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Prurigo Pigmentosa - A Multi-institutional Retrospective Study

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**Title:** Prurigo Pigmentosa - A Multi-institutional Retrospective Study 1 Authors: Amy Shen BS<sup>1</sup>, Carol E. Cheng MD<sup>1</sup>, Rhea Malik MS<sup>2</sup>, Erica Mark BS<sup>3</sup>, Natalia 2 3 Vecerek MD<sup>4</sup>, Nolan Maloney MD<sup>5</sup>, Joan Leavens MD<sup>6</sup>, Vinod E. Nambudiri MD MBA EdM<sup>2</sup>, 4 Art P. Saavedra MD PhD<sup>3</sup>, Marcia Hogeling MD<sup>1\*</sup>, Scott Worswick MD<sup>4\*</sup> 5 6 Affiliations 7 <sup>1</sup>Division of Dermatology, Department of Medicine, David Geffen School of Medicine at 8 UCLA, Los Angeles, CA 9 <sup>2</sup>Harvard Medical School, Department of Dermatology, Brigham and Women's Hospital, 10 Boston, MA 11 <sup>3</sup>Department of Dermatology, University of Virginia Medical Center, Charlottesville, VA 12 <sup>4</sup>Department of Dermatology, University of Southern California, Los Angeles, CA <sup>5</sup>Department of Dermatology, Stanford University, Palo Alto, CA 13 14 <sup>6</sup>Lake Washington Dermatology and Dermatopathology, Kirkland, WA 15 16 \*Joint senior author 17 18 **Corresponding author:** Amy Shen, BS 19 10833 Le Conte Ave, Los Angeles, CA 90095 20 21 Phone: 209-556-8123 22 Email: shuishen@mednet.ucla.edu 23 24 Funding sources: None 25 26 Conflicts of Interest: None 27 28 Patient consent: Not applicable 29 30 IRB approval status: Reviewed and approved by 31 UCLA IRB#: 21-000840 32 Mass General Brigham IRB#: 2022P001488 33 University of Virginia IRB: #HSR220101 34 35 Manuscript word count: 494 References: 5 36 37 Figures: 0 38 Supplementary figures: 0 39 Tables: 1 40 Supplementary tables: 0 41 42 Keywords: prurigo pigmentosa; general dermatology; medical dermatology; retrospective study; 43 ketogenic diet; Sjogren's; connective tissue disease; lupus; comorbidities; treatment; papules; 44 papulovesicles; diabetes; hyperlipidemia; rash; topical steroids, antibiotics; minocycline; 45 doxycycline; autoimmune; reticulated hyperpigmentation, distribution

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Prurigo pigmentosa (PP) is an inflammatory skin disease characterized by a sudden eruption of pruritic, erythematous papules in a reticular pattern followed by hyperpigmentation [1]. In recent years, PP has been anecdotally associated with the ketogenic diet [2]. However, studies on PP have been limited to small case series. Our retrospective study examined the largest pool of PP patients to date from three academic centers in the United States over a ten-year period to identify potential triggers, comorbidities, and treatment.

We searched dermatology records from the University of California, Los Angeles, the University of Virginia, and the Mass General Brigham Hospital System from 2011-2021 using the search term "prurigo pigmentosa." We identified 30 patients with a confirmed diagnosis of PP; demographic and clinical factors were abstracted.

Of the 30 patients, 21 were female (Table 1). The average age was 41 years old, and the median age was 37 (range: 18-74 years). The majority of patients were white (56%), followed by Asian (30%). 29 patients had a BMI > 25. All patients presented with erythematous papules coalescing into plaques with a background of reticulated hyperpigmentation with or without scale, and all patients endorsed pruritus. The most commonly affected sites were back (46%) and chest (43%). 40% (12/30) of the patients were on a ketogenic diet prior to the onset of symptoms. Three patients were prediabetic, and none were diabetic. Eleven patients had hyperlipidemia. None of the patients had autoimmune conditions. Histopathological findings for PP were subtle and non-specific --- common features of the 13 available records were very mild spongiosis and a predominantly lymphoplasmacytic perivascular and interstitial infiltrate. Neutrophils and eosinophils were rare. All patients received treatment. Topical corticosteroids (12/30) only provided temporary relief, while oral antibiotics (11/30) led to complete resolution in all treated patients.

Our study supports the classic presentation of PP as documented in the literature and finds pruritus to be a constant feature. Most individuals with PP across the institutions were either white or Asian females, consistent with demographics reported in the literature [3]. PP did not seem to be associated with autoimmune conditions, despite previous reports detailing a likely association [4]. Atopic diathesis has been postulated to be associated with PP [5]. Our data did not see a strong association of PP with the three most common atopic conditions. Our finding of higher BMI in nearly all PP patients hasn't been reported elsewhere and may be worth exploring in future studies.

Based on our results, we did not clearly identify a unifying, common trigger of PP, suggesting different etiologies not limited to the ketogenic diet. Biopsy may assist in ruling out other diagnoses but is not diagnostic of PP on its own. PP affects a wide range of ages and both genders, with a female predilection. Furthermore, a course of oral antibiotics (doxycycline or minocycline 100mg twice daily for 1-2 months) may be considered in PP cases where transition to normal diet or topical treatment did not lead to complete resolution.

 Table 1. Clinical and demographic characteristics of individuals with prurigo pigmentosa (n=30)

	Number of patients, n (%)		Number of patients, n (%)
Demographics	<b>T</b> , (,	Comorbidities	1 ()
Gender		Hyperlipidemia	11 (36)
Female	21 (70)	Hypothyroidism	3 (10)
Male	9 (30)	H. Pylori	1 (3)
Race	,	Allergic Rhinitis	1 (3)
White	17 (56)	Asthma	0 (0)
Asian	9 (30)	Diabetes Mellitus	0 (0)
Black	2 (6)	Hyperthyroidism	0 (0)
Other	2 (6)	Pregnancy	0 (0)
Other	2 (0)	Sjogren's	0 (0)
Morphologies		Lupus	0 (0)
Hyperpigmentation	30 (100)	Dermatomyositis	0 (0)
Papules	30 (100)	Scleroderma	0 (0)
Scaling	15 (50)	Rheumatoid Arthritis	0 (0)
Scaring	13 (30)	Mixed Connective-	0 (0)
Cumptoms		-Tissue Disease	0 (0)
Symptoms Pruritus	20 (100)	-11ssue Disease	0 (0)
Pain	30 (100)	Chin anasifia samauhiditias	
raiii	0 (0)	Skin specific comorbidities	2 (10)
D'-4'14'		Atopic dermatitis	3 (10)
Distribution	12 (42)	Acne vulgaris	3 (10)
Chest	13 (43)	Allergic contact dermatitis	3 (10)
Back	14 (46)	Psoriasis	2 (6)
Upper extremities	3 (10)	Pityriasis rosea	1 (3)
Lower extremities	2 (6)	Prurigo nodularis	1 (3)
		Telogen effluvium	1 (3)
Diet			
Ketogenic diet	12 (40)	Treatment	
Low carbohydrate diet	3 (10)	Topical corticosteroids	12 (40)
Normal diet	15 (50)	Triamcinolone 0.1%	6 (20)
		Betamethasone 0.05%	3 (10)
BMI		Hydrocortisone 2.5%	1 (3)
≤18.5	0 (0)	Fluocinolone 0.025%	1 (3)
18.5-24.9	1 (3)	Clobetasol 0.05%	1 (3)
25.0-29.9	21 (70)	Oral doxycycline	6 (20)
$\geq$ 30.0	8 (26)	Oral minocycline	5 (16)
		Topical ketoconazole	2 (6)
		Topical tacrolimus	1 (3)
		Oral prednisone	1 (3)
		Transition to normal diet	3 (10)

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