# A Case of an Atypical Mycobacterial Ulcer Following an Intramuscular

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**Citation**

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**Abstract**

Buruli ulcer is a chronic, indolent, necrotizing disease of the skin and soft tissue. Buruli ulcer is the third most common mycobacterial disease of immunocompetent hosts, after tuberculosis and leprosy, and is caused by toxin-producing mycobacteria named Mycobacterium ulcerans. Over the last 2 decades, a re-emergence of cases has occurred, leading to the 1998 World Health Organization (WHO) Buruli Ulcer Initiative and the Fifty-Seventh World Health Assembly Resolution on Buruli Ulcer, which have stimulated ongoing research into diagnosis, pathogenesis, and effective treatment.

## INTRODUCTION

Pemphigus vulgaris (PV) is a relatively uncommon mucocutaneous autoimmune disorder which preferentially affects the oral mucosa, skin, and conjunctivae. Rarely seen, however, is extra-oral mucosal involvement. We present a patient with biopsy-proven PV involving the anal canal and a review of the literature.

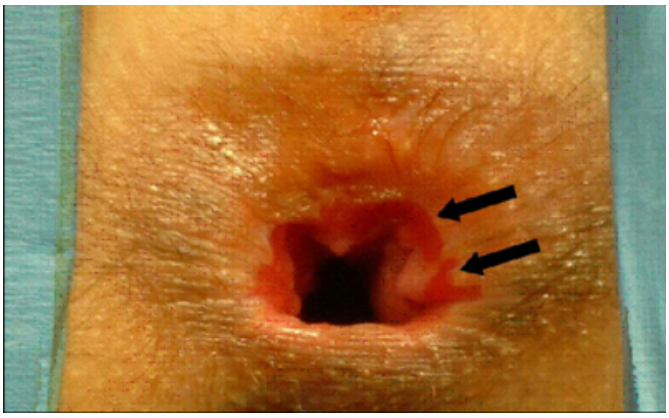
## REPORT OF A CASE

A 28-year-old female presented with a chief complaint of perianal irritation and painful bowel movements for two months and was referred for surgical evaluation. She had been diagnosed with oral pemphigus in 1998. She had no skin lesions, but her oral ulcers were steroid dependent. The patient had not tolerated a trial of Imuran. In August 2003 she developed pruritis ani and defecatory pain despite resolution of her oral lesion with steroids. She had undergone a trial of Diflucan for possible anal candidiasis without success. She had rare rectal bleeding, one soft bowel movement every one to two days, and normal continence to gas and stool. She denied a history or diarrhea or constipation.

The patient appeared well, with moon-like facies. Examination of her oropharynx revealed a 1cm mucosal ulcer. Examination of the anus and rectum under anesthesia demonstrated a circumferential denuded anoderm extending to the dentate line (Figure 1). The anal transition zone and the perianal skin were normal. Digital rectal examination was also normal. Multiple biopsies were obtained of the serpiginous ulceration and the margin between the ulceration and normal anoderm.

**Figure 1**

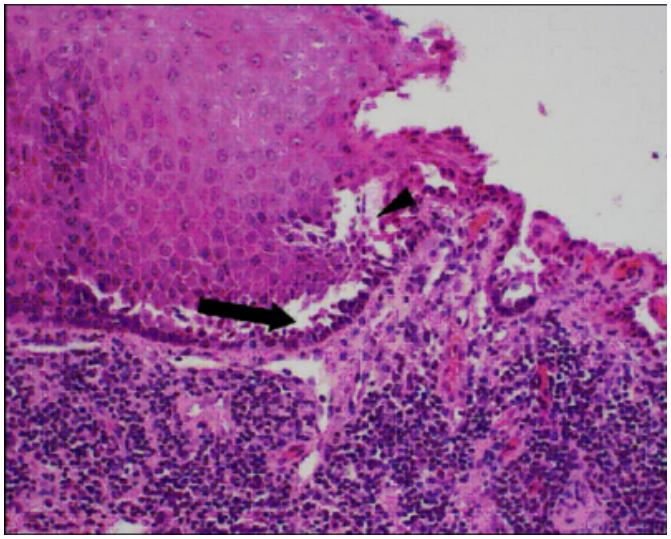
Figure 1. Intraoperative photograph of the anus with the patient in prone-jackknife position. There is circumferential, serpiginous ulceration of the anoderm (arrows)



Histologic examination revealed a strip of anoderm surface with stratified squamous epithelium (Figure 2). A suprabasilar acantholytic process was identified. The underlying connective tissue exhibited a patchy chronic inflammatory cell infiltrate composed of lymphocytes and plasma cells. The histopathology was consistent with PV. Direct immunofluorescent studies demonstrated localization of IgG antibody to the intercellular spaces of the suprabasilar epithelial cells and were negative for IgA, IgM and C3. These findings confirmed the diagnosis of PV.

**Figure 2**

Figure 2. Biopsy of anoderm demonstrating suprabasilar epithelial separation (arrow) and apoptotic keratinocytes are noted (arrowhead).(Hematoxylin and eosin; x180)



The patient was treated with an increased dose of oral corticosteroids as well as a course of mycophenolate mofetil to which she responded well. The anal lesions resolved completely within a few months and she has not had a recurrence at this site in five and a half years follow-up.

## DISCUSSION

PV is the most common and severe form of a rare group of potentially fatal autoimmune vesiculobullous diseases that affect mucus membranes and the skin. First described in 1791 by Wichman, PV has been reported in many countries, with an incidence ranging from 0.08 to 3.2 per 100,000 and a predilection towards people of Mediterranean and Ashkenazi Jewish descent (1-3). Although some studies show a slight female predominance, the disease generally affects men and women equally with a mean age of onset between 40 and 60 years (4-6). The gastrointestinal tract is an unusual site for PV manifestations and anorectal involvement is particularly rare. Most case series report no such findings and, in one of the largest studies published to date, rectal lesions were seen in only 2% of 1209 patients with PV (4). Malik et al. report a significantly higher incidence of anal involvement in their smaller cohort (10% of 160 PV cases) and propose that this may even be an underestimate due to misdiagnosis or underreporting (7). However, apart from these two studies, only four other cases suggestive of anorectal involvement have been previously documented and none with biopsy-proven diagnosis (8-11). To our knowledge, the case we describe here is one of the first with histopathologic confirmation.

While the precise etiology of PV is unknown, blistering is thought to result from circulating pathogenic IgG autoantibodies directed against desmoglein, a desmosome-associated glycoprotein strongly expressed in stratified squamous epithelium. Desmosomes constitute the primary adhesion structure of the epidermis and autoimmune targeting of its components can trigger a cascade reaction leading to the loss of this intercellular cohesion, known as acantholysis. In PV, this breakdown occurs just above the basement membrane (2, 3). This results in characteristic non-pruritic flaccid bullae that rupture easily leaving painful, weeping erosions (3). In 50-70% of cases, blistering begins in the oropharynx and nearly 100% of patients have oral lesions (1, 2). The disease usually progresses to the skin and then may remain localized or become widespread, involving up to 50% of the body surface in severe cases (3, 12). Cutaneous lesions generally include the upper trunk, head, neck, and intertriginous areas. Mucous membrane involvement can be found on any surface lined by stratified squamous epithelium, including the pharynx, larynx, conjunctiva, esophagus, anus, and genitalia (1, 3, 13). Patients present with a variety of systemic symptoms including weight loss (poor food intake due to painful oral lesions), malaise, epistaxis, dysphagia, and hoarseness when the larynx is involved (13).

The differential diagnosis of PV includes other autoimmune blistering disorders such as paraneoplastic pemphigus, pemphigus foliaceus, bullous pemphigoid, and dermatitis herpetiformis, as well as drug-induced, infectious and other acquired bullous diseases (2, 3,13). Diagnosis is based on four criteria: clinical findings, histologic evidence of suprabasilar intraepithelial acantholysis in affected tissue, direct immunofluorescence studies of perilesional tissue demonstrating intercellular deposition of IgG, and finally, the detection of circulating anti-Dsg autoantibodies using either indirect immunofluorescence or enzyme-linked immunosorbent assays (ELISA) (1,12,3).

Before the introduction of systemic corticosteroids in the 1950s, the mortality rate of PV was estimated to be 50% at 2 years, and nearly 100% at five years, usually due to mucocutaneous infection leading to sepsis (14,15). In the modern era, mortality has been dramatically reduced to less than 10%, with reported remission rates of 18-75% (6,12). There have been no large-scale controlled trials concerning management of PV and therefore, therapy is mainly empirical and highly variable. A recent guideline recommends initial treatment with prednisone, usually at a

dose of 1-1.5mg/kg/day (15), as higher doses have been found to increase morbidity with no therapeutic advantage (5). Adjuvant immunosuppressive drugs (azathioprine, cyclophosphamide, mycophenolate mofetil) are often used in addition for synergistic and steroid-sparing effects. However, a multi-modal approach is the key to successful management of anal PV as demonstrated by a 2006 review of 16 patients with widespread involvement including the anus. The authors report full recovery in this group using a combination of diet modification, anal hygiene protocols, and occasionally sublesional steroid injections in addition to the more traditional systemic and topical treatments (7).

## CONCLUSIONS

Pemphigus vulgaris is a crippling, potentially fatal mucocutaneous autoimmune disorder which preferentially affects the oral cavity, skin, and conjunctivae. Although anorectal involvement has been suggested in up to 10% of affected patients, this case represents one of the first documented instances of biopsy-proven anal disease. Patients with PV are frequently immunosuppressed and at risk for competing infectious and neoplastic anorectal pathologies. Therefore, in order to avoid misdiagnosis and expedite proper treatment, biopsy of suspected lesions is recommended.

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## References

1. Feldman (2002) Sleisenger & Fordtran's Gastrointestinal and Liver Disease, 7th ed. Saunders, Inc. 343, 631-2.
2. Keehn CA, Morgan MB. (2004) Clinicopathologic attributes of common geriatric dermatologic entities. Dermatol Clin 22: 115-23.
3. Sami N, Wei Yeh S, Ahmed AR. (2004) Blistering diseases in the elderly: diagnosis and treatment. Dermatol Clin 22: 73-86.
4. Chams-Davatchi C, Valikhani M, Daneshpazhooh M, et al. (2005) Pemphigus: analysis of 1209 cases. Int J Dermatol 44: 470-6.
5. Fernandes NC, Perez M. (2001) Treatment of pemphigus vulgaris and pemphigus foliaceus: experience with 71 patients over a 20 year period. Rev Inst Med Trop S Paulo 43(1) 33-6.
6. Scully C, Paes De Almeida O, Porter SR, Gilkes JJH. (1999) Pemphigus vulgaris: the manifestations and long-tern management of 55 patients with oral lesions. Br J Dermatol 140: 84-9.
7. Malick M, El Tal AE, Ahmed AR. (2006) Anal involvement in pemphigus vulgaris. Dis Colon Rectum 49 (4): 500-6.
8. Epstein JH, Feigen GM, Epstein NN. (1958) Pemphigus vulgaris with lesions of the rectal mucosa. AMA Arch Dermatol 78: 36-8.
9. Haskell B, Rovner H. (1958) Pemphigus of the Anal Canal. Dis Colon Rectum 1(6): 439-42.
10. Stone DD. (1971) Rectal lesions and toxic dilatation of the colon in a case of pemphigus vulgaris. Am J Dig Dis 16: 163-6.
11. Yoshida K, Takae Y, Saito H, et al. (2005) Cutaneous type pemphigus vulgaris: a rare clinical phenotype of pemphigus. J Am Acad Dermatol 52(5): 839-45.
12. Harman KE, Albert S, Black MM. (2003) Guidelines for the management of pemphigus vulgaris. Br J Dermatol 149: 926-37.
13. Bickle KM, Roark TR, Hsu S. (2002) Autoimmune bullous dermatoses: a review. Amer Fam Phys 65 (9): 1861-70.
14. Anhalt GJ, Diaz LA. (2004) Pemphigus vulgaris — a model for cutaneous autoimmunity. J Am Acad Dermatol 51(1): S20-1.
15. Mimouni D, Nousair CH, Cummins DL, Kouba DJ, David M, Anhalt GJ. (2003) Differences and similarities among expert opinions on the diagnosis and treatment of pemphigus vulgaris. J Am Acad Dermatol 49(6): 1059-62.

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