Pyostomatitis vegetans (PSV)-pyodermatitis vegetans (PDV): A clinicopathologic study of 7 cases at a tertiary referral center

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Background: Pyostomatitis vegetans (PSV)-pyodermatitis vegetans (PDV) is a rare inflammatory mucocutaneous disease associated with inflammatory bowel disease.

Objective: We sought to evaluate the clinicopathologic findings of PSV-PDV in a series of 7 patients.

Methods: We conducted a retrospective review of all cases of PSV-PDV at the Mayo Clinic from 1995 to 2014.

Results: Seven patients with PSV-PDV were included, and all had inflammatory bowel disease. Three had Crohn's disease and 4 had ulcerative colitis. Three patients had peripheral blood eosinophilia. Two had concomitant pyoderma gangrenosum in which pyoderma gangrenosum lesions were recalcitrant to therapy. Primary sclerosing cholangitis was seen in 3 patients. Two patients had direct and 3 had indirect immunofluorescence findings. Tissue eosinophilia was seen in the majority of mucosal and cutaneous lesions.

Limitations: Limited sample size and retrospective study design are limitations.

Conclusions: PSV-PDV is associated with inflammatory bowel disease and primary sclerosing cholangitis and may precede gastrointestinal symptoms. Immunofluorescence findings in select PSV-PDV cases may indicate possible overlap with autoimmune bullous disease. Tissue eosinophilia may be helpful in distinguishing PSV-PDV from pyoderma gangrenosum. Strict control of bowel disease and close monitoring of patients with subclinical disease is warranted. (J Am Acad Dermatol 2016;75:578-84.)

Key words: Crohn's disease; inflammatory bowel disease; pemphigus; primary sclerosing cholangitis; pyoderma gangrenosum; pyodermatitis; pyostomatitis; ulcerative colitis; vegetans.

Pyostomatitis vegetans (PSV)-pyodermatitis vegetans (PDV) is a rare inflammatory mucocutaneous disease strongly associated with inflammatory bowel disease (IBD), particularly ulcerative colitis (UC).[1,2] The cutaneous manifestations of this disease are referred to as "pyodermatitis vegetans" and the oral form is termed "pyostomatitis vegetans," although most reports support the notion that they are virtually the same disease because they have similar histopathological features.[3]

Abbreviations used:

BMZ:	basement membrane zone
DIF:	direct immunofluorescence
IBD:	inflammatory bowel disease
PBE:	peripheral blood eosinophilia
PDV:	pyodermatitis vegetans
PG:	pyoderma gangrenosum
PSC:	primary sclerosing cholangitis
PSV:	pyostomatitis vegetans
UC:	ulcerative colitis

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The symptoms of PSV-PDV can be preceded by bowel involvement by several years; however, the skin and oral lesions of PSV-PDV may also be markers of subclinical disease.[4] It is typically a clinical diagnosis that can be supported by histologic findings.[5] Clinically, PSV is characterized by confluent pustules and erosions on mucosal surfaces with the typical appearance termed a "snail track," and PDV presents as exudative plaques that can involve the scalp, face, and intertriginous regions. Some reports suggest that peripheral blood eosinophilia (PBE) may accompany PSV-PDV.[5] Histopathological findings include epidermal hyperplasia, focal acantholysis, and a dense mixed inflammatory infiltrate with intraepithelial and subepithelial eosinophilic microabscesses.

Recently, it has been suggested that PSV-PDV may fall on a spectrum of diseases that can overlap with an IgA variant of pemphigus vegetans.[6] Herein, we present a survey of 7 patients with PSV-PDV and examine its correlation to IBD activity, concomitant cutaneous disorders, histopathologic and immunofluorescence findings, PBE, and therapeutic modalities.

CAPSULE SUMMARY

- Pyostomatitis vegetans (PSV)-pyodermatitis vegetans (PDV) is a mucocutaneous dermatosis associated with inflammatory bowel disease.
- PSV-PDV may be seen with primary sclerosing cholangitis and pyoderma gangrenosum and may herald subclinical bowel inflammation.
- Close monitoring of PSV-PDV disease activity may allow for better control of inflammatory bowel disease, whereas treatment of inflammatory bowel disease may ameliorate PSV-PDV.

METHODS

A retrospective study was conducted. After institutional review board approval, the Mayo Medical Center Research Database was queried for cases from May 1995 to November 2014. The terms "pyostomatitis," "pyodermatitis," and "pyoderma" were included. Data were collected from the medical charts of patients diagnosed with PSV in this time frame at the Mayo Clinic in Rochester, MN. Clinical notes with clinical descriptions, photographs, laboratory data, and histopathology specimens were reviewed by 2 board-certified dermatopathologists (M. J. C. and A. G. B.) to confirm the rendered diagnosis.

Eight cases of PSV-PDV were rendered during this time frame. One patient was excluded as no histopathology was available and the patient was not seen by a dermatologist. Patient characteristics at the time of diagnosis of PSV-PDV are shown in Table I. The onset of PSV-PDV, measured in months, was determined by the time frame between confirmed clinical diagnosis of IBD and patient descriptions of when oral symptoms began. Gastrointestinal symptoms were documented as "yes" or "no" depending on if the patient, at time of PSV diagnosis, had worsening of IBD. Remission of disease was determined by the time period between the start of therapy for PSV-PDV and a follow-up visit documenting no evidence of oral lesions on physical examination. The site of involvement of PSV-PDV was determined by clinical documentation of the physical examination and clinical photographs. Histopathology and immunofluorescence characteristics of mucocutaneous lesions are listed in Table II.

RESULTS

Clinical description or photograph was available for all 7 patients. In our cohort, all patients had PSV-PDV associated with IBD. Specifically, 3 of the patients had a previous diagnosis of Crohn's disease and 4 had UC. There were 5 men and 2 women with an average and median age of 36.9 and 30 years, respectively. All patients were Caucasian. In each case, the diagnosis of IBD preceded the diagnosis of PSV-PDV. The average and median time from the diagnosis of IBD to PSV-PDV was 11.8 and 9.3 years, respectively (average 141.6 months; median 111 months) (range 53.7-281.1 months). At the onset of oral symptoms, 71% of patients had active disease. The most common sites of oral involvement included the gingiva (5/7), buccal mucosa (4/7), and upper and/or lower mucosal lip (2/7). Confluent pustules were seen in all patients (Fig 1).

Four patients (57%) had cutaneous PDV lesions, 3 of whom had associated UC. Two patients had intertriginous involvement (1 groin only, 1 groin and axilla), whereas 2 patients had involvement of extremities and scalp. The cutaneous lesions demonstrated ulcerative/erosive papules and plaques with yellow crust (Fig 2). Interestingly, 3 patients (43%) also had primary sclerosing cholangitis (PSC), 1 having Crohn's disease and 2 having UC. Two patients (29%) also had pyoderma gangrenosum (PG). In 1 case, the PG and PSV started simultaneously; however, PSV resolved in 27 days whereas the PG was unresponsive to therapy. In the second case, PG resolved later than PSV.

Six patients had oral biopsy specimen findings available for review. Pseudoepitheliomatous/epithelial hyperplasia, intraepithelial neutrophils, and submucosal neutrophils were the most common histopathologic