Original Article

Clinicopathological Profile of Pyoderma Gangrenosum: A 10-Year Retrospective Study from a Tertiary Care Center in South India

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Abstract

Background: Pyoderma gangrenosum (PG) is a reactive neutrophilic inflammatory dermatosis with a varied clinicopathologic presentation. It commonly manifests as rapidly progressive painful ulcers, mimicking varied conditions including infections, vasculitis, and malignancies, and is a diagnosis of exclusion. There are scarce data on PG from the Indian subcontinent. **Aim and Objectives:** The aim of the study was to study the clinicopathologic profile of patients with PG and their underlying systemic associations. **Materials and Methods:** A retrospective observational study was done between 2011 and 2021, and patients diagnosed as PG based on the diagnostic tool proposed by Maverakis *et al*. were recruited and their demographic, clinical, and histological findings were obtained. **Results:** Among 54 patients with suspected PG, 17 patients (eight males and nine females) fulfilled the diagnostic criteria, and the mean age of disease onset was 32.1 years (range: 3–60 years). Ulcerative variant was the most common type (9/17, 52.9%), and 29.4% had systemic associations including autoinflammatory syndromes. The onset at atypical sites such as face and hand were noted in one patient each. Histopathology revealed a polymorphous dermal infiltrate with neutrophilic predominance in the majority (94.1%). Systemic steroids (dose ranging from 0.5–1 mg/kg prednisolone equivalent) were used in 11/17 (64.7%) patients. The commonly used alternative drugs included clofazimine (47%), minocycline (29%), thalidomide (23.5%), adalimumab and mycophenolate mofetil in 17.6% each, dapsone and ciclosporine in 11.7% each. Remission was achieved between two weeks and three months in 10 (58.8%) patients after treatment initiation and two mortalities (11.7%) were recorded. **Conclusion:** PG can affect any age group and may be localized to rarer, atypical sites. The possibility of underlying autoinflammatory conditions should be considered in addition to the evaluation of other disorders like inflammatory bowel disease, hematological disorders, and rheumatological disorders.

Keywords: *Autoinflammatory syndromes, neutrophilic dermatosis, pyoderma gangrenosum*

Introduction

Pyoderma gangrenosum (PG) is a non-infectious, reactive inflammatory neutrophilic dermatosis with a variable prognosis and is often associated with systemic disorders. PG can have a heterogeneous clinical presentation and is characterized by neutrophilic infiltrate on histopathology.[1] Though both the innate and adaptive immune system components are involved in the evolution of PG, the exact pathogenetic mechanisms have not been fully delineated. PG is clinically characterized by painful raised lesions that ulcerate resulting in excruciatingly painful single or multiple ulcers mostly on the anterior lower legs, with raised undermined borders, tender dusky red or purplish margins, and a boggy or necrotic base.[2,3] The onset could be either explosive with rapid spread of lesions or indolent and gradually progressing. The ulcers usually heal with thin, atrophic cribriform scars. The pathergy phenomenon, which refers to the development of new lesions or exacerbation of preexisting lesions at sites of trivial trauma, is noted in about 20% of cases.[2] The clinical variants include the classic ulcerative type (the most common), bullous, pustular, vegetative, peristomal, and postoperative types. PG is considered to be a diagnosis of exclusion as there are no specific biologic markers.[4] The criteria initially proposed by Su *et al*.[4,5] have been replaced by the later validated diagnostic criteria by Maverakis *et al*.[6,7] and the PARACELSUS score.[8] There are a few Indian studies relating to this entity, and we therefore did a retrospective 10-year analysis

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to understand the clinicopathologic patterns and systemic associations of PG.

Materials and Methods

A retrospective observational study was conducted by collecting demographic, clinical, and histopathological data of patients in all age groups who had attended the dermatology department between January 2011 and 2021, in whom PG was diagnosed based on the improved diagnostic tool by Maverakis *et al.*[6] [Table 1]. Patients who did not fulfill the diagnostic criteria were excluded from the study. The study was approved by the institutional review board. The clinicopathological profile and concomitant systemic diseases were estimated in this cohort of patients by descriptive statistics using Microsoft Excel.

Results

Among 54 patients with a diagnostic suspicion, 17 were diagnosed with PG based on the diagnostic criteria by Maverakis *et al.*[6] [Figure 1]. The male-to-female ratio was 0.89:1 (eight males and nine females), and the mean age at presentation was 34.9 years (range: 18–62 years, SD: 14.15 years). The age of disease onset ranged from 3–60 years, with a mean age of disease onset of 32.1 years (SD: 14.86 years). Two males had juvenile onset at 3 and 15 years of age. The mean disease duration at presentation was 35.1 months. The demographic and clinical characteristics have been summarized in Table 2.

Morphology and site of involvement

The initial lesions observed were papules (nine patients, 52.9%), pustules (four patients, 23.5%), and blisters (five patients, 29.4%). The most common site of disease onset was the leg in 11 patients (64.7%). However, the disease progressed to involve other sites such as the thighs, abdomen, forearms, shoulder, and face in 5/11 patients (45%). Lower limb involvement alone was noted in nine patients (66.6% – confined only to the legs and 33.3% – thighs). The onset was noted on the face in one patient with facial confinement, hands in one patient with progression to the hip, and perianal area in one patient who had subsequent peristomal PG.

The progression was gradual in 10 patients and rapid in the remaining seven patients. The lesions were solitary in five patients (29.4%), 4/17 had 2–4 lesions (23.5%), and eight (47.05%) had multiple lesions. Pathergy could be elicited in only two patients, was negative in four, and the reports were not available in the remaining 11 patients.

The most common clinical type was the ulcerative variant in nine (52.9%) patients. Scarring was recorded in 9/17 patients [Figures 2–6]. There was no overlap with any other neutrophilic dermatosis.

Comorbidities and systemic associations

Systemic associations commonly reported in PG were seen in 5/17 patients (29.4%) and included inflammatory bowel disease (IBD)- Crohn's disease in two patients, of whom one had peristomal PG, one patient had seronegative arthritis, and two had autoinflammatory conditions (1- pyogenic arthritis, PG and acne (PAPA) and 1- PG, acne and suppurative hidradenitis (PASH) each), diagnosed based on the clinical features and not genetically proven.

Table 1: Diagnostic criteria of ulcerative pyoderma gangrenosum by Maverakis *et al.*[6,7]

Major criterion
Biopsy from the ulcer edge demonstrating neutrophilic infiltrate
Minor criteria
(1) Infective causes excluded
(2) Positive pathergy test
(3) History of inflammatory bowel disease or inflammatory arthritis
(4) History of papule, pustule, or vesicle ulcerating within four days of onset
(5) Peripheral erythema, undermining border, and tenderness at the site of ulceration
(6) Multiple ulcerations, at least one on the anterior lower leg
(7) Cribriform or "wrinkled paper" scar (s) at healed ulcer sites; and
(8) Decreased ulcer size within 1 month of initiating immunosuppressive medications
1 major criterion and 4/8 minor criteria are needed to diagnose pyoderma gangrenosum.

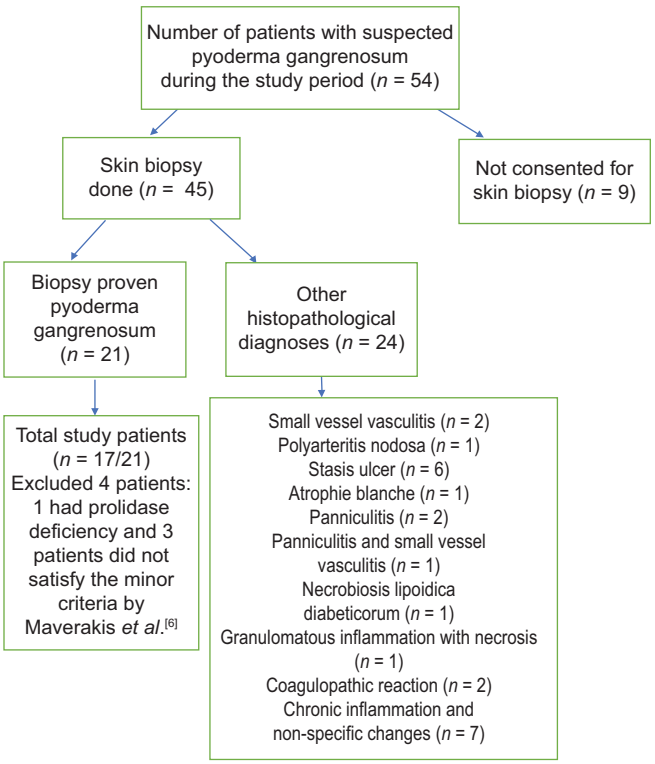

Figure 1: Patient flowchart

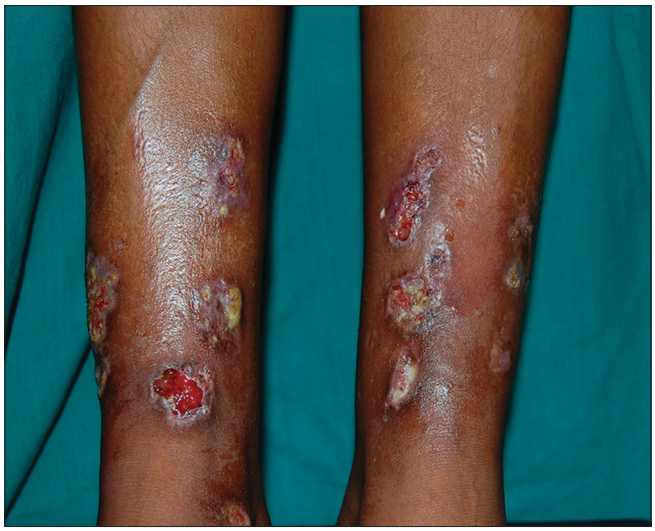

Figure 2: Pustular morphology preceding PG on the legs

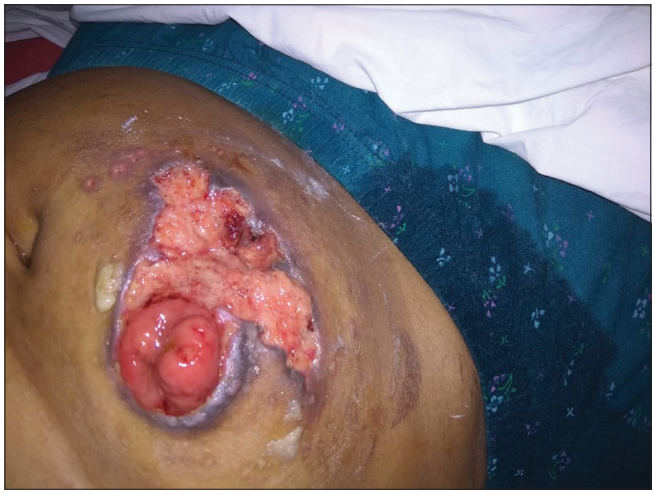

Figure 4: Peristomal PG in a patient with Crohn's disease

An early age of disease onset (three years) was noted in the male patient with PAPA syndrome. Electrophoresis was done in 7/17 patients, among whom 2/7 (28.6%) showed an abnormal pattern with an increase in the alpha and gamma regions, respectively. These patients did not follow up for further evaluation and hence haematological malignancies could not be established. Fecal calprotectin levels measured in eight patients were negative. Rheumatoid arthritis factor done in 9/17 patients was negative. Various comorbidities including diabetes mellitus, hypertension, anemia were also observed as listed in Table 2.

Histopathology

The most common histopathological feature was dermal inflammatory infiltrate, exclusively neutrophilic in 8/17 (47%) patients and polymorphous with predominant neutrophils in 16/17 (94.1%) patients. Vasculitis was present in 4/17 (23.5%) biopsies. Granulomatous inflammation with neutrophilic dermal infiltrate was noted in 2/17 (11.7%) cases. [Figure 7a and b]

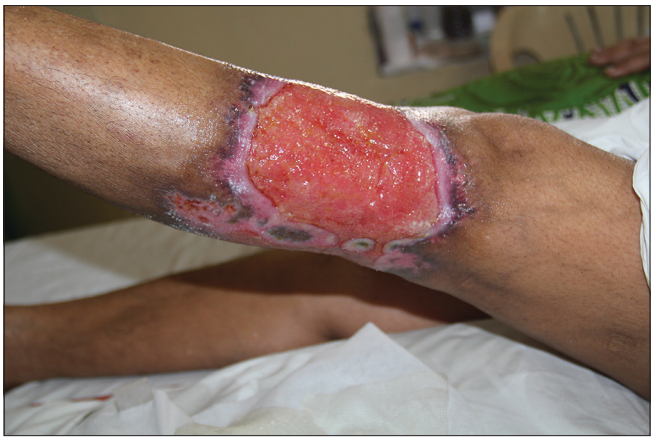

Figure 3: Classic ulcerative pyoderma gangrenosum on the leg

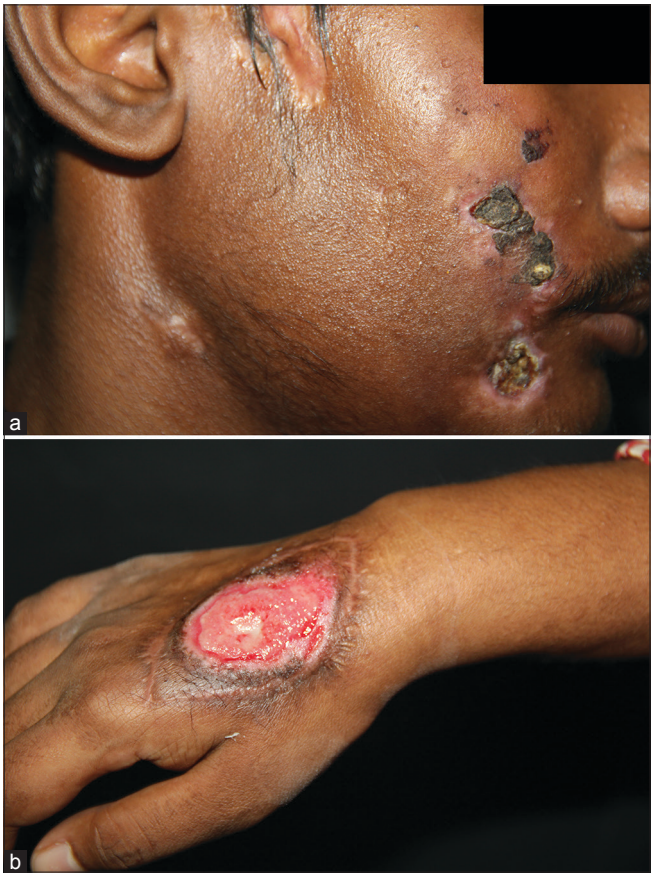

Figure 5: (a) PG at atypical sites – on the face with crusting and (b) hand

Treatment

Eleven (64.7%) patients received systemic steroids at presentation ranging from prednisolone equivalent doses of 15 mg to 75 mg in a tapering schedule (maximum dose of 1 mg/kg). All patients received combination therapy with steroid-sparing agents. Pus culture sensitivity was assessed in relevant cases, and secondary infections were treated adequately with antibiotics. The steroid-sparing agents used included clofazimine 100 mg in eight patients, minocycline 100 mg in five patients, thalidomide 50–100 mg in four

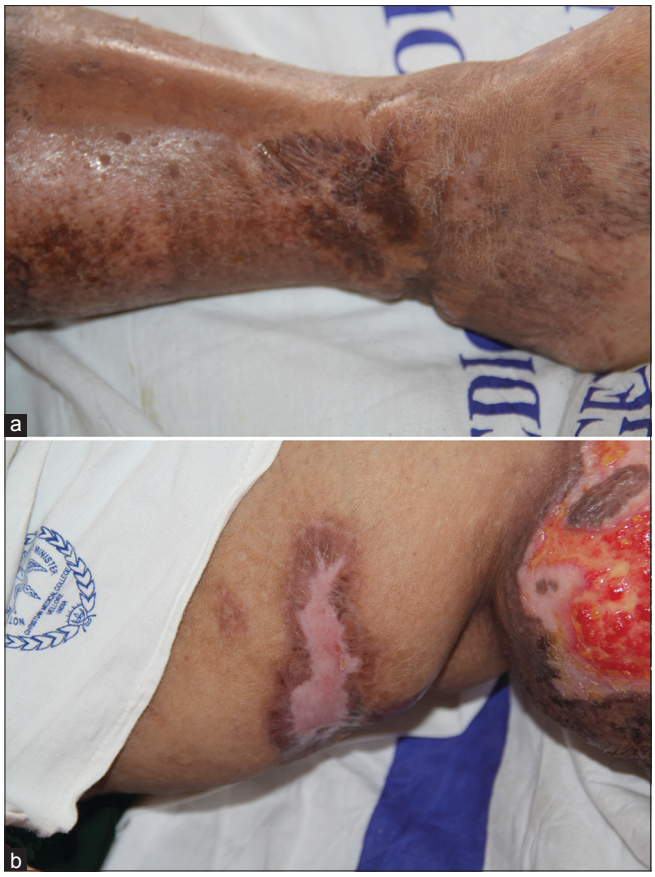

Figure 6: (a and b) Cribriform scarring at the site of healed PG

cases, mycophenolate mofetil (1.5–2 g/day) in three patients, dapsone 100 mg, ciclosporine (2–3 mg/kg), IVIG (intravenous immunoglobulin, 1–2 g/kg), and azathioprine (maximum dose of 100 mg/day) in two patients each, and one patient received methotrexate (10 mg weekly). Three patients received subcutaneous adalimumab at 40-160 mg biweekly doses in a treatment span of 1 month to three years [Table 2]. The patient with PASH syndrome received 2 g of intravenous immunoglobulin; however, due to the development of new lesions and the refractory nature of preexisting ulcers, an infliximab 400 mg infusion was given in four doses at 0, 2, 6, and 14 weeks, with remission lasting only two months. Three patients had received only antibiotics (initiated in view of secondary infection at the first visit) as 2 were lost to follow up and one died shortly after the first visit.

Follow-up

Three patients didnot review after the first visit and the follow-up period for the remaining 14 patients ranged from seven days to three years. Resolution of symptoms and healing were noted in 10/14 patients in a span ranging from two weeks to three months, and only one patient had complete remission at two weeks. One patient expired following acute coronary syndrome, and the other patient subsequently succumbed to fungal pneumonia

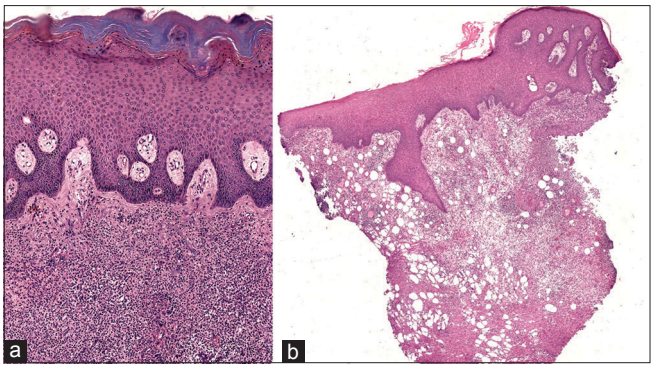

Figure 7: (a) H and E, 10x, showing diffuse dermal neutrophilic infiltrate in the upper dermis. (b) H and E, 10x, showing few ill-defined granulomas in the upper dermis with predominant neutrophilic infiltrate

and septicemia. In four patients, ulcers recurred while on treatment after remission, with the relapse duration ranging from two months to one year. One patient with ulcerative PG on adalimumab developed paradoxical psoriasis at one year of follow-up. The entire cohort didnot have relapse of the systemic disease during follow-up. On telephonic follow-up, On telephonic follow-up, 8/15 survivors could be traced, among whom 6/8 patients were in remission off treatment (duration ranging from 2-7 years), one female patient died due to other causes; and one male patient had a recurrence of ulcer on the leg of 1-week duration after being in remission for 3 years.

Discussion

The Maverakis *et al.*[6] criteria, which was proposed by the Delphi consensus, was chosen as it has a sensitivity of 86% and a specificity of 90%. Our study showed a female predilection (9/17, 52.9%), which was in concordance with the studies by Adışen *et al.*,[9] Ye and Ye,[10] Al Ghazal *et al.*,[11] and Schosler *et al.*[12] The mean age of presentation in our study was 34.9 years similar to other studies.[3,13] PG usually has an adult onset between 25 and 54 years of age. However, we had two male patients who had a juvenile onset at 3 and 15 years of age, which is quite rare and could represent autoinflammatory disorders with PG. In the case series by Bhat *et al.*,[13] the mean age was 34.39 years and there were five (27.77%) paediatric cases. The most common site of onset was the leg (11/17, 64.7%) and the classic ulcerative variant was the most common type (9/17, 52.9%), as noted in most other studies.[3,9,10,13–15] The reason for localization to the leg is not clear, and coexisting diabetes or peripheral vascular disease could contribute to the disease worsening.[9,15] Lesions can occur at other sites, as noted in three of our patients who had involvement of the face, hand, and peristomal area along with perianal involvement, respectively. PG at atypical sites such as the face and hand has been reported in literature.[12,16,17] Pustular PG was observed in three patients in our study. Bullous PG often considered as a marker of hematological malignancies was noted in 4/17 patients, among whom three had rapid disease progression, but hematological associations

Table 2: Demographic and clinical characteristics of our study patients

No	Gender (M/F)	Age of onset of disease (years)	Initial morphology	Sites of involvement	No. of lesions (*n*)	Type of PG	Systemic association/ comorbids	Steroid at outset (Y/N)	Steroid maximum dosage given	Treatment given	Follow-up duration
1	M	35	Pustule	Thigh	One	Pustular	Nil	N	Nil	Antibiotics	Lost
2	M	45	Papule	Leg	One	Ulcerative	Treated TB	Y	0.5 mg/kg prednisolone	Prednisolone and thalidomide	Seven days
3	F	24	Bulla, pustule	Leg and abdomen	Two	Ulcerative	Crohn's disease	N	Nil	Ciclosporine, minocycline, clofazimine, and mesacol	Four days
4	M	3	Pustule	Leg, thigh, and face	Multiple	Pustular	PAPA syndrome	Y	Prednisolone 0.5 mg/kg	Prednisolone, adalimumab, and methotrexate	Three months
5	F	60	Papule	Thigh	Multiple	Ulcerative	Hypertension	N	Nil	Antibiotics	Expired due to acute coronary syndrome
6	M	15	Papule	Leg	Multiple	Ulcerative	Nil	Y	Prednisolone 1 mg/kg	Prednisolone, ciclosporine, and clofazimine	14 days
7	M	38	Papule	Leg	Two	Ulcerative	Iron deficiency anemia, hypokalemia, and vitamin D deficiency	Y	Prednisolone 1 mg/kg	Prednisolone, clofazimine, and thalidomide	One month
8	F	34	Bulla	Hands and hip	Three	Bullous	Seronegative arthritis, herpes simplex, and iatrogenic Cushing's syndrome	Y	Prednisolone 1 mg/kg	Predisolone, antibiotics, and MMF	One month
9	F	30	Papule	Legs, face, and trunk	Multiple	Ulcerative	PASH syndrome	Y	Prednisolone 0.5 mg/kg	Prednisolone, IVIg, HCQS, dapsone, clofazimine, cyclophosphamide, adalimumab, MMF, minocycline, azathioprine, thalidomide, and infliximab	Two years
10	F	43	Bulla	Leg	One	Bullous	Hemophilia A carrier state, pancytopenia, hypertension, and esophageal candidiasis	N	Nil	Clofazimine	Three years
11	F	52	Papule	Leg	One	Ulcerative	Nil	Y	Methylprednisolone 1 mg/kg	Methylprednisolone, IVIg, clofazimine, and minocycline	Expired after 3 months follow up due to fungal pneumonia
12	M	18	Papule	Face	One	Ulcerative	Nil	Y	Prednisolone 1 mg/kg	Prednisolone and minocycline	Five months

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Table 2: Contd...

No	Gender (M/F)	Age of onset of disease (years)	Initial morphology	Sites of involvement	No. of lesions (*n*)	Type of PG	Systemic association/ comorbids	Steroid at outset (Y/N)	Steroid maximum dosage given	Treatment given	Follow-up duration
13	M	19	Pustule	Legs, forearm, and trunk	Multiple	Pustular	Nil	Y	Prednisolone 1 mg/kg years	Prednisolone and dapsone	16 months
14	F	27	Bulla	Thigh	Multiple	Bullous	Type 1 diabetes	N	Nil	Antibiotics	Lost
15	F	31	Bulla	Leg	Multiple	Bullous	Suspected Turner's syndrome	Y	Prednisolone 0.5 mg/kg	Prednisolone, clofazimine, minocycline, and MMF	One month
16	M	50	Papule	Legs, shoulders, and forearms	Two	Ulcerative	Anemia and dyslipidemia	N	Nil	Antibiotics and clofazimine	Lost
17	F	22	Papule	Peristomal, perianal, and axilla	Multiple	Peristomal	Crohn's disease, perianal fistula, TB lymphadenitis, and TNF-associated paradoxical psoriasis	Y	Prednisolone 0.5 mg/kg	Prednisolone, antibiotics, thalidomide, azathioprine, and adalimumab	Three years

MMF=Mycophenolate mofetil; HCQS=Hydroxychloroquine; TNF=Tumor necrosis factor; DM=Diabetes mellitus; TB=Tuberculosis; IV Ig=Intravenous immunoglobulin

could not be established in any of them. Most of our patients had multiple ulcers (70.5%), which was in concordance with the findings by Binus *et al.,*[15] but contrary to the study by Riyaz *et al*.[3]

Systemic associations have been reported in 50–70% of cases of PG with conditions such as IBD, inflammatory arthritis, hematological disorders, and internal malignancies.[10,18] In our study, systemic involvement was noted only in 29.4%, which is similar to the findings by Riyaz *et al*.[3] Many studies however showed a higher percentage of association.[10,13,19] Among patients with IBD (both had Crohn's disease), one had a peristomal PG, which developed seven months after ileostomy, and the other had the classic ulcerative variant. We had one case each of PAPA and PASH syndrome. Genetic studies were not done for the same. The different genes described in inflammasome formation include *MEFV*, *PSTPIP1*, *NLRP3*, *NLRP12*, *LPIN2*, and *NOD2*, which cause increased release of interleukin-1β, which has been documented in both syndromic and sporadic cases of PG.[20,21] The comparative data with other studies are depicted in Table 3.

In our study, even though 54 patients were suspected to have PG, only 17 patients could be recruited, as the majority did not satisfy the diagnostic criteria. This highlights the difficulty in labeling the diagnosis of PG, excluding the umpteen other causes of leg ulcers. A misdiagnosis rate of 10% was reported by Weenig *et al*.[22] in their study on 95 patients. A biopsy from the ulcer edge demonstrating neutrophilic infiltrate is the major criterion proposed by the Delphi consensus of international experts.[6,7] Histopathological findings in our study correlated with most of the other studies where dermal neutrophilia was noted in all patients. The infiltrate was polymorphous with neutrophil predominance in most of our patients (94.1%). Though controversial, leukocytoclastic vasculitis was observed in 4/17 (23.5%) patients in our study, similar to the findings recorded by Chakiri *et al*.[23] Granulomatous inflammation can also occur in PG, especially in cases with coexisting Crohn's disease.[24] Granulomas were noted in two cases in our study, among whom one had Crohn's disease and presented with peristomal PG. Necrosis was not a feature in any of the samples, though there are rare reports.[25]

There is insufficient evidence to clearly define the preferred therapeutic ladder for each PG variant.[20] In our cohort, 11/17 (64.7%) patients received either oral or parenteral steroids at presentation in tapering doses. The duration of steroid administration ranged from 2 weeks to 3.5 years, maximum dosage being 1 mg/kg/day equivalent to prednisolone. Systemic steroids were required for a prolonged duration in 4/10 patients (duration range: 1–3.5 years). In all cases, other steroid-sparing agents were also combined at the outset. Steroids were the

Table 3: Comparative data from other studies

Study and year	Country of study and study design	Duration (years)	Sample size (*n*)	Male (*n*)/ Female (*n*)	Mean age at presentation (years)	Diagnostic criterion used	Type of PG (*n*)	Systemic associations/co-morbidities (*n*)
Current study	India; R	10	17	8/9	34.9	Maverakis *et al.*[6]	Ulcerative – 9 Bullous – 4 Pustular – 3 Ulcerative and peristomal – 1	IBD (2), PAPA and PASH syndrome (1 each), seronegative arthritis (1), hemophilia A carrier state (1), anemia (3), diabetes (1), hypertension (2), and treated TB (2)
Schosler L *et al.*[12] (2021)	Denmark; R	6	64	29/35	63.4	Daniel Su *et al.*[5]	Ulcerative – 55 Bullous – 2 Peristomal- 4 Vegetative –2 Pustular – 1	Hematological diseases (5), IBD (18). cardiac disease (13), diabetes (16), malignancy (13), thyroid disease (8), other autoimmune diseases (8), renal disease (3), hidradenitis suppurativa (3), rheumatoid arthritis (8), gout (5), parkinsonism (2), and hemophagocytosis (1)
Riyaz *et al.*[3] (2017)	India; P	10	61	32/29	39.7	Clinical and histologic correlation, exclusion of other causes	Ulcerative – 55 Plaque – 6	IBD (10), arthritis (2), diabetes (2), and others (6)
Adışen *et al.*[9] (2015)	Turkey; R	12	27	10/17	53.2	Clinical and histologic correlation, exclusion of other causes	Ulcerative – 24 Others – 3	Behçet's disease (2), IBD (2), myelodysplastic syndrome (1), solid neoplasia (3), hypertension (9), diabetes (5) thyroiditis (4), hyperlipidemia (4), congestive heart failure (3), cervical radiculopathy (1), venous insufficiency (8), tuberculosis (1), chronic renal insufficiency (1), Meniere disease (1), deep vein thrombosis (1), anemia (14), depression (1), and hepatitis (1)
Ye and Ye[10] (2014)	Australia; R	10	23	7/16	62.8	Clinical and histological correlation, exclusion of other causes	Ulcerative – 18 Bullous – 2 Vegetative – 2 Pustular – 1	IBD (2), ankylosing spondylitis (1), hematological (2), solid neoplasia (5), CREST (1), and surgery (6)
Al Ghazal *et al.*[11] (2013)	Germany; P	1	259	117/142	58	Modified main and additional diagnostic criteria for PG	-	Arthritis (41), IBD (24), thyroid disease (29), diabetes (66), solid neoplasia (22), hematological (8), and CTCL (2)
Binus *et al.*[15] (2011)	USA; R	8	103	25/78	51.6	Clinical and histological correlation, exclusion of other causes	-	IBD (35), hematological malignancy (11), MGUS (10), arthritis (30), diabetes (29), and thyroid disorder (4)
Bhat *et al.*[13] (2010)	India; P	14	18	10/8	34.39	Clinical and histologic correlation, exclusion of other causes	Ulcerative – 17 Bullous – 1 Ulcerative and pustular – 2	Seronegative arthritis (4), ulcerative colitis (3), myeloproliferative disorder (2), and SLE (1)

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Table 3: Contd...

Study and year	Country of study and study design	Duration (years)	Sample size (*n*)	Male (*n*)/ Female (*n*)	Mean age at presentation (years)	Diagnostic criterion used	Type of PG (*n*)	Systemic associations/co-morbidities (*n*)
Mlika *et al.*[19] (2002)	Tunisia; R	20	21	11/10	41.8	Clinical and histologic correlation, exclusion of other causes	Ulcerative – 16 Bullous – 3 Vegetative – 2	IBD (5), hematological disease (4), Sjögren syndrome (1), and rheumatoid arthritis (1)
von den Driesch *et al.*[14] (1997)	Germany; R	10	44	14/30	50	Standardized major and minor diagnostic criteria	Ulcerative – 41 Bullous – 2 Vegetative – 1	Arthritis (5), IBD (6), solid neoplasia (5), hematological (3), and CTCL (1)

R=retrospective study; P=prospective study; IBD=inflammatory bowel disease; PAPA=pyogenic arthritis, pyoderma gangrenosum, and acne; PASH=pyoderma gangrenosum, acne, and suppurative hidradenitis; CREST=calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia; CTCL=cutaneous T-cell lymphoma; MGUS=monoclonal gammopathy of undetermined significance

mainstay of treatment in the studies by Al Ghazal *et al*., Riyaz *et al*., and Bhat *et al*.[3,11,13] However, both monotherapy and combination therapy are reported to be equally efficacious in the management of aggressive PG.[10,26] Ciclosporine is the most commonly used combination/steroid-sparing drug in many studies and is best documented in the literature.[27] In a Danish study on 64 patients with PG, the most common 2-drug therapy combination was prednisolone/methotrexate given to 37.5% of the patients, among whom 83.3% had complete remission, whereas the combination therapy of prednisolone with ciclosporine attained 100% remission in their study, with the mean time to remission being 13.9 months. However, the time to achieve remission was shortest for the combination of prednisone and the TNF-α inhibitor, infliximab.[12] The combination with ciclosporine was initiated in only two patients in our study who did not follow-up adequately.

In our cohort, only 1/3 patients who received adalimumab concomitantly with steroids showed ulcer resolution in less than three months, similar to the findings by Ye and Ye.[10] The other two patients were started on infliximab and azathioprine, respectively. One patient who did not show any response even after four doses of adalimumab, was switched over to infliximab with which she achieved remission. There are reports of thalidomide showing a good response in recalcitrant PG and genital PG and was utilized in 23.5% of our patients.[28,29] Thalidomide is an immunomodulator that modulates cytokine synthesis, decreases polymorphonuclear cell chemotaxis, and reduces tumor necrosis factor-alpha (TNF–α) levels, thus controlling neutrophilic inflammation. It also promotes the proliferation and migration of keratinocytes, thus enabling re-epithelialization.[28]

The recurrence rate was 23.5% in our study, relapse duration ranging from two months to one year. A higher recurrence rate of 33.3% was observed by Bhat *et al*.[13] and 39% was observed by Ye and Ye,[10] in contrast to our findings. None of our patients were considered for surgical treatment interventions considering the possibility of pathergy.

Limitations

Even though 54 patients in our study were suspected to have PG, only 17 patients could be labeled with the diagnosis of PG as we strictly adhered to the criteria, and this could also be due to the difference in diagnostic criteria used in various studies. Accordingly, we have used only descriptive statistics to analyze the data. As this was a retrospective study, there were lacunae in obtaining various details, assessing treatment efficacy, and follow up.

Conclusion

We would like to highlight that PG can affect any age group with even a juvenile onset and may be localized to rarer,

atypical sites such as the face or upper torso. Screening for syndromic associations is highly important, besides systemic associations like IBD. Prospective longitudinal studies and comparative therapeutic trials may henceforth help us in better understanding and management of PG.

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Conflicts of interest

There are no conflicts of interest.

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