Acrodermatitis continua of Hallopeau, also known as acrodermatitis perstans and dermatitis repens, is a rare inflammatory pustular dermatosis of the distal fingers and toes. It is considered a variant of pustular psoriasis or, less commonly, its own pustular psoriasis-like independent entity. Precise pathophysiology and incidence are unknown. Case literature suggests predominance in women, but the disease affects both sexes and, rarely, children.

Acrodermatitis continua of Hallopeau initially presents as erythema overlying the distal digits that evolves into pustules. The nail bed is often involved, with paronychial and subungual involvement and atrophic skin changes. Most patients experience a chronic, relapsing course involving the proximal digit as the condition worsens. Acrodermatitis continua of Hallopeau has been reported to both evolve into and stem from generalized plaque or pustular psoriasis. The present patient was noted to have plaque psoriasis lesions nearly 1 year after the onset of her disease. Psoriatic arthritis is a rare complication, but distal phalangeal osteolysis is an important comorbidity.

The differential diagnosis includes infectious paronychia of viral, fungal, or bacterial etiology, infected contact dermatitis, and dyshidrotic eczema. Gram stain, potassium hydroxide mount, culture, and microscopy may be useful in diagnosis. Histopathologically, ACH is characterized by neutrophil-rich spongiform pustules within the epidermis, dermal edema, and lymphohistiocytosis. As in pustular psoriasis, biopsy from the nail bed often reveals acanthosis and spongiform pustules.

Treatment with topical corticosteroids, tacrolimus, fluorouracil, calcipotriol, methotrexate, acitretin, cyclosporine, and phototherapy have produced inconsistent responses. Successful treatment with tumor necrosis factor inhibitors and the IL-1 inhibitor anakinra have been reported. However, these agents are not always efficacious and may even have the potential to incite pustular psoriasis.

Two cases of ACH treated with concomitant ustekinumab and acitretin have been reported, one successfully, the other unsuccessfully. The present case is the first to our knowledge to be successfully treated with ustekinumab as monotherapy. Given that the literature supports ustekinumab as monotherapy and concomitant therapy for pustular psoriasis, ustekinumab was a reasonable choice for our patient and succeeded when other agents had failed.

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Additional Contributions: We thank the patient for granting permission to publish this information.


Treatment of Refractory Pityriasis Rubra Pilaris With Novel Phosphodiesterase 4 (PDE4) Inhibitor Apremilast

Pityriasis rubra pilaris (PRP) is an inflammatory dermatologic disorder of unknown cause characterized by hyperkeratotic follicular papules combining into salmon-colored scaling plaques, palmoplantar hyperkeratosis, and sharply demarcated islands of spared skin. We report a case of refractory PRP treated with the immune modulator apremilast.

Report of a Case | A white man in his 70s presented with an 8-month history of scaling, pink, pruritic papules, originating on his back (Figure, A), which coalesced to encompass nearly his entire body surface. There was extensive erythema with scaling and waxy, hyperkeratotic scaling of the palms and soles.

A punch biopsy was performed for hematoxylin-eosin staining and demonstrated alternating parakeratosis and orthokeratosis with spongiosis and mild superficial lymphocytic infiltrate. Given these findings, PRP was diagnosed.

The patient started acitretin therapy and showed initial improvement, but after 4 months, his disease continued to progress. He was transitioned to methotrexate therapy with prednisone bridging. However, the methotrexate regimen was discontinued after 8 weeks owing to lack of response and was replaced with cyclosporine. After 4 weeks of marginal response with cyclosporine and prednisone, infliximab was added, based on literature reports of improvement of PRP with tumor necrosis factor (TNF) inhibition.

The patient showed marked improvement after 1 infliximab infusion of 5mg/kg. Unfortunately, 4 weeks later, he was diagnosed with small-cell lymphocytic leukemia (SLL). Because TNF inhibitors have been associated with an increased risk of lymphoma, infliximab therapy was discontinued. The patient sought care for SLL, and rituximab and bendamustine chemotherapy was initiated. After completion of the chemotherapy, his PRP worsened.

The challenge was to identify a PRP-directed treatment in a patient with refractory disease and contraindication to
TNF inhibitors. After exhausting therapeutic options, we considered apremilast (Otezla; Celgene Corporation). The patient started with a dose of 10 mg/d and titrated over 5 days to the recommended maintenance dose of 30 mg twice daily.

Four weeks later, the patient reported significant improvement in symptoms. He complained of mild gastrointestinal upset but denied other new symptoms. The most common adverse effects reported with apremilast are diarrhea, nausea, and headache. At 8- and 12-week follow-ups, he reported further decreases in body surface area involvement of PRP. At 6- and 8-month follow-ups, he showed nearly complete resolution of skin findings (Figure, B).

Discussion | There is no unifying etiology to the pathogenesis of PRP. Current treatments are mainly empirical or based on case reports. Systemic retinoids and methotrexate are often used as first-line treatments. Other therapies include azathioprine, cyclosporine, fumaric acid, mycophenolate mofetil, vitamin D analogues, and phototherapy.3,4

Eastham et al3 demonstrated, in the most comprehensive analysis to date, that TNF inhibition is effective for refractory PRP. The patient in the present case responded well to infliximab but could not continue therapy because he developed SLL. Based on the proposed immune-driven mechanism of PRP, we explored novel treatments targeting regulation of inflammatory responses.

Apremilast is an oral, small-molecule inhibitor of phosphodiesterase 4 (PDE4) approved for treatment of moderate to severe plaque psoriasis and psoriatic arthritis.5 Expressed in various cell types including keratinocytes, PDE4 participates in regulation of immune and inflammatory processes by regulating intracellular cyclic adenosine monophosphate (cAMP) and downstream protein kinase A pathways and phosphorylating CREB (transcription factor cAMP-response element binding protein).6 Activation of this pathway results in downstream inhibition of proinflammatory cytokines, including TNF. Given our patient’s improvement with TNF inhibition, the choice of apremilast was a logical next step for the patient in the present case. The drug is appealing owing to its minimal adverse effects and monitoring requirements.6 At 12-month follow-up, the patient remained disease free.

Our experience suggests that apremilast may be an effective treatment for refractory PRP. Additional studies are necessary to further establish the role of PDE4 inhibitors as an option for refractory PRP.

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To the Editor The Editorial by Hill and colleagues is interesting and informative. They suggest that online reviews are a fast, efficient, transparent way to obtain actionable feedback from patients. However, we believe one of the major problems of physician online ratings is actually the lack of transparency. It is very difficult or even impossible to tell if the rater is a patient or someone posing as a patient, such as an unhappy employee or a competitor. Furthermore, even positive ratings have limited utility; the origin of shining comments might be anonymous prose of the physician. Not to mention, most of the for-profit online rating sites often create a handful of posts. The size of such a sample, compared with the number of patients that any given physician takes care of, lacks any statistical significance. Thus, available online reviews of physicians lack accountability or quality control.

In a recent publication, Schlesinger and colleagues opined that “Most existing websites post comments submitted from any source, with no assurance that they come from real patients. Clinicians’ legitimate concerns about accuracy and representativeness may discourage them from using comments for improving clinical practice.” They further opined that “Without active policy intervention, the pernicious influences of comments may outweigh the positive.” They emphasize that the key is to hold narrative data to the standard of scientific rigor. Most importantly, there should be some assurance that narratives are representative of patient experience.

Gray and colleagues conducted research to investigate the validity of physician website ratings by measuring the association between US physician website ratings and traditional quality measures of clinical and patient experience. They found no evidence that physician website ratings were associated with clinical quality measures.

Nevertheless, physician online ratings may have some serious unintended consequences to physicians. Zgierska and colleagues have suggested that patient satisfaction survey utilization may promote job dissatisfaction, attrition, and inappropriate clinical care among some physicians. Fifty-nine percent reported that their compensation was linked to patient satisfaction ratings. Seventy-eight percent reported that patients’ satisfaction surveys moderately or severely affected their job satisfaction; 28% had considered quitting their job or leaving the medical profession.

With these problems in mind, we question the validity of online reviews related to physicians and their performance and raise a concern about the potential negative impact of this largely invalid pool of data related to physicians’ work.

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In Reply The problems with online reviews—few data, invalid instruments, lack of representativeness, and lack of transparency—are all predicated on physicians not actively encouraging online reviews. Should physicians choose to participate, large numbers of ratings can be obtained, validated instruments can be used, representativeness can be improved, and the high quality of care that physicians provide can be made transparent to the public.

Online reviews are not going to suddenly disappear. More and more, the public seeks out reviews for information on physicians. By leaving only unhappy outliers to enter ratings, physicians can become victims of online reviews. Worse, by attempting to suppress physician ratings, physicians would look like they had something to hide. The alternative is for physicians to embrace online reviews and encourage our patients to participate. One company, Medical Justice, that tried to help physicians by suppressing online ratings recognized this was a huge mistake and now encourages online rating. Having more representative reviews results in higher scores and dramatically diminishes the impact of outliers and fake reviews.

In-office kiosks, patient portals, and verification code systems can help assure that reviews come from actual patients. Reminders at checkout or on patient instruction sheets can include instructions on how patients can leave online reviews. Validated online measures are feasible, and people put more trust in recommendations from large numbers of reviewers. Some fake ratings may occur, but their effect would be swamped by extensive patient participation; a few negative ratings may even increase patients’ perception that positive ratings are valid.

Sticking our heads in the sand and avoiding online reviews because of their limitations would be a self-fulfilling disaster for physicians and our patients. The public is online, and we would do well to use the internet to get feedback from our patients and make that feedback transparent to help patients see the high quality of care that we provide.

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