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Pyodermatitis-pyostomatitis vegetans: a case report and review of literature

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Abstract

Pyodermatitis-pyostomatitis vegetans is a rare inflammatory dermatosis. There is a strong association between pyodermatitis-pyostomatitis vegetans and inflammatory bowel disease, particularly ulcerative colitis. Herein, we report a case of pyodermatitis-pyostomatitis vegetans with positive direct immunofluorescence staining findings and review the literature for the past 18 years to characterize the disease, its epidemiologic characteristics, its associations, and the pathology and direct and indirect immunofluorescence findings. The total number of cases was 38, including 22 men and 16 women, with an average age of forty. Direct immunofluorescence staining had been performed for 32 patients, of which 12 had positive findings. Of those with positive direct immunofluorescence, 6 patients showed IgA cell surface staining. A recent approach suggests that these immunological findings may not be accidental and indicates a possible overlap with autoimmune bullous diseases discussed in this review.

Keywords: pyodermatitis-pyostomatitis vegetans, inflammatory bowel disease

Introduction

Pyodermatitis-pyostomatitis vegetans is a mucocutaneous dermatosis and oral lesions are characterized by multiple yellow or white pustules that may rupture, leaving erosions and a characteristic 'snail track' appearance. The involvement of skin surfaces usually includes exudative or crusted vegetating plaques on flexural areas [1, 2]. Histological findings include epidermal hyperplasia, dense mixed inflammatory cell infiltrate in the underlying dermis, and intraepithelial and subepithelial eosinophilic and neutrophilic microabscesses. Acantholysis, if present, is usually focal and mild [3]. Herein we report a case of PPV with direct immunofluorescence study suggestive of IgA pemphigus and review the literature of the past 18 years to characterize the disease, its epidemiologic characteristics, associations, pathology, and direct and indirect immunofluorescence studies.

Case Synopsis

A 32-year-old woman presented with a 2-month history of multiple crusted plaques on her scalp, back, and lower abdomen. She had a 16-year history of Crohn disease, which was under control with

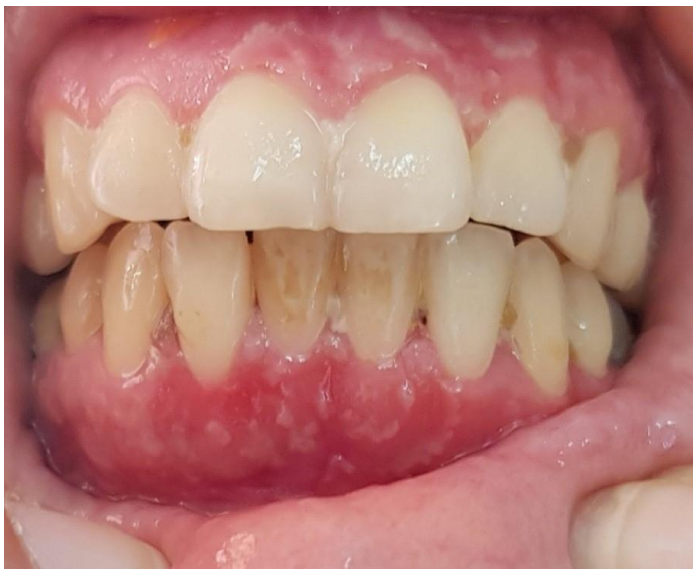

Figure 1. *Gingival erythema with overlying confluent pustules arranged in serpentine lines.*

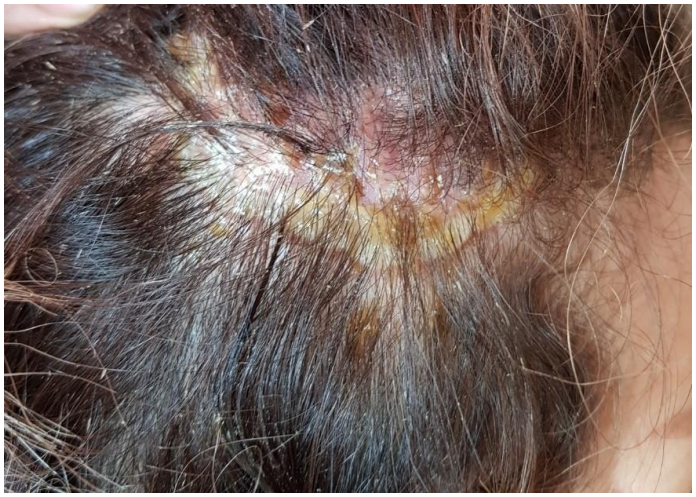

Figure 2. *A large well-defined annular erosive vegetative plaque with overlying yellow crust.*

mesalazine and azathioprine 150mg daily. She had been taking her medication without any interruption and had a well-documented compliance history. The patient also had a history of painful oral sores for about one year prior to the presentation of the cutaneous lesions. She had been previously received various antibiotics including ciprofloxacin and cephalexin for her cutaneous lesions, without an adequate therapeutic effect. Physical examination revealed gingival erythema with overlying confluent pustules arranged in serpentine lines (**Figure 1**). On the scalp, there was a large well-defined annular erosive vegetative plaque with overlying yellow crust that extended throughout the vertex (**Figure 2**). Vegetative plaques were present on her back, lower abdomen (on the site of caesarian section scar), and medial side of her left arm; these lesions had an active erosive border with central crusting. There were also multiple separate pustules on her back. No other abnormal findings were noted on her physical examination.

Laboratory assessment showed an absolute eosinophil count of 2100 cells/mm3 without leukocytosis; viral serologies for HIV and hepatitis viruses as well as autoimmune antibody studies were all negative. There was also microcytic hypochromic anemia, hemoglobin of 10.1g/dl, and serum iron 44µg/dl. She had a past medical history of minor β thalassemia. Fecal occult blood tests were positive. Tzanck smear was negative for herpes simplex virus.

Histopathology of the biopsy taken from the pustular border of her abdominal lesion showed epidermal hyperplasia with some acantholysis and mixed epidermal and dermal inflammation with numerous eosinophils (**Figure 3**). Direct immunofluorescence showed a +2 intercellular epidermal deposition of IgA without any deposits of IgG and C3 (**Figure 4**). Intraepithelial and subepithelial microabscesses containing numerous eosinophils as well as neutrophils and a moderate mixed inflammatory cell infiltrate in the underlying dermis were found on histology of the oral lesions.

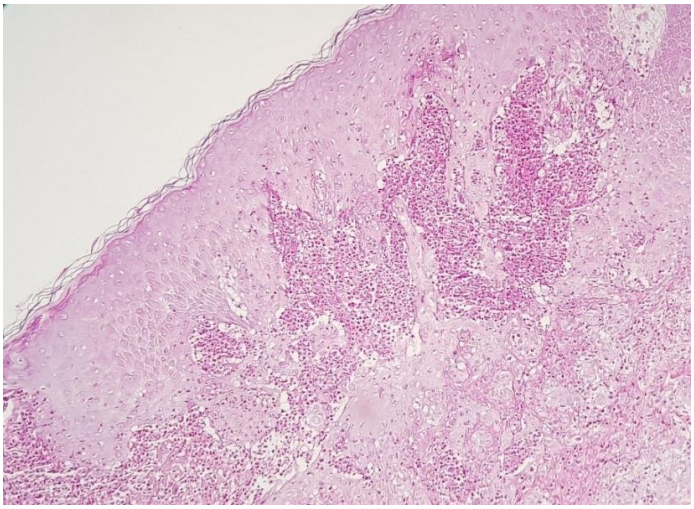

Figure 3. *Epidermal hyperplasia with some acantholysis and mixed epidermal and dermal inflammation with numerous eosinophils. H&E, 100×.*

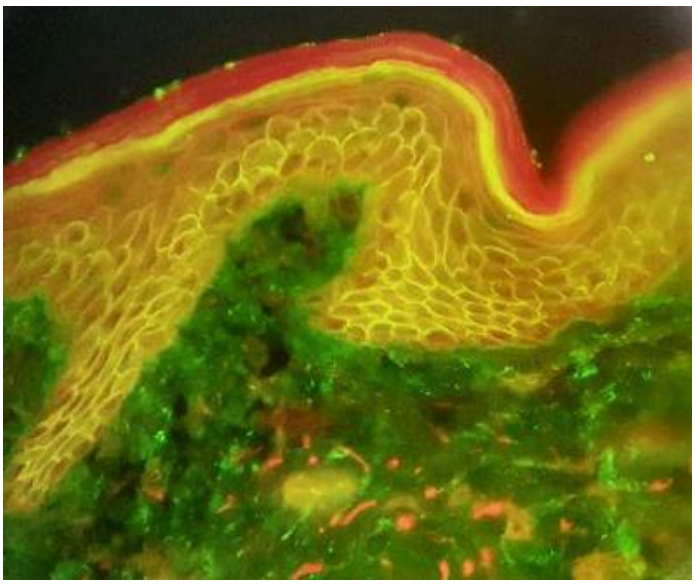

Figure 4. *A +2 intercellular epidermal deposition of IgA without any deposits of IgG and C3, 400×.*

Based on the patient's underlying condition, clinical and histopathologic features, which included the presence of oral lesions and their characteristic 'snail track' appearance, acantholysis, and mixed inflammatory cell infiltrate with prominent eosinophils, the diagnosis of PPV was made. Therefore, treatment with clobetasol 0.05% lotion once daily and clobetasol ointment twice daily for scalp and truncal lesions, respectively, was initiated leading to a marked improvement of the cutaneous lesions within 7 days. However, because the patient was still distressed by painful oral lesions, oral dapsone 25mg once daily was added to the therapeutic regimen. After two months of treatment with dapsone 25mg daily the mucosal lesions and symptoms had a significant improvement.

Case Discussion

Pyodermatitis-pyostomatitis vegetans is a rare mucocutaneous dermatosis. There is strong association between PPV and IBD, particularly ulcerative colitis[4, 5]. The most important entity in the differential diagnosis of PPV includes autoimmune blistering disorders such as pemphigus vulgaris, particularly its pemphigus vegetans subtype, IgA pemphigus, epidermolysis bullosa acquisita, and dermatitis herpetiformis. These entities are usually differentiated based on their clinical and histopathological features. Negative direct immunofluorescence is of high value for distinguishing PPV from these autoimmune blistering diseases [3].

There have been 3 reviews on pyostomatitis vegetans patients between the years 1979 and 2011 including 37 cases, all of them stating that negative immunofluorescence distinguishes PPV from pemphigus [6-8]]. In some of these studies; the cases with positive immunofluorescence were considered as pemphigus vegetans and therefore excluded from the study [5, 6, 9]. However, in many other cases, these direct immunofluorescence findings including C3, IgG, and IgA deposits at the basal membrane zone or epithelial cell surfaces have been often explained as the secondary response to epithelial damage, rather than the actual cause of the disease [9]. In our case, direct immunofluorescence examination showed a 2+ intercellular IgA deposition suggesting a diagnosis of IgA pemphigus. However, the existence of multiple eosinophils forming intraepidermal and dermal abscesses and moreover the typical manifestations of the patient's oral lesions ('snail track' appearance), absence of pruritis, and association with Crohn disease were highly suggestive of PPV. In IgA pemphigus mucous membranes are usually spared, cutaneous lesions are highly pruritic, and association with Crohn disease is much less common [4].

Considering the fact that in more recent studies the number of cases subjected to immunofluorescence studies and therefore positive immunofluorescence findings have considerably increased, a new approach is suggested. The immunological findings in these cases may not be a false-positive finding corroborating the hypothesis that PPV may be a part of a spectrum of diseases, including pemphigus vegetans and IgA pemphigus. The presence of immunological depositions may play a causative role in the disease pathogenesis [1, 9, 10]. This new interesting theory has led us to review the literature once again to study more recent cases with more focus on the IF information.

Therefore, what is the explanation for negative direct immunofluorescence findings in a certain number of patients? One possible explanation could be that most cases of PPV occur in the setting of an underlying disease, mostly IBD for which patients are usually on immunosuppressive or immunomodulating medications that can result in negative findings. In our study at least 11/19 patients with negative direct immunofluorescence findings were found in the presence of immunosuppressive or immunomodulators for their underlying conditions before PPV diagnosis and immunofluorescence staining.

Although PPV often presents in IBD patients who have recently stopped medications or are functionally untreated, our patient was under treatment with mesalazine and azathioprine daily. Despite the fact that it is documented that the clinical course of PPV usually parallels the activity of IBD, in this study more than half of the patients with

IBD did not have gastrointestinal symptoms at the onset of the mucocutaneous lesions [3]. However, lack of gastrointestinal symptoms, does not rule out IBD activity. Activity assessment of IBD is based on a combination of symptoms, clinical and endoscopic findings, and biomarkers such as C reactive protein and fecal calprotectin. Therefore, assessing the correlation between PPV and IBD activity could not be fully investigated owing to lack of laboratory and endoscopic findings in a majority of cases [11].

Depending on the existence and severity of the underlying disease a variety of therapeutic strategies have been used in the treatment of PPV. These include corticosteroids, both systemic and topical, which are considered one the most common and effective treatment options in many cases [3]. Although it is documented that antimicrobials, rather than corticosteroids or antineutrophil therapies, are indicated if the cause is uncertain [4], there are a few case reports showing the resolution of lesions mainly based on antibiotic therapy. This may highlight the possible role of microbial agents in PPV pathogenesis [1, 12]. Other reportedly successful treatments include topical tacrolimus, dapsone, azathioprine, mycophenolate mofetil, cyclosporine, and tumor necrosis factor inhibitors such as adalimumab and infliximab. When associated with ulcerative colitis subtotal/total colectomy are other treatment options since the clinical course of PPV tends to parallel that of ulcerative colitis [1, 2, 13].

We performed a review of pyodermatitis-pyostomatitis vegetans cases indexed in PubMed detailed in case reports and case series between 2001 and 2018. To be included in this review, articles had to be available in the English language, and the diagnosis of pyodermatitis-pyostomatitis vegetans had to be confirmed by skin biopsy. The terms ''pyostomatitis,'' ''pyodermatitis,'' and ''pyoderma'' were included. Patients without cutaneous lesions were excluded from the study. Age, sex, pathology, and treatments were recorded in all patients. Localization of the lesions were categorized into two groups of oral and skin lesions. Direct and indirect immunofluorescence studies as well as peripheral blood eosinophilia and the presence of gastrointestinal symptoms at the onset of cutaneous lesions, if available, are listed in **Table 1**. Demographics features and lesion locations are recorded in **Table 2**.

Histopathology of lesions showed epidermal hyperplasia in 25/38 cases. A total of 32/38 had inflammatory infiltration in their biopsy specimens, dominantly composed of intraepithelial and dermal neutrophils and eosinophils, whereas 24/31 of cases had variable degrees of eosinophilic tissue infiltration. Acantholysis was present in 12/38 cases and was mainly mild, suprabasilar, and focal.

Direct immunofluorescent staining had been performed for 32 patients, of which 20 showed negative results and 12 had positive findings. Of the 12 with positive direct immunofluorescent, 6 patients showed IgA cell surface staining of which one also showed IgG and C3 at basement membrane zone and IgG cell surface staining. Only three patients had epithelial cell surface staining with IgG and C3. One patient showed linear IgG basement membrane zone staining and one only had mild granular C3 at the dermoepithelial junction. Indirect immunofluorescent staining was available in 14 patients of which more than half (8/14) showed negative results. In our review, 65% of patients had an antecedent, coincident or subsequent history of IBD (70% of those with IBD had ulcerative colitis).

A correct and early diagnosis of PPV is necessary for a thorough investigation of the patient for IBD and other associated medical conditions which may require prompt treatment. Moreover, the correct diagnosis of PPV can aid in choosing the best therapeutic approach leading to the best outcome with the least side effects.

Conclusion

Herein, we report PPV in a 32-year-old woman with Crohn disease and review the literature on this rare entity from 2001 to 2018. Based on this study we suggest that PPV is a diagnosis primary based on clinical features including the presence of characteristic oral findings, a strong association with IBD, and vegetative cutaneous lesions. Although immunofluorescence examination can be of value in diagnosing this disorder, the diagnosis cannot

merely depend on it. We hope that these data aid in the future studies to clarify this confusing clinical entity and its pathogenesis.

Potential conflicts of interest

The authors declare no conflicts of interests.

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Table 1. *Pyostomatitis-pyodermatitis vegetans case reports since 2001, Unavailable data are recorded as N/A.*

	No.	Age	Sex	Localization	Pathology	DIF	IIF	GI symptoms	PBE	Other Medical Conditions	Rx
Gheisari et al. 2018	1	32	F	Oral: + Skin: Scalp, Trunk, Extremities	Skin: Epidermal hyperplasia, acantholysis and mixed epidermal and dermal inflammation with numerous eosinophils	+2 Intercellular epidermal deposition of IgA without any deposits of IgG and C3	N/A	–	+	CD	sCS Dapsone
Nayak et al. 2017 [14]	2	33	F	Oral: + Skin: Intertriginous, Extremities, Trunk	Epidermal hyperplasia, moderate spongiosis, suprabasal cleft with multiple intraepithelial abscesses with numerous eosinophils and neutrophils	-	-	+	-	UC	sCS
Maseda et al. 2017 [15]	3	49	M	Oral: - Skin: Extremities	Epidermal hyperplasia, neutrophilic infiltration	N/A	N/A	N/A	N/A	CD	Ciprofloxacin 500 mg BD Clarithromycin 250mg BD
Dodd et al. 2017 [13]	4	30s	F	Oral: + Skin: Scalp Intertriginous, Trunk	Epidermal acanthosis, spongiosis, acantholysis, eosinophilic pustular inflammation and mixed dermal infiltrate	Linear C3 and IgG deposition along the BMZ	-	-	N/A	CD	Infliximab Intralesional Triamcinolone Dapsone
Keitley et al. 2017 [16]	5	54	F	Face	Acantholysis, spongiosis. neutrophilic infiltration within the epidermis and dermis. Intradermal eosinophils, Intraepithelial abscesses within eccrine glands	N/A	N/A	-	+	CD Ileal adenocarcinoma	Ciclosporin Prednisolone lymecycline 0.3% tacrolimus in clobetasol 17-propionate ointment

Clark et al. 2016 [3]	6	30	M	Oral: + Skin: Scalp	Oral: Epithelial hyperplasia with intraepithelial and submucosal neutrophils and eosinophils Skin: ulceration, epidermal hyperplasia, subcorneal neutrophilic pustule, intraepidermal eosinophils, papillary dermal neutrophils and eosinophils	N/A	N/A	-	+	UC PG PSC	Nystatin sCS Dapsone Azathioprine Intralesional corticosteroids Petrolatum
	7	44	F	Oral: + Skin: Extremities Trunk, Scalp	Skin: Subcorneal neutrophilic pustule, scattered intraepidermal and dermal neutrophils and eosinophils	-	N/A	-	+	UC PG	sCS tCS Petrolatum
	8	21	M	Oral: + Skin: Intertriginous	Oral: focal epithelial ulceration, epithelial hyperplasia, acantholysis, intraepithelial and subepithelial clefting and mixed inflammation with numerous neutrophils Skin: pseudoepitheliomatous hyperplasia, focal acantholysis, intraepidermal and dermal neutrophils and eosinophils	Cell surface IgA deposition	IgG and IgA cell surface antibodies	+	+	UC PSC Psoriasis	Dapsone tCS
	9	58	M	Oral: + Skin: Intertriginous	Oral: epithelial hyperplasia with focal suprabasilar acantholysis, intraepithelial	Cell surface IgG,IgA & complement C3 & BMZ IgG & complement	IgG and IgA cell surface antibodies IgG BMZ antibodies with	-	-	CD	sCS Dapsone tCS Ketoconazole

					neutrophils and eosinophils and submucosal neutrophils and eosinophils	C3 deposition	epidermal pattern				
Dupuis et al. 2016 [17]	10	48	M	Oral: + Skin: Trunk, Extremities, Face	Skin: mixed inflammatory infiltrate, intraepidermal microabscess formation, subepidermal collections of eosinophils and neutrophils	Mild granular staining for C3 at the dermoepidermal junction	N/A	-	N/A	Indeterminate colitis	sCS
Carvalho et al. 2016 [18]	11	78	F	Skin: Trunk, Extremities, Genital	Skin: Pseudoepitheliomatous hyperplasia with dense inflammatory infiltrate composed of neutrophils and eosinophils in the upper dermis	-	-	-	+	-	Oral amoxicillin/ clavulanic acid 875/ 125mg BD Prednisolone 40mg/day
Yorulmaz et al. 2015 [19]	12	67	M	Skin: trunk and extremities Oral: N/A	Pseudoepitheliomatous hyperplasia with dermal inflammatory infiltrate mainly comprising neutrophils, eosinophils, focal suprabasal acantholysis	-	N/A	-	N/A	Asthma CD	Adalimumab, sCS tCS
Mansouri et al. 2015 [20]	13	11	F	Oral: + Skin: Face	Skin: Pseudoepitheliomatous hyperplasia and intraepithelial or dermal microabscess composed of eosinophils, eosinophilic spongiosis, mixed dermal infiltration	-	N/A	-	+	PCID recurrent HSV	sCS not good response

Uzunçakmak et al. 2015 [21]	14	16	M	Oral: - Skin: Scalp, Trunk, Extremities	Acanthosis, plasma cells, eosinophils, and lymphocytic infiltrate in the dermis	-	N/A	-	N/A	UC	sCS
Stingeni et al. 2015 [22]	15	17	M	Oral: + Skin: Scalp Face, Trunk,	Acantholytic blisters, Intraepidermal micro abscesses and mixed inflammatory cell infiltrates with eosinophils in the surrounding derma.	Weak linear IgG and granular C3 deposits along the BMZ	N/A	-	+	UC	Prednisone Azathioprine Mesalazine
KAJIHARA et al. 2013 [23]	16	78	M	Oral: - Skin: Scalp, Trunk, Extremities	Oral: Skin: erosion, ulcer and irregular epidermal hyperplasia, Dermal inflammatory infiltration composed mostly of eosinophils and neutrophils	-	N/A	-	-	MM	sCS Colchicine Topical tacrolimus
Wolz et al. 2013 [9]	17	21	M	Oral: + Skin: Intertriginous	Acantholysis, intraepidermal & dermal/epidermal separation, and mixed dermal inflammation with numerous neutrophils	IgA cell surface staining	Positive for cell surface IgA antibodies on monkey esophagus	+	N/A	UC	tCS Dapsone
	18	58	M	Oral: + Skin: Intertriginous	Suprabasilar acantholysis, intraepidermal pustules, subepithelial pustules	IgA cell surface staining discontinuous strong linear IgG deposition along the bmz with weak staining of epithelial cell surfaces.	Positive for cell surface IgA antibodies on monkey esophagus (1:160) and for cell surface IgG antibodies with an epidermal pattern on	+	N/A	CD	Dapsone Prednisone

						Discontinuous C3 staining of the bmz.	human salt-split skin				
Mesquita KC, Costa IMC. 2012 [24]	19	12	M	Oral: + Skin: Anogenital	Oral and Skin: Suprabasal acantholytic, mixed inflammatory infiltration with eosinophils	-	N/A	-	-	-	sCS Azathioprine Dapsone
Abellaneda et al. 2011 [10]	20	38	M	Oral: + Skin: Anogenital, Trunk, Extremities, Scalp	Epidermal acanthosis with spongiosis, PMN leukocyte exocytosis, intraepidermal and dermal mixed inflammatory infiltrates with abundant neutrophils and eosinophils	Positive (IgA)	Positive (weak)	+	+	UC	Prednisone, Mesalazine, Mercaptopurine
Canpolat et al. 2011 [25]	21	64	M	Oral: - Skin: Extremities	Pseudoepitheliomatous hyperplasia with a dense inflammatory infiltrate composed of neutrophils in the upper dermis	-	N/A	+	N/A	UC	Scs Amoxicillin-clavulanate Aluminium subacetate dressing Sulfasalazine Metronidazole
Moloney et al. 2011 [26]	22	50	F	Oral: + Skin: Face	Mixed inflammatory cell infiltrate with prominent eosinophils and neutrophils	-	N/A	+	+	UC	Azathioprine, Mesalazine, Dapsone
Matias et al. 2011 [27]	23	47	F	Oral: - Skin: Intertriginous , Face, Trunk	Epithelial hyperplasia, focal subepidermal cleft, mixed dermal inflammatory infiltrate predominantly eosinophilic with papillary eosinophilic abscesses and eosinophilic spongiosis.	-	-	-		-	Prednisone

Kitayama et al. 2010 [28]	24	51	F	Oral: + Skin: Anogenital Face	Acanthosis, dense mixed dermal inflammatory infiltration consisted mainly of eosinophils and neutrophils	-	-	+	-	UC	Cyclosporine, Azathioprine, Prednisone
Adişen et al. 2009 [29]	25	74	M	Oral: - Skin: Extremities	Skin: pseudoepitheliomatous hyperplasia, fibrosis, vascular proliferation, and a mononuclear predominant infiltration with occasional neutrophils in dermis	N/A	N/A	-	-	-	sCS
Ko et al. 2009 [30]	26	47	M	Oral: + Skin: Anogenital Trunk	Skin: Epidermal hyperplasia with microabscess formation consisting of eosinophils and neutrophils	-	N/A	-	N/A	-	Methylprednisolone
	27	24	F	Oral: N/A Skin: Scalp	Skin: epidermal hyperplasia with intraepithelial eosinophilic microabscesses	-	N/A	-	N/A	-	Methylprednisolone
Yasuda et al. 2008 [31]	28	37	M	Oral: + Skin: Face Scalp	Skin: epidermal hyperplasia, dense neutrophilic and eosinophilic inflammation with microabscesses involving the dermal papillae and epidermis.	-	N/A	+	+	UC PSC Colon cancer	Total colectomy, Topical tacrolimus

Harish et al. 2006 [32]	29	35	M	Oral: - Skin: Face, Trunk	Hyperkeratosis, irregular acanthosis. Subepidermal micro-abscess containing neutrophils and eosinophils superficial and deep dermal inflammatory infiltrate composed mainly of neutrophils.			+	N/A	UC	tCS cCS Mesalamine, Metronidazole
Konstantopoulou et al. 2005 [33]	30	19	M	Oral: + Skin: Scalp Trunk Intertriginous	Skin: intense infiltrate of lymphocytes, eosinophils and plasma cells, thinning of the epidermis, and a subepidermal bulla	N/A	N/A	+	+	UC Sclerosing cholangitis	Metronidazole Prednisolone Dapsone Hydrocortisone butyrate emulsion
Leibovitch et al. 2005 [34]	31	29	M	Oral: + Skin: Trunk Intertriginous Face Extremities	Skin: epidermal hyperplasia, eosinophilic dermal abscesses, eosinophilic exocytosis into the epidermis. Subtle focal suprabasal acantholysis, mixed dermal inflammatory infiltration	-	-	-	+	UC	Prednisone

Ahn et al. 2004 [35]	32	33	F	Oral: N/A Skin: Trunk Anogenital Face Scalp	Skin and oral: Epidermal hyperplasia and intraepidermal splitting, with predominant neutrophilic and eosinophilic infiltrates, and a few acantholytic cells. Dense mixed dermal inflammatory cell infiltrates, microabscesses consisting of neutrophils, eosinophils, and acantholytic cells within the epidermis and dermal papillae.	+ linear deposition of IgG along the BMZ	+ using salt-split skin showed IgG deposition on the epidermal side at 1:320 dilution	-	-	UC	Prednisone, Dapsone, Colchicine
RIEDERJ et al. 2004 [36]	33	42	M	penis	Epithelial hyperplasia and extensive dermal inflammation with neutrophils, lymphocytes and eosinophils	N/A	N/A	N/A	N/A	HIV Recurrent HSV	Circumcision
Nigen et al. 2003 [6]	34	23	F	Oral: + Skin: Intertriginous Extremities Anogenital	pseudoepitheliomatous hyperplasia with intraepithelial microabscesses and eosinophilic spongiosis	-	-	-	N/A	-	tCS Prednisone
	35	57	F	Oral: + Skin: Intertriginous Extremities Anogenital	Eosinophilic spongiosis with suprabasal clefts and a few acantholytic cells Dermal inflammatory infiltrate with predominance of eosinophils	-	N/A	-	+	-	Prednisone tCs

Brinkmler et al. 2001 [1]	36	23	M	Oral: + Skin: Trunk Intertriginous extremities	hyperkeratotic and acanthotic epidermis with intraepidermal abscess formation. A dense perivascular cell infiltrate was composed of histiocytes, a few plasma cells and eosinophils	-	N/A	-	-	-	Prednisolone, Cyclosporine Isotretinoin
Papadopoulos et al. 2001 [12]	37	24	F	Oral: - Skin: Extremities	Skin: pseudoepitheliomatous hyperplasia, acanthosis, hyperkeratosis, neutrophilic infiltration in the stratum corneum	-	-	-	-	highly elevated serum IgE level Hidradenitis suppurativa	Intravenous Imipenem–cilastatin 500 mg qid for 8 days, Whirlpool therapy, Local wound care
Bianchi et al. *2001* [37]	38	48	F	Oral: - Skin: Extremities, Trunk	Skin: Pseudoepitheliomatous hyperplasia, intraepidermal and subepidermal neutrophilic microabscesses, dense superficial and deep dermal inflammatory infiltrate composed mainly of neutrophils, lymphocytes and, eosinophils	-	-	+	N/A	UC	Intravenous piperacillin Mesalazine, Metronidazole

Table 2. *Demographics and lesion locations.*

	Number (percentage)
Gender	
Male	22
Female	16
IBD	25 (66%)
UC	18 (72%)
CD	7 (28%)
GI Symptoms at disease onset	17 (46%)
Cutaneous involvement	38
Trunk	18
Extremities	14
Intertriginous	13
Scalp	13
Anogenital	11
Face	11
Mucosal involvement	26
Gingiva and buccal mucosa	11
Mucosal lips	8
Tongue and palates	6