Recurrent Vegetative Pyoderma Gangrenosum of the Face: A Case Report

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ABSTRACT

A 46-year-old male presented with an erythematous papule progressing into a vegetative plaque on the right cheek that resolved with cribriform scarring. Eight months after, a similar looking erythematous papule appeared on his left cheek. This papule rapidly progressed into a vegetative plaque within a week, and was associated with a pain score of 7 out of 10. Histopathology of the second lesion revealed suppurative dermatitis with diffuse dense infiltrates composed mostly of neutrophils. Cultures revealed few colonies of *Enterobacter cloacae* which was inconclusive. Pathergy test was negative. High dose systemic corticosteroids were started, with an observed rapid reduction of pain, inflammation, and ultimately resolution of the lesion with formation of cribriform scarring, confirming a case of vegetative pyoderma gangrenosum. It is important to note that not all inflamed and purulent lesions are infectious-neutrophilic dermatoses should always be considered.

Keywords: pyoderma gangrenosum, face, glucocorticoids, case report

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INTRODUCTION

Pyoderma gangrenosum (PG) is a rare, non-infectious neutrophilic dermatosis that presents as a painful ulcerating plaque.[1-3] There are currently five subtypes which include classic ulcerative, bullous, pustular, vegetative, and peristomal.[1] It initially starts as a small papule or pustule, eventually coalescing into an ulcerating plaque.[4] Pyoderma gangrenosum rarely affects the face as it usually involves the lower extremities.[2,4] Pyoderma gangrenosum is considered a diagnosis of exclusion.[2] We present a case of recurrent vegetative pyoderma gangrenosum occurring on the contralateral side of the face eight months after its initially presented and resolved on the right cheek.

CASE DESCRIPTION

A 46-year-old male with no known comorbidities presented with an erythematous papule on the right cheek that progressed into a vegetative plaque with subsequent purulent discharge (Figure 1). There was no reported weight loss, pallor, joint pain, abdominal pain, or bowel movement changes. While he had no prior history of pulmonary tuberculosis or treatment, his father was treated for pulmonary tuberculosis. Otherwise, he had an unremarkable personal and family history.

The plaque then rapidly increased in size, encompassing the entire right cheek within seven days. There was no history of trauma or manipulation. Patient was initially admitted in a different tertiary institution, where computed tomography scan of the facial bones and orbits was done, revealing

bilateral maxillary sinusitis and unremarkable orbits. Wedge biopsy was done showing suppurative dermatitis. Tissue samples revealed no microorganisms seen after 48 hours on gram stain and culture, and acid-fast bacilli was not seen and detected on tissue PCR. Due to reported abdominal pain, esophagogastroduodenoscopy (EGD) was done and revealed gastric ulcer and duodenal ulcer Forrest III, erosive gastroduodenitis, and a positive test for *H. pylori*. This points less likely to inflammatory bowel disease as EGD would reveal strictures, fistulas, and erythema.[5] He was managed as a case of necrotizing soft tissue infection, probably necrotizing cellulitis and was started on Piperacillin-Tazobactam and Clindamycin. The wound was cleaned with gauze soaked in sodium chloride solution three times a day, in addition to mupirocin ointment and copper sulfate, zinc oxide, sucralfate cream being applied on the right cheek. The patient was discharged with resolution of the plaque on the right cheek with cribriform scarring.

Eight months after the initial presentation of the lesion, the patient noted a similar-looking papule, but this time on the left cheek. There was associated pain with a numeric rating scale score 7 out of 10, but no history of trauma. Within one week, it evolved into a solitary, well-defined, irregularly shaped, vegetative, erythematous plaque with overlying pustules, purulent non-foul-smelling discharge and a pinkish to violaceous border. The plaque measured 8.2 cm x 6.5 cm x 0.5 cm and occupied the left cheek from the nasolabial fold to the infraorbital region (Figure 2A). No other ulcerative lesions on other locations. There was also cribriform-like scarring on the right cheek extending to the left side of the nose, noted from the previous admission. On further physical examination, there was presence of a solitary painful oral ulcer in the mucosa of the lower lip. Pathergy test was done and revealed to be negative with no erythema, papule, or pustule formation at the site of intradermal injection of isotonic saline on the forearm after 24 hours.

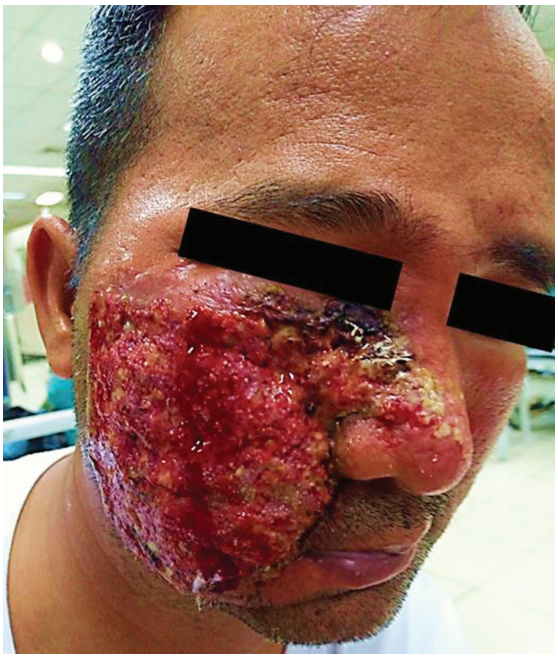

Figure 1. ***First incident of vegetative plaque on the right cheek.*** Solitary vegetative plaque with subsequent purulent discharge on the entire right cheek.

A wedge biopsy was taken from the border of the plaque on the left cheek, which revealed sections of diffuse dense infiltrates composed mostly of neutrophils, consistent with suppurative dermatitis (Figure 3). Wound discharge was negative for acid-fast bacilli and showed no growth on gram stain and culture. Tissue samples were negative for acid-fast bacilli, bacterial gram stain TB culture, and fungal culture. Tissue bacterial culture had few colonies of *Enterobacter cloacae* which was considered inconclusive as a possible contaminant as *E. cloacae* is a commensal microflora of the intestinal tracts of humans and animal.[6] Laboratory testing was available in the tertiary institution and covered during the admission. With the following results, the diagnosis of vegetative pyoderma gangrenosum on the face and herpetic stomatitis for the oral mucosa were considered. Other condsiderations for the face include facial cellulitis, lupus vulgaris, (PAPA) pyogenic arthritis, PG, and acne syndrome and Behcet disease.

For associated diseases of PG, complete blood count initially had slightly decreased hemoglobin at 114 g/L which prompted peripheral blood smear which revealed normocytic, normochromic, marked poikilocytosis which was not consistent with hematologic malignancy but anemia from acute inflammation. HbA1c was normal. Auto-antibody screening, rheumatoid factor, antiphospholipid screening were not done due to the absence of systemic symptoms. The previous EGD was also not consistent with inflammatory bowel disease. Creatinine was initially elevated at 121 umol/L which then slowly decreased due to possible acute kidney injury secondary to infection with secondary anemia, not in uremia. Before initiating anti-inflammatory medication, chest x-ray was done and revealed to be essentially normal as well. Herpes 1 and 2 IgG were both reactive. HIV rapid screen revealed to be non-reactive.

Initiation of methylprednisolone pulse therapy (MPPT) 1 gram for three days resulted in a dramatic decrease in size, thickness, and purulent discharge (Figure 2B). Therapy was eventually shifted to intravenous hydrocortisone 100 mg every 8 hours for two days. As the patient was stable and the plaque continued to decrease in size, thickness and induration, he was discharged. Medication was shifted to oral prednisone 60 mg per day for his home medications. The patient's steroids were then decreased by 10 mg every week until tapered off completely. After 30 days of corticosteroid treatment, there was more than a 50% decrease in ulcer size (Figure 2D) with evidence of cribriform scarring (Figure 2C and E).

Patient was advised that the treatment will need a prolonged course of oral prednisone which would require

monitoring of resolution of the lesion. Systemic symptoms for pyoderma gangrenosum or adverse effects from prednisone intake such as infections, nausea, vomiting, bone pain, and blood pressure monitoring were also assessed. The importance of follow-up every week was emphasized as the lesion may recur and medications were given every week. Patient also reported the associated pain decreased from 7 to 3 out of 10.

DISCUSSION

Vegetative pyoderma gangrenosum, also known as superficial granulomatous pyoderma[7] classically presents as a localized vegetative or ulcerative lesion that lacks the violaceous undermined borders and is located usually on the trunk.[1] Only nine cases affecting one side of the face have

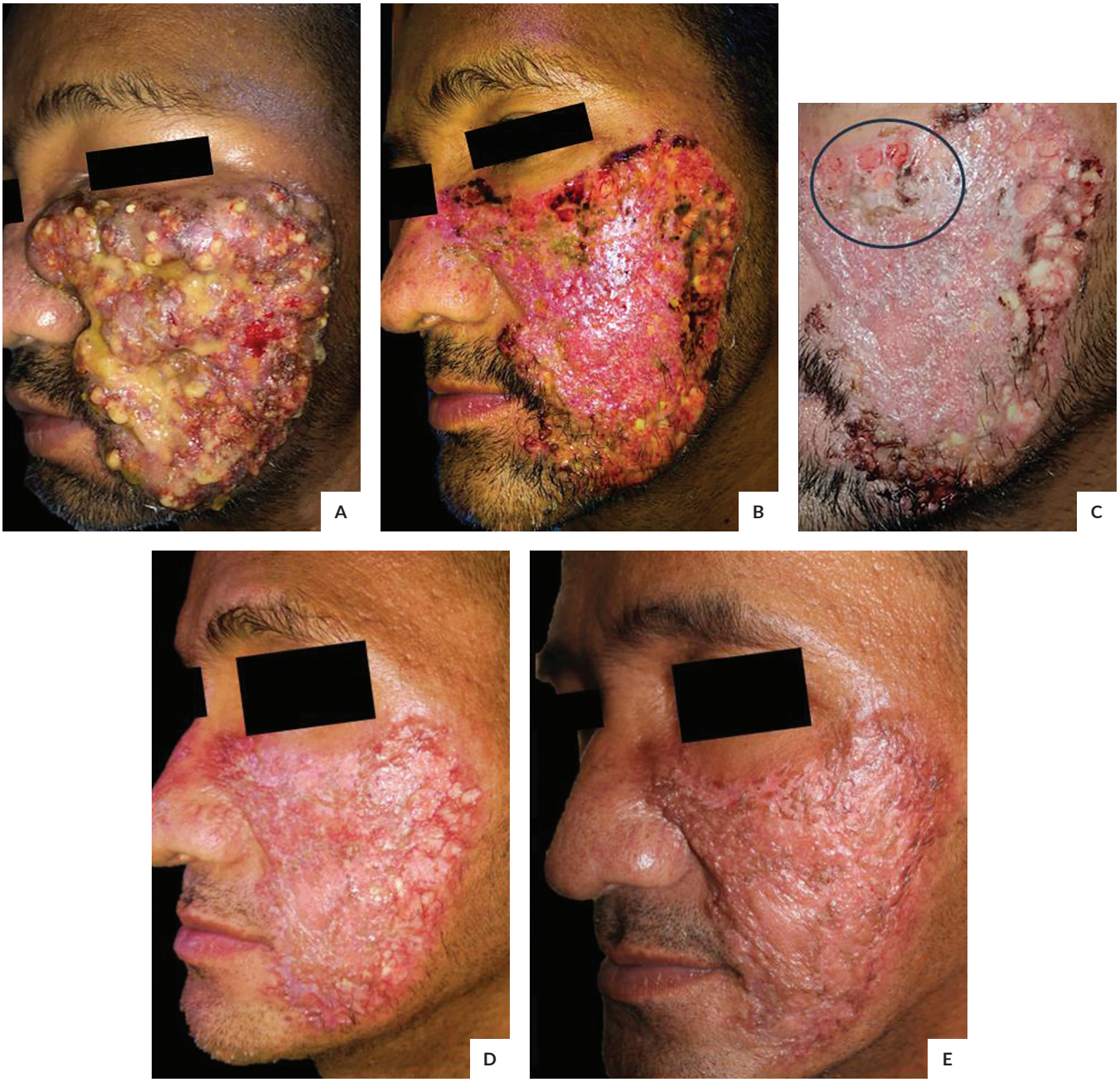

Figure 2. ***Use of high dose glucocorticoids in a case of vegetative pyoderma gangrenosum, left cheek.*** Prior to the initiation of methylprednisolone 1 g pulse therapy **(A)** for three days, which resulted in the decrease in induration **(B)** and formation of cribriform scarring **(C)** followed by Intravenous hydrocortisone 100 mg every 8 hours until discharged **(D)** then shifted to oral prednisone 60 mg/tab decreased by 10 mg every week until tapered off. After 30 days on glucocorticoids, there was more than 50% decrease in ulcer **(E)**.

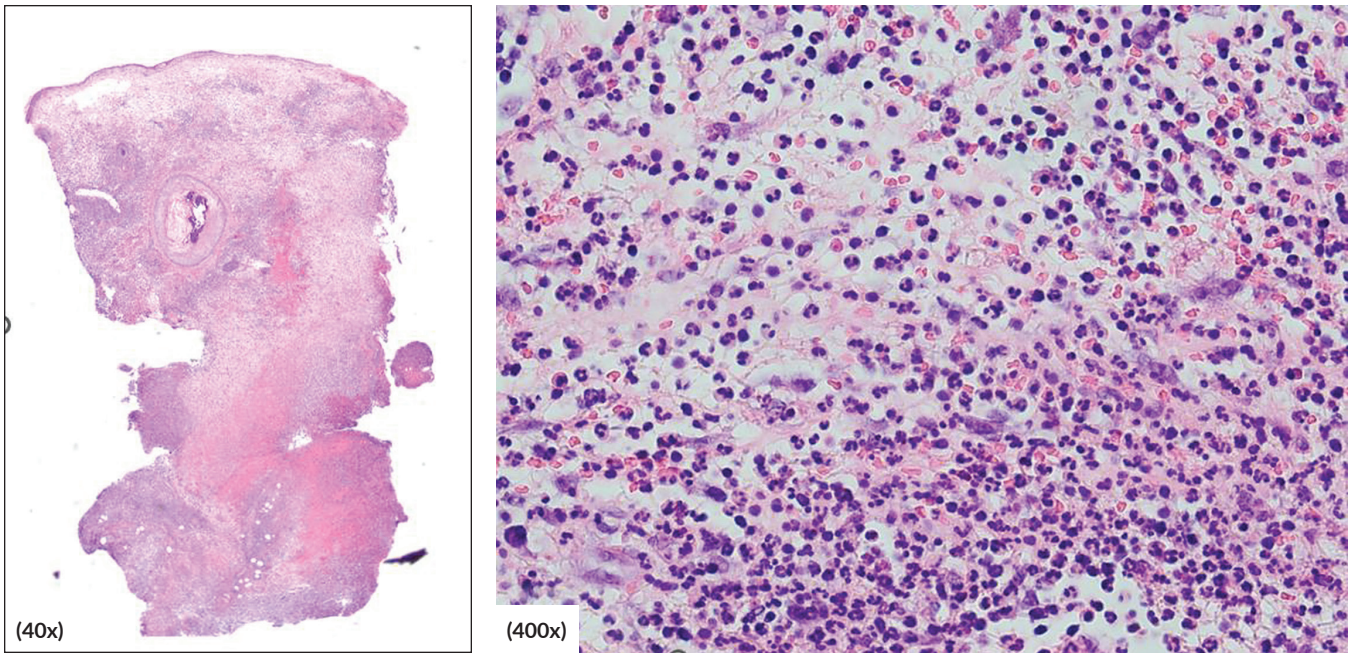

Figure 3. ***Histopathologic review of 4 mm-punch biopsy taken from the border of the ulcer on the left cheek (Hematoxylin and Eosin: 40x, 400x).*** Sections show diffuse dense infiltrates composed mostly of neutrophils, consistent with suppurative dermatitis.

Table 1. ***Diagnostic Criteria of Ulcerative Pyoderma Gangrenosum: A Delphi Consensus of International Experts[9]:*** Major criteria is required and 4 out of the 8 minor criteria for the diagnosis of pyoderma gangrenosum

Major criteria (1)	Minor criteria (8)
(1) biopsy of ulcer edge demonstrating neutrophilic infiltrate	(1) exclusion of infection
	(2) pathergy
	(3) history of inflammatory bowel disease or inflammatory arthritis
	(4) history of papule, pustule, or vesicle ulcerating within four days of appearing
	(5) peripheral erythema, undermining border, and tenderness at ulceration site
	(6) multiple ulcerations, at least one on an anterior lower leg
	(7) cribriform or "wrinkled paper" scar(s) at healed ulcer site
	(8) decreased ulcer size within one month of initiating immunosuppressive medication(s)

been reported.[2,7] This case is especially unique, since in our patient, the lesions occured on opposite sides of the face on two separate occasions spaced months apart.

Based on the diagnostic criteria of pyoderma gangrenosum (Table 1)[8,9], our patient met the major criteria and 4 of the 8 minor criteria for the diagnosis of pyoderma gangrenosum. The required major criteria of biopsy of the edge of the ulcer or plaque demonstrating diffuse dense infiltrates composed mostly of neutrophils, consistent with suppurative dermatitis (Figure 3) was met. The minor criteria that were met include the following: (1) history of a papule, pustule, or vesicle ulcerating within four days of appearance, (2) the decreased ulcer size within one month of initiating immunosuppressive medications (Figures 2A-D), (3) presentation of cribriform or wrinkled paper scar(s) at healed ulcer sites (Figure 2E), and (4) the exclusion of infection. The diagnostic criteria for pyoderma gangrenosum can maximize discrimination with 86% sensitivity and 90% specificity.[9] Other considerations included such as facial cellulitis and lupus vulgaris which were ruled out based on the biopsy and tissue culture. Non-infectious causes such as foreign body injectables like fillers were excluded due to a negative history.

Pyoderma gangrenosum is most commonly associated with inflammatory bowel disease (IBD), arthritis, and hematologic disorders in 78% of cases.[1] The remaining cases are considered idiopathic.[1] With regards vegetative pyoderma gangrenosum, there have been no specific associated diseases reported as of this writing.[1,7]

Vegetative pyoderma gangrenosum is considered a superficial variant and presents as a solitary, less aggressive plaque.[4] Hence, it is often responsive to topical modes of therapy, including topical and intralesional steroids and antimicrobial therapy.[1,4,7] There are no definitive guidelines for the treatment of pyoderma gangrenosum and management is

based on the severity and extent of the disease.[3,10] Despite the case being the vegetative variant of pyoderma gangrenosum, systemic therapy using high dose corticosteroids, such as the infusion of 1 gram methylprednisolone pulse therapy was initiated since the affected area was on the face. This was then followed by slow tapering of oral corticosteroids to avoid relapse of the plaques.[3] Gentle cleansing to prevent secondary infection is recommended.[7] Debridement or surgery is contraindicated due to pathergy, which is a minor criteria and occurs in 25-50% of cases.[4] The initial presentation on the right cheek may have slowly resolved because of the anti-inflammatory properties of the antibiotics[10] that were given to the patient on his past admission. With the treatment, the patient was relieved as the pain decreased but was worried as to recurrence of the disease. The financial limitation of the patient is factor for his health-seeking behavior. This also affects his follow-up consultations which would be managed in the out-patient basis as additional work up for screening of systemic associations despite the absence of symptoms such as abdominal pain was not done.

CONCLUSION

As pyoderma gangrenosum is a diagnosis of exclusion, cultures to exclude differentials diagnoses such as deep fungal infections, tuberculosis verrucosa cutis, and pyoderma vegetans[7] are essential to clinch the diagnosis. It is important to remember that not all inflamed and purulent lesions are infectious. Neutrophilic dermatoses should always be considered as prompt management can prevent prolonged therapy, delayed wound healing, and debilitating scarring.

Informed Consent

Patient consent was obtained on the use of photographs and histological slides, and publication of the case.

Statement of Authorship

All authors certified fulfillment of ICMJE authorship criteria.

Author Disclosure

All authors declared no conflicts of interest.

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