pulmonary PG. Misdiagnosis can prove deadly, as reducing immunosuppressive treatment may cause a pulmonary PG flare, whereas continuing immunosuppressants allows progression of aspiration pneumonia or chemical pneumonitis.

Here we present an unusual case of facial and perianal PG in a patient with occult underlying CD complicated by pulmonary disease of uncertain etiology. Patients with PG faciale should be assessed for dysphagia, and pulmonary disease must be adequately investigated to determine best management strategies.

REFERENCES
1. Binus AM, Qureshi AA, Li VW, Winterfield LS. Pyoderma gangrenosum: a retrospective review of patient characteristics, comorbidities and therapy in 103 patients. Br J Dermatol. 2011;165(6):1244-1250.
2. Ashchyan HJ, Butler DC, Nelson CA, et al. The Association of age with clinical presentation and comorbidities of pyoderma gangrenosum. JAMA Dermatol. 2018;154(4):409-413.
3. Maverakis E, Ma C, Shinkai K, et al. Diagnostic criteria of ulcerative pyoderma gangrenosum: a delphi consensus of international experts. JAMA Dermatol. 2018;154(4):461-466.
4. Agnew JL, Strombom PD, Fong CF, Kelly TJ, Martz JE. Perianal pyoderma gangrenosum after excision and fulguration of anal condyloma acuminatum. Int J Surg Case Rep. 2015;17:51-54.
5. Alvite-Canosa M, Monjero-Ares I, Alonso-Fernández L, et al. Pyoderma gangrenosum with extensive perianal involvement. Rev Esp Enferm Dig. 2012;104(2):92-93.
6. Ahronowitz I, Harp J, Shinkai K. Etiology and management of pyoderma gangrenosum: a comprehensive review. Am J Clin Dermatol. 2012;13(3):191-211.
7. Larsen S, Bendtzen K, Nielsen OH. Extraintestinal manifestations of inflammatory bowel disease: epidemiology, diagnosis, and management. Ann Med. 2010;42(2):97-114.
8. Reese GE, Constantinides VA, Simillis C, et al. Diagnostic precision of anti-Saccharomyces cerevisiae antibodies and perinuclear antineutrophil cytoplasmic antibodies in inflammatory bowel disease. Am J Gastroenterol. 2006;101(10):2410-2422.
9. Borda LJ, Wong LL, Marzano AV, Ortega-Loayza AG. Extracutaneous involvement of pyoderma gangrenosum. Arch Dermatol Res. 2019;311(6):425-434.
10. Gupta AS, Greiling TM, Ortega-Loayza AG. A Systematic review of pyoderma gangrenosum with pulmonary involvement: clinical presentation, diagnosis and management. J Eur Acad Dermatol Venereol. 2018;32(7):e295-e297.