Clinical Features and Treatment of Peristomal Pyoderma Gangrenosum

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Peristomal pyoderma gangrenosum (PPG), an unusual variant of pyoderma gangrenosum, has been reported almost exclusively in patients with inflammatory bowel disease (IBD) and is frequently misdiagnosed.

Objective To better characterize the clinical manifestations, diagnosis, and management of PPG.

Design, Setting, and Patients Retrospective analysis of 7 patients with PPG observed in a university-affiliated community setting between 1988 and December 1999.

Main Outcome Measures Clinical and histopathologic features, associated disorders, and microbiologic findings.

Results Two patients had Crohn disease, 2 had ulcerative colitis, and 3 had abdominal cancer. Five patients had at least 1 relapse of PPG after initial healing. Although 3 of 4 patients with IBD had active bowel disease, a parallel course with PPG occurred in only 1 patient. Both patients whose stoma was relocated developed an ulcer at the new site. Effective therapies included topical superpotent corticosteroids, intralesional injection of triamcinolone acetonide at the ulcer margin; topical cyclosporine, mycophenolate mofetil; and intravenous infliximab.

Conclusion Our experiences demonstrate that although PPG has been most often reported in patients with IBD, it may occur in the absence of IBD. Biopsy of the skin lesion is not diagnostic but excludes other causes. Relocation of the stoma may be associated with a new ulceration and should be avoided. Trauma to the skin of a predisposed patient may elicit the pustules or ulcerations associated with pathergy.

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Figure. Peristomal Pyoderma Gangrenosum Following Relocation of a Stoma

Patient 4 had a total proctocolectomy for ulcerative colitis 25 years prior to ulceration at the previous site of the ileostomy. A, This new ulcer developed 1 month after the relocation of her stoma and failed to respond to systemic antibiotics for a presumed wound infection. Note the ulcer’s violaceous, overhanging, scalloped border. B, Healing of the lesion in the same patient following therapy with oral dapsone.

Table. Clinical Characteristics of Patients With Peristomal Pyoderma Gangrenosum (PPG)

<table>
<thead>
<tr>
<th>Patient No./Sex/Age, y and Underlying Disease</th>
<th>Stoma Type</th>
<th>Time to PPG Onset After Surgery</th>
<th>Effective Therapies (Time to Healing)*</th>
<th>Ineffective Therapies*</th>
<th>Current Status of PPG</th>
<th>Other Medical History</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/F/72 Crohn disease</td>
<td>Total proctocolectomy with ileostomy</td>
<td>1 y</td>
<td>Topical clobetasol propionate (2 y); topical cromolyn sodium (1 y); infliximab (1 mo)</td>
<td>Sulfasalazine, metronidazole, triamcinolone acetonide</td>
<td>Mildly active</td>
<td>None</td>
</tr>
<tr>
<td>2/M/47 Crohn disease</td>
<td>Total proctocolectomy with ileostomy</td>
<td>13 y, and within 1 mo after stomal relocation</td>
<td>Dapsone, 200 mg/d (complete response for 2 episodes within 6 mo); cyclosporine, 4 mg/kg per day (4 mo)</td>
<td>Sulfasalazine, minocycline, prednisone (60 mg/d), topical clobetasol propionate, triamcinolone acetonide</td>
<td>Resolved</td>
<td>Deep vein thrombosis, prednisone-associated glaucoma, diabetes, cataracts</td>
</tr>
<tr>
<td>3/F/36 Ulcerative colitis</td>
<td>Total abdominal colectomy with retained rectal stump and ileostomy</td>
<td>2 mo</td>
<td>Partial response to prednisone, 20 mg/d, and cyclosporine, 5 mg/kg per day</td>
<td>Sulfasalazine, mesalamine, metronidazole, triamcinolone acetonide</td>
<td>Resolved</td>
<td>Axillary scarring due to presumed hydradenitis suppurativa, cushingoid facies</td>
</tr>
<tr>
<td>4/F/56 Ulcerative colitis</td>
<td>Total proctocolectomy with ileostomy</td>
<td>25 y, within 1 mo after stomal relocation</td>
<td>Dapsone, 100-150 mg/d (3 mo for 3 separate occurrences)</td>
<td>Prednisone (20 mg/d), topical clobetasol propionate</td>
<td>Resolved after third recurrence</td>
<td>Rheumatoidlike polyarthritis, pyoderma gangrenosum on the leg (10 y prior to peristomal lesion)</td>
</tr>
<tr>
<td>5/M/83 Bladder cancer</td>
<td>Urostomy</td>
<td>20 y</td>
<td>Triamcinolone acetonide, topical clobetasol propionate (6 wk)</td>
<td>None</td>
<td>Resolved</td>
<td>Hypertension, peptic ulcer disease</td>
</tr>
<tr>
<td>6/M/73 Bladder cancer</td>
<td>Urostomy</td>
<td>11 mo</td>
<td>Dapsone (11 mo for first occurrence), mycophenolate mofetil (8 mo)</td>
<td>Prednisone (20 mg/d), topical cromolyn sodium, clobetasol propionate</td>
<td>Still active</td>
<td>Steroid-induced diabetes, diverticulitis</td>
</tr>
<tr>
<td>7/M/64 Rectal adenocarcinoma</td>
<td>Proctosigmoidectomy with colostomy</td>
<td>22 y</td>
<td>Triamcinolone acetonide, topical clobetasol propionate (3 mo)</td>
<td>None</td>
<td>Resolved</td>
<td>Hypertension, gout, hyperlipidemia</td>
</tr>
</tbody>
</table>

*Triamcinolone acetonide is usually administered intralesionally as 3 mg/mL every 3 to 4 weeks.
Patient 4 had a lesion of classic pyoderma gangrenosum on her leg 10 years prior to the development of PPG and has had chronic polyarthritis. She had no colon or rectum at the time of either ulcerative lesion and has had no evidence of active IBD since her colectomy, yet PPG occurred. Three of the 4 patients with IBD had active bowel disease in association with the occurrence or recurrence of PPG. Patient 3 had a subtotal colectomy; following proctotectomy her lesions promptly healed and have not recurred for more than 3 years, unlike the situation in patient 4. In patients 1 and 2, systemic antibiotics that improved the intestinal symptoms did not lead to improvement of PPG. Patient 1 was treated with infliximab and had a prompt and sustained response of her bowel disease. Her PPG almost completely resolved after the third month’s infusion, but recurred within 4 weeks. Five patients have had multiple episodes of PPG.

Therapy was empiric and the responses varied (Table). All patients had been treated with 1 or more courses of broad-spectrum antibiotics for a presumed skin infection prior to the diagnosis of PPG. Topical clobetasol propionate (a class I topical corticosteroid) used in conjunction with intralesional (dermal) injection of triamcinolone acetonide was effective in 3 patients, ineffective in 2 patients, and possibly useful as adjunctive therapy in 2 patients. Topical 2% cromolyn sodium solution was effective in 1 patient and ineffective in another. Three patients required the addition of an immunosuppressive agent (cyclosporine [n=2] or mycophenolate mofetil [n=1]) because of systemic corticosteroid failure or adverse effects. Patient 4 had repeated responses to oral dapsone with each of her 3 relapses (Figure); in contrast patient 6’s initial episode responded to dapsone, but a relapse failed to respond.

COMMENT

Classic pyoderma gangrenosum is associated with systemic disease in half of such patients,1-5 whereas all of our small group of patients with PPG had a systemic disorder. Inflammatory bowel disease occurs in 15% to 20% of patients with classic pyoderma gangrenosum, and until recently accounted for almost all patients with PPG.11,12 Tumors of the colon, bladder, prostate, bronchus, ovary, breast, and adrenal gland have been associated with sporadic cases of classic pyoderma gangrenosum.2 Malignancies were the reason for stoma creation in 3 of our patients.

There was wide variability in the time from formation of the enterostomy/colostomy to the onset of PPG (2 months to 25 years).11-13 Dermatologists consider pathergy to be a process whereby, in susceptible persons, trauma to the skin results in pustules and/or ulcers. Pathergy may have functioned with the debridement, grafting, and relocation of the stoma in our patients to cause PPG. A seemingly insignificant degree of trauma related to irritation of the adhesive of the appliance or leakage of urine or feces may invoke pathergy, resulting in PPG in predisposed persons. Pathergy has been suspected in up to 30% of patients with pyoderma gangrenosum,2,5 and occurs in other neutrophilic dermatoses including Behçet disease, Sweet syndrome, and the blind loop syndrome.14

The PPG in 2 of our 7 patients responded to topical superpotent corticosteroids and intralesional injection of triamcinolone acetonide. However, the majority (6/7) required some form of systemic therapy, including dapsone, cyclosporine, mycophenolate mofetil, or infliximab. The latter agents allowed discontinuation of systemic corticosteroids in 4 patients with corticosteroid-induced diabetes mellitus, cataracts, glaucoma, or facial swelling.

The approach to treatment of patients with PPG is empiric. In patients with active IBD, measures to treat the underlying disease must be started along with conservative local wound care and topical application of a potent or superpotent corticosteroid. Patients without active bowel disease may try a topical corticosteroid. In cases that fail to respond, topical cromolyn sodium solution or tacrolimus ointment may be effective.11 After 2 to 4 weeks of local therapy without response, systemic therapy should be considered. Prednisone, 0.5 to 1.0 mg/kg per day, is standard therapy for pyoderma gangrenosum, whether peristomal or on other sites; however, we often consider other agents because of the adverse effects that commonly accompany use of systemic corticosteroids. A variety of agents can be tried, but our first-line therapy is dapsone for 4 to 6 weeks, then cyclosporine13 or mycophenolate mofetil.

Peristomal pyoderma gangrenosum may cause serious morbidity in patients who require placement of a stoma. The majority of our patients were initially misdiagnosed as having a skin infection, tending to confirm our belief that PPG is underreported.7,11

REFERENCES