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CASE REPORT

Pyoderma gangrenosum: A single-center case series study of 32 cases

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ABSTRACT

Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis with multiple different clinical presentations and associated comorbidities. It often presents as ulcerated lesions with a violet/erythematous border and an irregular undercut margin. In this largest single-centre case series study in Antalya-Turkey, we reviewed 32 PG patients diagnosed consecutively within the last 5-year period. Consistent with the literature, PG morphologically often presented with the ulcerative clinic (90.6%), and inflammatory bowel disease (15.6%) was the most common etiologic factor. In our study, female gender predominance (78.2%), lower extremity localization (93.7%), and the rate of multiple ulcers at diagnosis (90.6%) were more prominent than in the literature. Unlike the literature, oral mucosa involvement and syndromic form (both, 6.2%) of the disease were detected more frequently. Our results indicate that PG patients may show differences according to geographical and ethnic differences and/or characteristics of the healthcare institution.

Keywords: pyoderma gangrenosum; case series; neutrophilic dermatosis

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1. Introduction

Pyoderma gangrenosum (PG) is an uncommon neutrophilic dermatosis characterized by ulcers with a violet-coloured active edge, undermined borders and peripheral erythema. The disease usually develops between 40–60 years of age and is more common in women. Several clinical variants have been described, including classical ulcerative, bullous, pustular, vegetative, peristomal and postoperative[1]. Bullous PG presents with blisters that are initially present and then transform into ulcerative lesions[2]. Vegetative lesions usually develop on the surface of previously classic ulcerative PG, often during treatment[3], whereas pustular lesions usually precede or accompany PG ulcers. Classical ulcerative PG is the most common clinical variant (**Figure 1**). Almost half of PG cases are associated with systemic diseases, including inflammatory bowel disease (IBD), hematologic disorders, solid organ malignancy and inflammatory arthritis[3].

While the data regarding PG in our country are limited[4–6], there is no epidemiological data for Antalya and its neighbourhoods. The hypothesis of the study was that PG patients may show differences according to geographical, ethnic differences and/or characteristics of the healthcare institution. This study aimed to describe the sociodemographic and clinical features, comorbidities and management of PG patients diagnosed in our clinic for the last 5 years and to compare them with the literature.

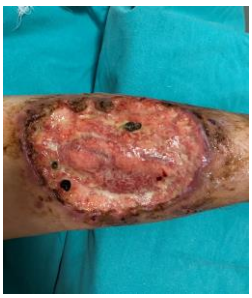

Figure 1. Ulcerative (classic) type PG located on the posterior aspect of the tibia.

2. Methods

The diagnosis of PG is made clinically and with histopathological confirmation in necessary patients. 32 consecutive PG patients admitted to our clinic between January 2017 and December 2021 were included in this case series study. The data of the patients were accessed through the patient registration programme with the approval of the ethics committee of our hospital (ethics committee approval number: 70904504/204). In case of missing data, they were contacted by telephone. Patients were evaluated in terms of sociodemographic characteristics (age, gender, age at onset, comorbidities), possible triggering factors, history of drug use, dermatological examination findings, clinical subtypes and treatment protocols used.

3. Results

The mean age was 48 years (range, 21–92), seven (21.8%) were male, and 25 (78.2%) were female. Among the comorbidities, the most common was inflammatory bowel disease (IBD) (ulcerative colitis, 3; Crohn's disease, 2; total 5 [15.6%]) which was followed by inflammatory arthritis (9.3%), hidradenitis suppurativa (6.2%), Behçet's disease (6.2%), PASH (pyoderma gangrenosum, acne and hidradenitis suppurativa) (3.1%), PAPASH (pyogenic arthritis, pyoderma gangrenosum, acne and hidradenitis suppurativa) (3.1%) and hematologic malignancy (3.1%).

The most common clinical subtype was ulcerative (classic) type PG (93.7%). Two or more lesions at diagnosis was observed in 90.6% of patients. Lower extremities (93.7%) were the most common localization, followed by trunk (28.1%), upper extremities (15.6%), oral mucosa (6.2%), scalp (6.2%) and face (3.1%). Pathergy reaction was positive in 34.3% of patients. The clinical features of our patients with PG are summarized in **Table 1**. Corticosteroids and cyclosporine were the most commonly preferred therapies in the systemic treatment of the patients (**Table 2**). During the 5-year follow-up period, we observed mortality in four patients (12.5%). Two of the deaths were associated with PG-induced immunosuppression. The other two were unrelated to PG.

Table 1. Clinical characteristics of patients with pyoderma gangrenosum.

Clinical characteristics		**Number of patients, *n*: 32 (%)**
Clinical type	Ulcerative	29 (90.6)
	Bullous	2 (6.3)
	Pustular	1 (3.1)
	Vegetative	-
Number of lesions	Single	3 (9.3)
	Multiple (2 or more)	29 (90.7)
Pathergy reaction	Surgery	6 (21.4)
	Insect bite	3 (9.3)
	Trauma	1 (3.1)
	Injection	1 (3.1)

Table 1. (*Continued*).

Clinical characteristics		Number of patients, *n*: 32 (%)
Comorbidities	Inflammatory bowel diseases	5 (15.6)
	Inflammatory arthritis	3 (9.3)
	Behçet's disease	2 (6.2)
	Hidradentis suppurativa	2 (6.2)
	Syndromic (PASH, PAPASH)	2 (6.2)
	Hematologic malignancies	1 (3.1)
Location	Lower extremity	30 (93.7)
	Trunk	9 (28.1)
	Upper extremity	5 (15.6)
	Oral mucosa	2 (6.2)
	Scalp	2 (6.2)
	Face	1 (3.1)

Table 2. Characteristics of systemic treatments of patients with pyoderma gangrenosum.

Treatment	Number of patients, *n* (%)
Systemic corticosteroids	11 (34.3)
Cyclosporine	5 (15.6)
Systemic corticosteroid + cyclosporine	14 (43.7)
Systemic corticosteroid + sulfasalazine	4 (12.5)
TNF-alpha inhibitors	4 (12.5)

4. Discussion

Our study is the largest case series study of PG in Turkey to date. Our results demonstrated that PG was frequently seen in women, morphologically often presented as ulcerative type, most commonly located in the lower extremities with multiple ulcers, and IBD was the most common etiologic factor.

The mean age ranged from 42 to 53 in previous studies, with 20, 25, and 27 patients conducted in Turkey. In our study, the mean age (48) was consistent with the Turkish data. In the literature, it has been reported that the disease is observed more frequently in women, with rates ranging between 52%–79%. Unlike the three studies conducted in Turkey, female predominance was more prominent in our study (55%–63% vs. 78.2%)[4–6]. PG ulcers are most commonly localized in the lower extremity and especially on the tibia. Binus and Qureshi[7] in a case series of 103 patients from the UK, reported lower extremity localization in 78% of patients. They found two or more ulcers in 65% of the patients in the same study. In previous studies from Turkey[4–6], lower extremity involvement ranged between 70%–92% and multiple lesions were found in 30%–70% of patients. In general, the PG localization (93.7%) was consistent with the literature in our study, while the rate of multiple ulcers at diagnosis (90.6%) was higher than in the literature.

Although it was not reported in any of the previous studies in Turkey, PG was observed in the oral mucosa in two patients (6.2%) in our study (**Figure 2**).

The most common comorbidity of PG in our study was IBD, which is in parallel with the results of a recent comprehensive meta-analysis, including 2611 patients. In a retrospective study involving 53 cases of PG in a South Korean population with a 20-year duration, 22 (41.5%) patients were treated with systemic corticosteroids and systemic corticosteroids were among the most commonly used treatment in our case series[8].

Our study had several limitations. Some data were self-reported an*d* the study included relatively limited number of cases. Therefore, caution should be exercised when generalizing the results to the population.

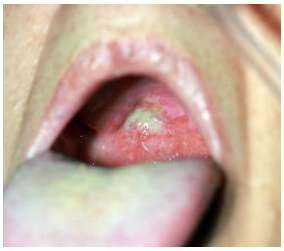

Figure 2. Ulcerative type PG located in the oral mucosa.

Conflict of interest

The authors declare no conflict of interest.

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