# Pyodermatitis vegetans after total colectomy

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## ABSTRACT

Pyostomatitis–pyodermatitis vegetans (PPV) is a rare dermatological manifestation of inflammatory bowel disease, characterized by erythematous, vesiculopustular, exudative, annular, and/or vegetating plaques over the intertriginous regions that may precede or appear at the same time as the mucosal lesions. Systemic corticosteroids, dapsone, sulfasalazine, azathioprine, cyclosporine, and subtotal/total colectomy are the most common treatment options. A 16-year-old male patient presented to our outpatient clinic with a four months history of thickly crusted erosions on his trunk, back, and lower extremity. He had ulcerative colitis for four years and total colectomy was done seven months ago. Clinical and histopathological examination of his lesions were consistent with pyostomatitis vegetans. Although subtotal/total colectomy has been reported as a treatment option for PPV, lesions reappeared three months after total colectomy in our patient.

**Key words:** Inflammatory bowel disease, pyodermatitis–pyostomatitis vegetans, ulcerative colitis

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## INTRODUCTION

Pyostomatitis–pyodermatitis vegetans (PPV) is a rare inflammatory disorder of the mucous membranes and skin, which is usually accepted as a marker of inflammatory bowel diseases (IBDs), especially ulcerative colitis.[1] Although this entity is commonly seen with IBDs, it was also reported with sclerosing cholangitis, chronic hepatitis, and pericolangitis in the literature.[1,2] Mucosal lesions are known as "pyostomatitis vegetans" and characterized by edematous mucosa, multiple white-yellow pustules, vegetating hemorrhagic erosions, and ulcerations. Cutaneous lesions are known as pyodermatitis vegetans and characterized by white-yellow pustules, crusted papules, and plaques that coalesce into annular lesions and resemble "snail tracks" that mainly affect the major intertriginous surfaces and scalp.[1,3] Pemphigus vegetans and IgA pemphigus are the differential diagnoses clinically.[2] Histopathological examination and immunflorescence (which is typically negative) and history of IBD are helpful in confirming the diagnosis.

Mucosal and cutaneous lesions usually show the same histological features including intraepidermal and superficial dermal microabscesses containing neutrophils and eosinophil polymorphs and mixed deeper dermal inflammatory infiltrate.[1-4] Acanthosis, acantholysis, and pseudoepitheliomatous hyperplasia are commonly seen in the oral lesions.

Topical therapies are usually not adequate in the treatment of PPV. Systemic corticosteroid (0.5–1 mg/kg/d) is still the firstline therapy.[5] Systemic dapsone, azathioprine, cyclosporine, isotretinoin, infliximab, etanercept, methotrexate, and subtotal/total colectomy are other treatment options.[1-6]

We present a young male with thickly crusted erosions on sternum, shoulders, back, and lower extremities, which appeared after total colectomy surgery for ulcerative colitis.

## CASE REPORT

A 16-year-old male patient presented to our



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outpatient clinic with a 4 months history of thickly crusted erosions on his scalp, sternum, right shoulder, back, and bilateral lower extremity [Figure 1]. He had ulcerative colitis since 4 years and was treated with systemic corticoteroids for 2 years. Total colectomy was performed 7 months ago and systemic corticosteroid therapy was discontinued after colectomy. His skin esions appeared 3 months after the withdrawal of systemic corticosteroid therapy.

Two punch biopsy specimens and a direct immunflorescence specimen were taken from the margins of erosions on his right shoulder and sternum with initial diagnosis of pemphigus vegetans, pyodermatitis vegetans, acne conglobata, and pyoderma gangrenosum.

Histopathological examination of the biopsy from the right shoulder revealed nuclear debris, crusts, and irregular acanthosis in the epidermis, and dense capillary proliferation, plasma cells, eosinophils, and lymphocytic infiltrate in the dermis [Figure 2]. Hyperkeratosis, acanthosis, exocytosis in epidermis, edema, and dense lymphocytic infiltrate were seen in dermis in the specimen that was taken from sternum. Direct immunflorescence was negative for IgA, IgG, and C3. He was diagnosed as pyodermatitis vegetans based on his history, clinical findings, and histopathological examination. His lesions almost totally regressed within 1 month with systemic methylprednisolone 40 mg/d and topical mupirocin [Figure 3].

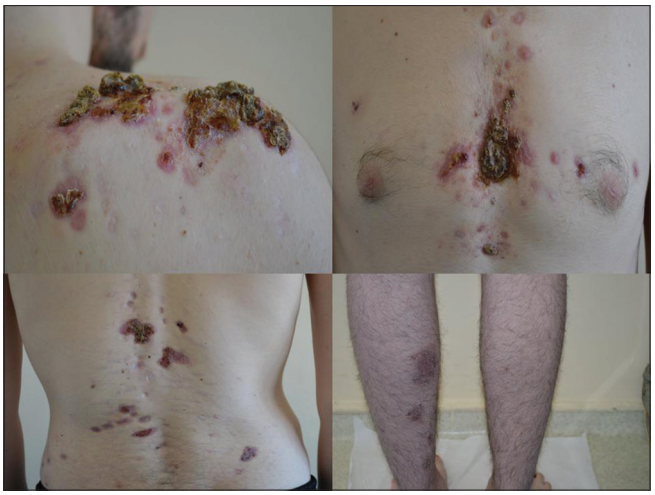

**Figure 1:** Hemorrhagic crusted erosions on sternum, right shoulder, back, and right leg

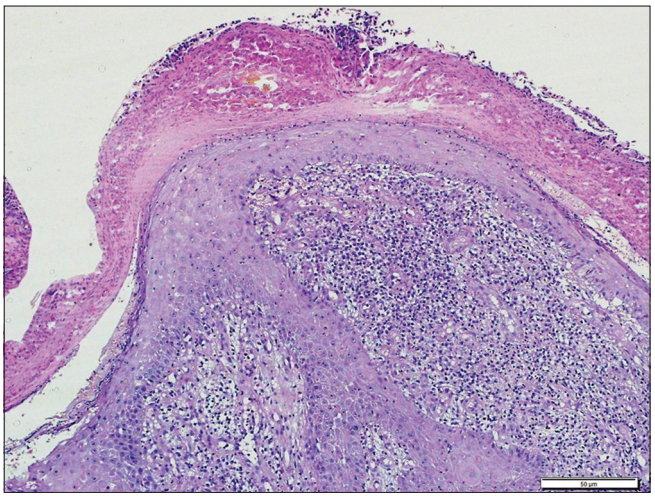

**Figure 2:** Crusts, irregular acanthosis in epidermis, dense capillary proliferation, and plasma cells, eosinophils and lymphocytic infiltrate in the dermis (H and E ×40)

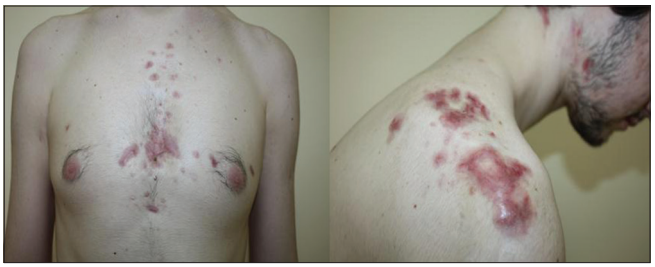

**Figure 3:** Almost complete reepithelization within 1 month with systemic methylprednisolone 40 mg/d

## DISCUSSION

Pyostomatitis–pyodermatitis vegetans (PPV) is a rare and well-known inflammatory dermatosis, which is usually associated with IBD.[1] It was first described in 1898 by *Hallopeau* in five patients who had oral pustular and cutaneous vegetating lesions.[2] Association of the pustular lesions with IBD was described by McCarthy *et al*. in 1949.[2] PPV is seen in adults after the third decade and more common among males (M:F = 3:1).[3] Our patient was a 16-year-old male, the youngest patient reported with this diagnosis.[2]

Erythematous, vesiculopustular, exudative, annular, vegetating plaques are the common cutaneous manifestations of PPV.[4] These features may however lead to confusion in the diagnosis. Konstantopoulou *et al.* reported a case report of PPV and ulcerative colitis in whom presentation was confused with acneiform lesions.[5] Although intertriginous surfaces are most commonly affected regions, extraordinary localizations have been reported in the literature.[6] Canpolat *et al.* reported a patient with pyoderma vegetans on the fingers of the left hand and the foot. Yasuda *et al.* reported a patient with erythematous, crusted, erosive and vegetating lesions of PPV on the mandible and lower lip that responded well to total colectomy and topical tacrolimus.[6,7] Leibovitch *et al.* reported a case report of pyodermatitis–pyostomatitis vegetans of the eyelid that completely healed with systemic steroids and sulfasalazine.[8] In our patient, lesions were located on shoulders, sternum, lower extremity and appeared 3 months after the total colectomy operation.

Systemic corticosteroids, dapsone, sulfasalazine, azathioprine, cyclosporine, and subtotal/total colectomy, and topical

tacrolimus are most preferred treatment options of PPV.[6,9] Although subtotal/total colectomy has been reported as a treatment option for PPV in the literature, lesions appeared after total colectomy surgery in our patient that responsed very well to systemic methylprednisolone within one month. *Kitayama et al*. reported a case similar to our patient who had PPV that appeared after subtotal colectomy.[10] Our case led us to consider that PPV could be a marker of continuing systemic inflammation.

The case is being reported for its rarity, young age of the patient, unusual localization of lesions and their occurence after total colectomy surgery.

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## Conflicts of interest

There are no conflicts of interest.

## REFERENCES

1. Cox NH, Coulson IH. Systemic disease and the skin. In: Burns T, Breathnach S, Cox N, Grifftihs C, editors. Rook's Textbook of Dermatology. 8th ed. United Kingdom: Wiley-Blackwell; 2010. p. 62.1-62.113.
2. Wang H, Qiao S, Zhang X, Liu C. A case of pyodermatitis-pyostomatitis vegetans. Am J Med Sci 2013;345:168-71.
3. Wolz MM, Camileri MJ, McEvoy MT, Bruce AJ. Pemphigus vegetans variant of IgA pemphigus, a variant of IgA pemphigus and other autoimmune blistering disorders. Am J Dermatopathol 2013;35:53-6.
4. Marzano AV, Borghi A, Stadnicki A, Crosti C, Cugno M. Cutaneous manifestations in patients with inflammatory bowel diseases: Pathophysiology, clinical features and therapy. Inflamm Bowel Dis 2014;20:213-27.
5. Kontsantopoulou M, O'Dwyer EM, Steele JC, Field EA, Lewis MA, Macfarlane AW. Pyodermatitis-pyostomamatis vegetans complicated by methicillin-resistant *Staphylococcus aureus* infection. Clin Exp Dermatol 2005;30:666-8.
6. Canpolat F, Cemil BÇ, Yılmazer D, Yeşilli O, Eskioğlu F. Pyoderma vegetans associated with ulcerative colitis: A case with good response to steroids. Case Rep Dermatol 2011;26:80-4.
7. Yasuda M, Amano H, Nagai Y, Tamura A, Ishikawa O, Yamaguchi S. Pyodermatitis-pyostomamatis vegetans associated with ulcerative colitis: Successful treatment with total colectomy and topical tacrolimus. Dermatology 2008;217:146-8.
8. Leibovitch I, Ooi C, Huilgol SC, Reid C, James CL, Selva D. Pyodermatitis-pyostomatitis vegetans of the eyelids case report and review of the literature. Ophthalmology 2005;112:1809-13.
9. Brikmeier T, Frosch PJ. Pyodermatitis-pyostomatitis vegetans: A clinical course of two decades with response to cyclosporine and low dose prednisolone. Acta Derm Venerol 2001;81:134-6.
10. Kitayama A, Misago N, Okawa T, Iwakiri R, Narisawa Y. Pyodermatitis-pyostomatitis vegetans after subtotal colectomy for ulcerative colitis. J Dermatol 2010;37:714-7.