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Unilateral Vulvar Pyoderma Gangrenosum: A Case Report

Alshami MA[1]*, Alshami HM[2] and Mohana MJ[2]

[1]Department of Dermatology, Faculty of Medicine and Medical Sciences, Sana'a University, Sana'a, Yemen

[2]Department of Dermatology, Faculty of Medicine and Medical Sciences, Sana'a University, Sana'a, Yemen

ABSTRACT

Pyoderma gangrenosum (PG) is a rare, chronic, relapsing, ulcerative neutrophilic dermatosis type with a peculiar morphologic presentation. PG of the vulva is extremely rare, and unilateral involvement of the vulva has not been previously reported. We present herein a case of unilateral PG of the vulva in a 22-year-old female patient. The patient responded excellently to treatment with oral prednisolone 40 mg/day tapered over 3 weeks, combined with azathioprine 50 mg thrice a day from the 10th day of treatment.

***Corresponding author**

Mohammad Ali Alshami, Department of Dermatology, Faculty of Medicine and Medical Sciences, Sana'a University, Sana'a, Yemen. Tel: 00967733760082.

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Abbreviations

PG: Pyoderma Gangrenosum

IL: Interleukin

Introduction

Pyoderma gangrenosum (PG)—a term coined by Brunsting et al. in 1930—is a rare, chronic, relapsing, ulcerative neutrophilic dermatosis type with a peculiar morphologic presentation [1]. It is associated with systemic disease. Diagnosis of PG is based on clinical findings, and it is often under-recognized and under-reported [2,3]. The typical PG lesion presents as an extremely painful ulcer with a dead attenuated border and a base that may be suppurative or vegetative [4]. Herein, we report a case of unilateral PG of the vulva in a 22-year-old female patient.

Case Presentation

The 22-year-old woman presented to the dermatology clinic with a painful ulcer over the left side of the vulva. It had appeared 3 months prior, initially as erythematous papules and eventually rupturing to form painful ulcers with serosanguinous discharge. She had no history of vesiculation, itching, vaginal discharge, or joint pain. Her general health was unremarkable. Cutaneous examination revealed a tender ulcer approximately 4×8 cm in size, with undermined edges and an indurated base. It was restricted to the left labium majus and minus and was covered with slough and hemorrhagic crusts (Figure 1). Several topical and systemic antibiotics prescribed by various physicians had been ineffective. On the basis of the patient's presentation and history of antibiotic ineffectiveness, we diagnosed PG of the vulva, which was supported by the patient's rapid response to oral prednisolone.

The patient had an excellent response to treatment with oral prednisolone 40 mg/d tapered over 3 weeks (Figure 2) and azathioprine 50 mg thrice a day—a steroid-sparing agent that was added from the 10th day of starting prednisolone. During a monthly follow-up, mild recurrence, induced by stopping prednisolone completely (Figure 3), was observed 6 months after starting treatment but disappeared after increasing the prednisolone dose to 20 mg twice daily in addition to continuing azathioprine 50 mg thrice a day.

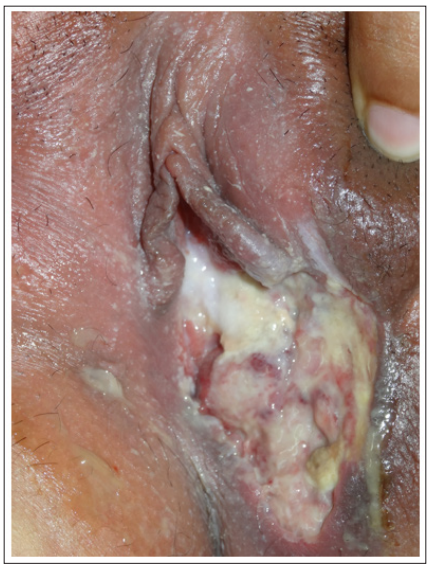

Figure 1: Before treatment, a large ulcer was observed. It had an irregular shape and undermined borders, and the base was covered with white creamy necrotic material, involving the lower third of the left vulva.

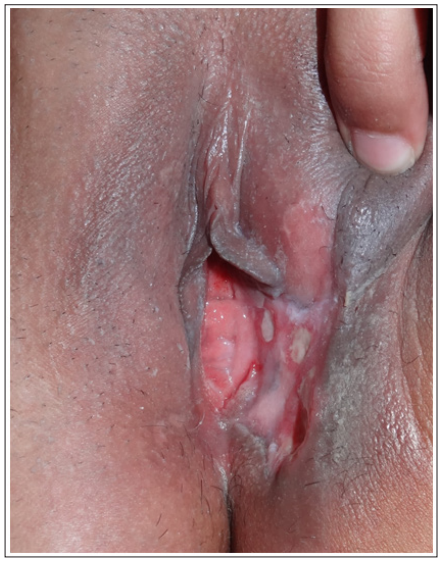

Figure 2: One month after treatment, almost complete healing of the ulcer had taken place; the base was clean except for two small, shallow ulcers.

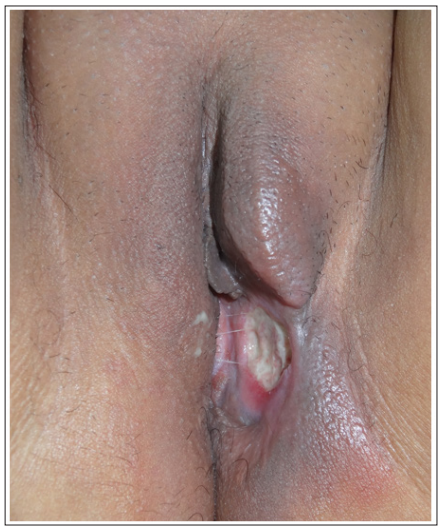

Figure 3: Six months after treatment, a large, single ulcer with a white necrotic base and edema of the left labium majus appeared 1 week after completely stopping prednisolone.

Discussion

PG of the vulva is extremely rare, and a unilateral affection of the vulva has not been previously reported. The yearly estimated incidence of PG is 3-10 cases per million [5]. Clinically, four major types are recognized: ulcerative, bullous, pustular, and superficial granulomatous [6]. It was first described by Brocq in 1916, though the term "pyoderma gangrenosum" was put forth by Brunsting et al. in 1930, who also promoted the theory of infectious etiology (streptococci and staphylococci) [7]. Women between 20 and 50 years of age are the most affected, and half of affected patients have an underlying systemic disease, such as inflammatory bowel disease, hematologic disorder, or arthritis. Infants and children represent 4% of patients with PG. Various monogenic autoinflammatory diseases present in association with PG features. Increased activity of the interleukin (IL)-1 pathway is thought to be involved in PG, similar to several autoinflammatory disorders [8]. This proposed pathogenesis is additionally supported by elevated serum levels of IL-1β and the positive effect of anti-IL-1β antibody canakinumab in these patients. The efficacy of corticosteroids in PG may be explained by their ability to suppress the synthesis of IL-1α and IL-1β [9]. Up to one third of patients with PG demonstrate pathergy phenomenon, an enhanced reaction to trivial trauma, that initiates and aggravates the cutaneous lesions [10]. The typical lesion of classic ulcerative PG is a painful ulcer over the anterior surface of the shin [11]. Based on clinical presentation, location, and associated diseases, other variants (bullous, pustular, and superficial granulomatous/vegetative) are recognized [12]. The case presented herein is the first of its kind, as PG affected the vulva in a unilateral fashion. Treatment was initially successfully with appropriate beside wound care and administration of oral prednisolone, then later of azathioprine. Clinicians should consider the possibility of PG on the vulva. It usually presents bilaterally, but in rare instances, it presents unilaterally. It requires early diagnosis and prompt appropriate treatment to avoid complications.

Conflicts of Interest

The authors declare that they have no conflict of interest.

References

1. David F, Lopes Freitas R, Brás-Cruz R, Rocha J Rosário C A (2022) Case of Recurrent Idiopathic Pyoderma Gangrenosum. Cureus 14: e25112.
2. Chen B, Li W, Qu B (2023) Practical aspects of the diagnosis and management of pyoderma gangrenosum. Front Med (Lausanne) 10: 1134939.
3. Hobbs MM, Ortega-Loayza AG (2020) Pyoderma gangrenosum: From historical perspectives to emerging investigations. Int Wound J 17: 1255-1265.
4. George C, Deroide F, Rustin M. (2019) Pyoderma gangrenosum - a guide to diagnosis and management Clin Med (Lond) 19: 224-228.
5. Monari P, Moro R, Motolese A, Misciali C, Baraldi C, et al. (2018) Epidemiology of pyoderma gangrenosum: Results from an Italian prospective multicentre study. Int Wound J 15: 875-879.
6. Bhat RM (2012) Pyoderma gangrenosum: An update. Indian Dermatol Online J 23: 7-13.
7. Ye MJ, Ye JM (2014) Pyoderma gangrenosum: a review of clinical features and outcomes of 23 cases requiring inpatient management. Dermatol Res Pract 2014: 461467.
8. Maronese CA, Pimentel MA, Li MM, Genovese G, Ortega-Loayza AG, et al. (2022) Pyoderma gangrenosum: an updated literature review on established and emerging pharmacological treatments. Am J Clin Dermatol 23: 615-634.
9. Yamamoto T, Yamasaki K, Yamanaka K, Komine M, Kawakami T, et al. (2023) Clinical guidance of pyoderma gangrenosum 2022. J Dermatol doi: 10.1111/1346-8138.16845.
10. Varol A, Seifert O, Anderson CD (2010) the skin pathergy test: innately useful? Arch Dermatol Res 302: 155-168.
11. Bissonnette C, Kauzman A, Mainville GN (2017) Oral pyoderma gangrenosum: diagnosis, treatment and challenges: a systematic review. Head Neck Pathol 11: 427-441.
12. Brooklyn T, Dunnill G, Probert C (2006) Diagnosis and treatment of pyoderma gangrenosum. BMJ 333: 181-184.