Follicular Psoriasis

Weill Cornell Medicine
Dermatopathology

Background

Psoriasis is a common autoimmune dermatosis representing an interplay between certain genetic predisposing factors along with clonally restricted Th1 T cells responding to epidermal keratinocyte derived antigen. A unique interleukin 17 interleukin 23 cytokine rich milieu is pathogenetically significant and conducive to its salient histomorphologic features such as epideram hyperplasia and intraepidermal influx of neutrophils. Many of the effector intraepidermal lymphocytes are of the CD8 subset; myeloid dendritic cells are also key players in the evolution of psoriasis (1). The worldwide prevalence of psoriasis is age and location dependent. In the US, the incidence of psoriasis is approximately 2%, with a higher frequency in the Caucasian population (2). The classic cutaneous manifestation is that of plaque psoriasis also referred to as psoriasis vulgaris. Characteristic lesions are described as elevated well-circumscribed erythematous plaques covered by silvery scales that exhibit the Auspitz's sign upon removal. Follicular psoriasis (FP) is an uncommon variant manifesting as a scaly folliculocentric hyperkeratotic eruptions of the trunk and extremities, irrespective of the presence or absence of conventional lesions of psoriasis vulgaris. The precise incidence of FP is unknown with roughly 23 reported cases emphasizing its rarity given the overall incidence of conventional psoriasis in the general population. Due to the lack of awareness, the clinical presentation is often misdiagnosed as other follicular dermatoses such as bacterial folliculitis, pityriasis rubra pilaris, keratosis pilaris, and follicular eczema.

Case Presentation

A 61-year-old white Caucasian female presented with a sudden and self-remitting pruritic papular rash of the bilateral forearms and bilateral lower legs of several months duration. She denied any constitutional symptoms. The patient had no known history of kidney, liver, thyroid, and hematological diseases. Other family members were not afflicted with a similar eruption. Physical examination revealed numerous erythematous crusted papules that extended to the superior and inferior back. Symptomatic palliation was seen with the use of a pimecrolimus cream (Elidel)(figure 1). Multiple shave biopsies were performed on the right superior back, right inferior back, right upper arm, and right forearm. Light microscopic examination revealed an extensive pustular folliculocentric process. There was permeation of the outer root sheath epithelium by neutrophils. In addition, the ostium of the follicle was occluded by keratin intimately admixed with neutrophils(figure 2a, 2b). The background epidermis exhibited classic features of a psoriasiform diathesis, the hallmarks being those of neutrophil imbued parakeratosis, granular cell layer loss and dilated dermal papillae capillaries lying in intimate apposition to the basal layer of the epidermis (figure 2c, 2d). Direct immunofluorescence revealed entrapment of immunoglobulin and complement within the stratum corneum along with variable granular deposition of complement along the dermal epidermal junction. A diagnosis of follliuclar psoriasis was made. The patient was placed on a tumor necrosis factor inhibitor which resulted in regression of the eruption. It was subsequently established that the patient had a remote history of psoriasis .

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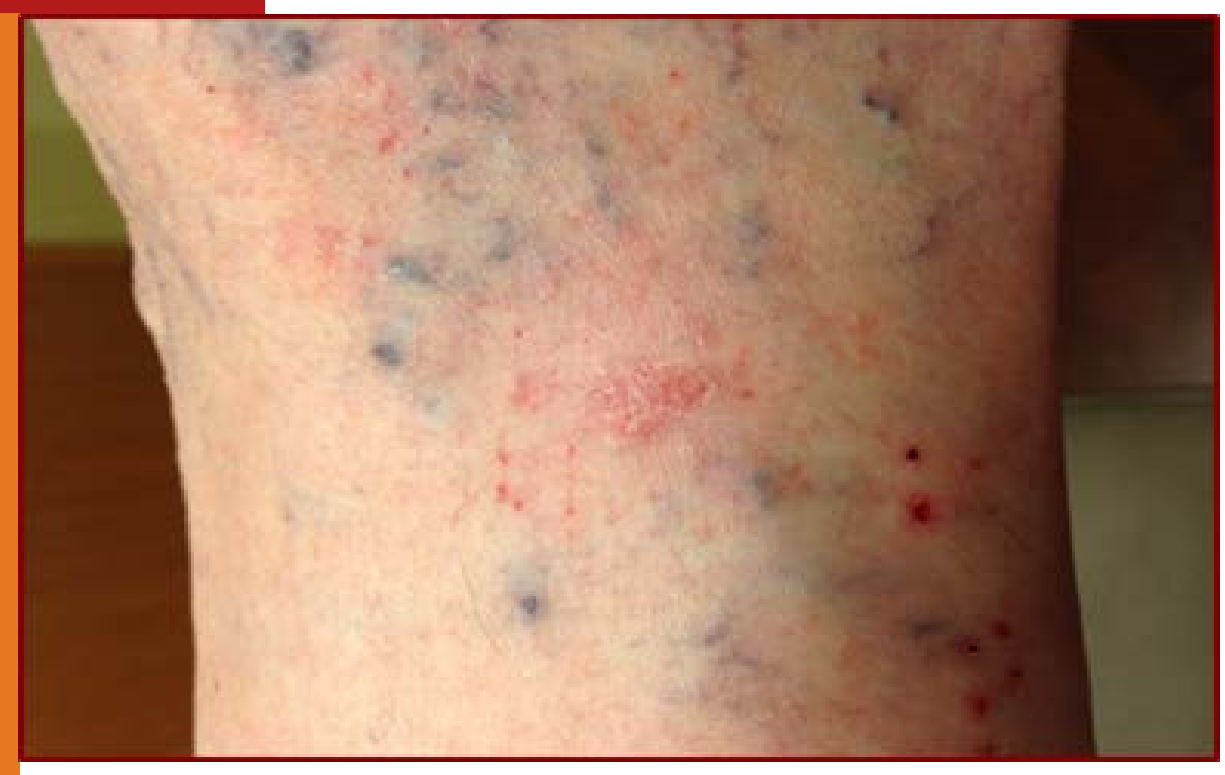

Discussion

We have presented a patient with diagnostic features of FP in the setting of an antecedent history of psoriasis vulgaris. Expectedly our patient demonstrated an excellent response to biologic therapy. Our case appears to be prototypic for this rare variant of psoriasis based on our review of the literature. From a historical perspective the first paper making reference to follicular psoriasis was in a paper published by Michelson in 1958 as the unusual in psoriasis; the reported cases describing this follicular presentation of psoriasis were in children. However the first paper recognizing FP as a distinct entity was first made by Stankler and Ewen. The authors subdivided FP into two broad categories : the adult type and juvenile type (3). The adult group was characterized by a total of 6 patients, 5 females and 1 male ranging in age from 18 to 69 years of age. All patients had a prior diagnosis of psoriasis . This group presented with bilateral follicular lesions on the thighs in a background of more conventional lesions of psoriasis vulgaris. The juvenile group comprised a total of 4 patients, 1 female and 3 males below the age of 10 years. A prior diagnosis of psoriasis under the age of 10 was rendered in all 4 patients . This group presented with asymmetric plaque-like follicular lesions on the trunk and axilla. In the series of FP by Ploysangam and Mutasim, a total of 5 patients, 4 females and 1 male, ranging in age from 23 to 73 years had clinical symptoms of erythematous hyperkeratotic follicular papules on the trunk and extremities (4). Only two patients had a prior history of psoriasis, both exhibiting a peculiar localization of psoriasis to the scalp. There appeared to be an association with diabetes mellitus and being of African ancestry. There are several additional anecdotal case reports describing FP. In particular Arps et al. described a 46-year-old diabetic African American woman with symptomatic pruritus with folliculocentric hyperkeratotic papules on the scalp, neck, back, and extremities (5). This patient had no previous diagnosis of psoriasis and lacked the classical findings for psoriasis vulgaris. Thomas et al. presented a case of FP in an 18-year-old Asian male who exhibited bilateral follicular lesions involving the thighs, calves, and arms. This patient was initially diagnosed with lichen planopilaris (6). Patil et al. described a 13-year-old boy who had symptomatic scaling and erythema of the skin that began as horny papules on the knees and elbows (7). These lesions evolved into erythematous scaly papules encompassing the entire body.

The clinical presentations and microscopic examinations in all of the previously described reports were similar to the findings in our case. Our case represents the 23th reported case of FP. A summary of these 23 cases is presented in table I.

The basis for the folliculocentricity of this variant of psoriasis is unclear. Secondary progressive involvement of the integument in patients with an established history of conventional psoriasis seems plausible given the document history of pre-existing psoriasis in most patients and the well known predilection of psoriasis for those areas of the skin devoid of follicles most notably palms, soles, and nails. Perhaps a further explanation lies in inherent tropism of common viruses such as varicella, measles, polyomavirus, human hyerpes type VI and human herpes type VII for the hair follicle. The innate immune response involves the recruitment of plasmacytoid and myeloid dendritic cells into the infected follicles a source of type I interferon which enhances both the adaptive and innate limbs of immunity. Hence the follicle could have a microenvironment conducive to the development of the psoriatic lesion given the known role of myeloid dendritic cells, type I interferons and T cells in the pathogenesis of the earlier phases of psoriatic inflammation.

Figure Legend

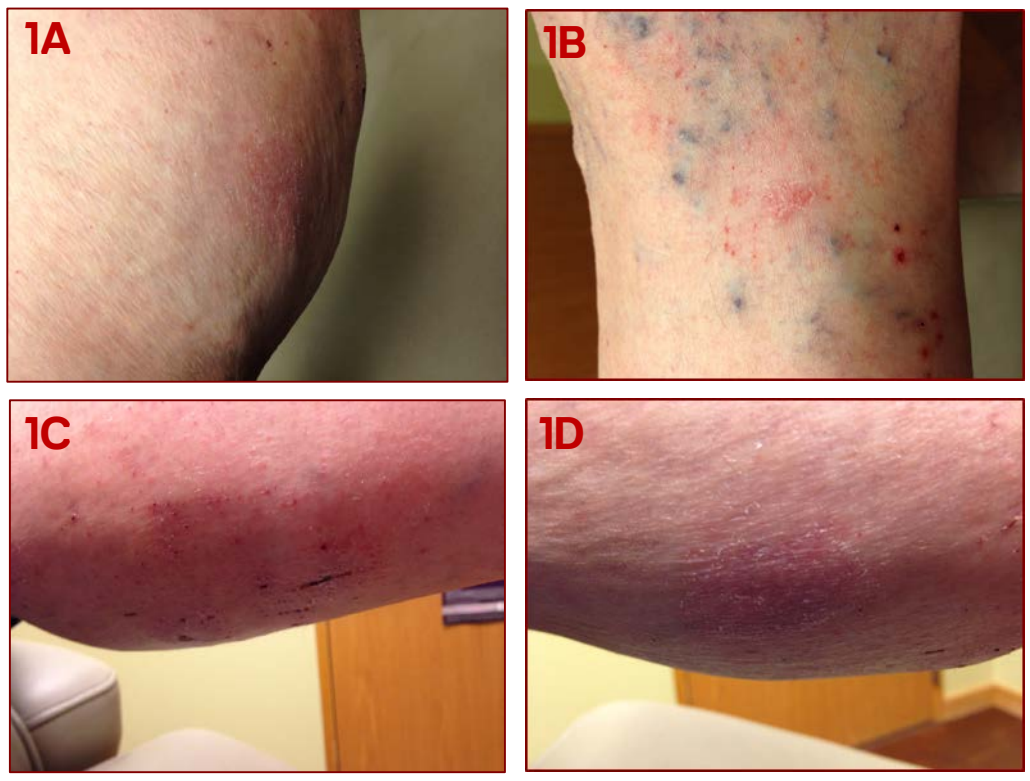

Figure 1A-D: Erythematous hyperkeratotic follicular papules of the arms and legs.

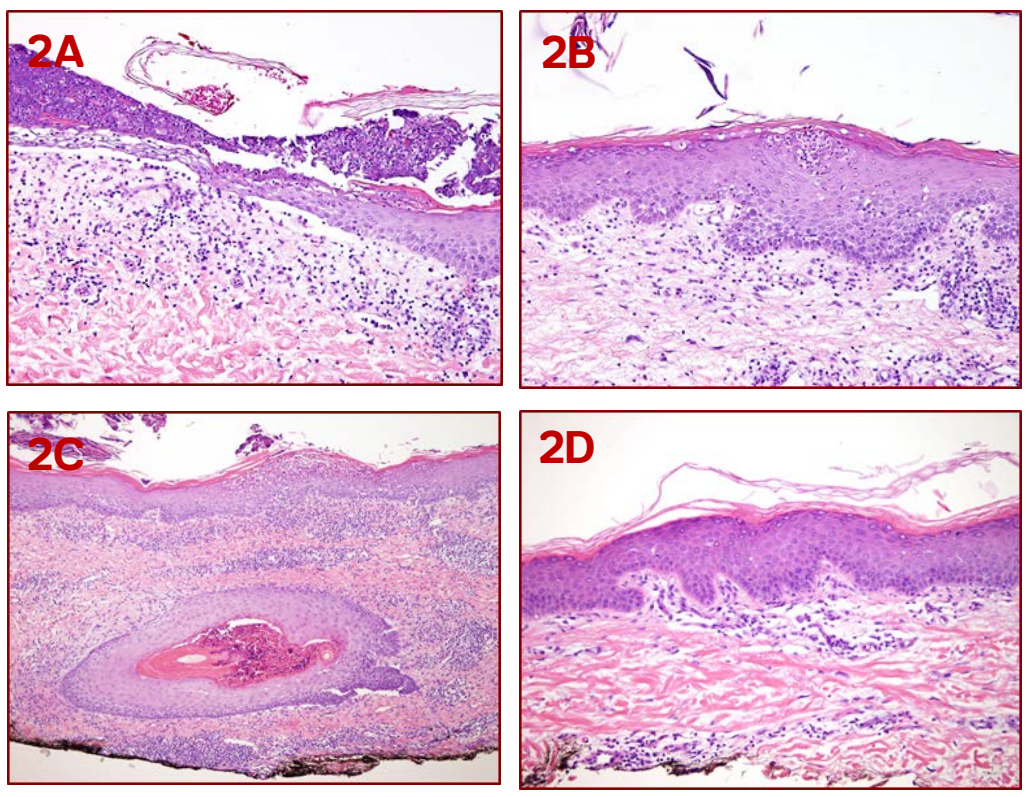

Figure 2A-D: A. 20x The biopsy showed a pustular folliculocentric process. B. Higher power magnification reveals marked infiltration of the follicular ostium by neutrophils intimately admixed with compact keratin. C. The interfollicular epidermis revealed subtle features of a psoriasiform diathesis revealed by lenticular shaped foci of neutrophil imbued parakeratosis and D. characteristic dilated capillaries lying in intimate apposition to the basal layer of the epidermis.

Under the direction of Dr. Cynthia M. Magro, the Weill Cornell Comprehensive Dermatopathology Service is a leading edge consultation service and CAP-accredited laboratory for dermatologists, plastic and general surgeons and other dermatopathologists. Dr. Magro is an internationally renowned dermatopathologist, educator and author. She is a Professor of Pathology and Laboratory Medicine at the Weill Cornell Medical College in Manhattan, and is board certified in anatomic pathology, dermatopathology and cytopathology. Dr. Magro is an expert in the diagnosis of complex inflammatory skin diseases. Her areas of expertise include cutaneous manifestations of auto-immune disease, systemic viral disease and vasculitis, atypical drug reactions, benign, atypical and overtly malignant lymphocytic infiltrates of the skin, and diagnostically difficult melanocytic proliferations.The award-winning author of *The Melanocytic Proliferation: A Comprehensive Textbook of Pigmented Lesions*, Dr. Magro has recently completed her second book, *The Cutaneous Lymphoid Proliferation, a comprehensive textbook on benign and malignant lymphocytic infiltrates*. She has co-authored over 280 peer reviewed papers and several textbook chapters. Dr. Magro frequently presents courses on inflammatory skin pathology and difficult melanocytic proliferations to the American Academy of Dermatology, the United States and Canadian Academy of Pathology, and the American Society of Clinical Pathology. Dr Magro has consistently been recognized in Who's Who in America®, Castle Connolly's renowned America's Top Doctors – New York Metro Area® edition and in the Super Doctors® list published in The New York Times Magazine.

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Table I

	Clinical Presentation	Histological Findings
Stankler and Ewan	Adult group: Bilateral follicular lesions on the thighs. Juvenile: group: Asymmetrical follicular plaque lesions affecting the trunk and axillae.	Early lesions: Follicular plugging with a perivascular and perifollicular infiltrate of lymphocytes and mast cells. Older lesions: Follicular plugging with marked parakeratosis of the ostium, contiguous with the ostium was mildly hyperplastic epithelium with loss of the stratus granulosum.
Ploysangam and Mutasim	Widespread discrete, erythematous, round hyperkeratotic papules measuring 2-4 mm in diameter on the torso and extremities.	The follicular epithelium was acanthotic with thinning or loss of the granular layer. Follicular hyperkeratotic plugging with parakeratosis containing neutrophils was seen. The epidermis contiguous with the ostium revealed psoriatic acanthosis with sparse dermal perivascular lymphocytic infiltrate.
Arps et al.	Pruritic follicularly-based hyperkeratotic papules on the scalp, neck, back, and extremities, occurring singly or in clusters.	Distended follicular infundibula with parakeratotic scale admixed with neutrophils was seen. The infundibular epithelium demonstrated mild acanthosis and hypogranulosis.
Thomas et al.	Asymptomatic symmetrical follicular lesions on the thighs, calves, and arms.	Biopsy revealed dilated central hair follicle with parakeratotic plugging with discrete areas of granular layer in the ostial-infundibular epidermis with a neutrophilic infiltrate.
Patil et al.	Dark rough horny papules on the knees and elbows that became diffuse erythematous scaly papules.	Follicular plugging and ostial parakeratosis with adjacent epidermal parakeratosis, hypogranulosis, elongated rete ridges with dermal papillary hyperplasia, suprapapillary thinning of epidermis, and dermal neutrophilic infiltrate was seen.
Cheng, Hanson, and Magro	Bilateral pruritic, erythematous papular rash on the forearms and lower legs with extension to the back.	Follicular parakeratosis with neutrophilic infiltration was identified. The follicular ostia exhibited hyperkeratosis with neutrophils. The adjacent epidermis showed neutrophil imbued parakeratosis, granular cell layer loss, and dilated dermal papillae capillaries lying in apposition to the basal layer of the epidermis

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