to improve in the next days (Figure 2). Two weeks after amino acid treatment, radiolabeled somatostatin analogue therapy was given to induce tumor reduction and allow for tumor resection without affecting the adjacent blood vessels.

Discussion | The diagnosis of NME is often challenging, both clinically and histologically. The skin lesions can mimic other dermatoses, and histologic analysis is considered a poor diagnostic tool, as was evident in the present case. Delay in diagnosis is also attributed to the rarity of the tumor, underrecognition of the dermatologic presentation, and the lack of specificity of the other symptoms. Therefore, a high index of suspicion should be maintained when a patient presents with suggestive skin and systemic symptoms.

Necrolytic migratory erythema significantly affects patient quality of life and is resistant to treatment. The use of intravenous amino acids as a treatment for NME has been reported, but the quick resolution of the skin lesions was not well documented. We showed the quick resolution of the skin and mucosal lesions after only 24 hours, leading to marked improvement in quality of life for the patient.

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Treatment of Acrodermatitis Continea of Hallopeau With Ustekinumab as Monotherapy

Acrodermatitis continua of Hallopeau (ACH), a rare pustular eruption affecting the distal digits, was successfully treated with ustekinumab as monotherapy.

Report of a Case | A woman in her 20s presented with recalcitrant and well-demarcated hand dermatitis unresponsive to topical medications including corticosteroids. Her medical history was remarkable for “eczema” dating back to childhood. Patch testing results were negative.

On examination, the left and right third fingers had sharply demarcated erythematous scaly plaques with fissures, scale crust, and nail pitting. The left third finger was confluentely covered, leaving the rest of the hand unaffected (Figure 1). This constellation of findings limited to the fingers suggested an eruption consistent with ACH. Given her condition’s recalcitrant history and the typical resistance of ACH to topical medicaments, she was started on a regimen of cyclosporine, 100 mg, twice daily (3.2 mg/kg), tacrolimus ointment nightly,
calcipotriene cream daily, and diluted bleach soaks daily. She experienced intolerable gastrointestinal upset, so her cyclosporine dose was decreased to 50 mg twice daily, and the topical applications were increased to twice daily.

One month later, the patient had only a partial response. Another cyclosporine formulation was tried at 75 mg twice daily. She developed worsening onychodystrophy, and the cyclosporine dose was increased to 175 mg/d with clobetasol twice daily.

Four months after initial presentation, cyclosporine treatment was discontinued owing to protracted nausea. She was started on a regimen of 45-mg ustekinumab, 1 subcutaneous injection followed 1 month later by a second injection with subsequent injections every 3 months thereafter. Ustekinumab is a human monoclonal IgG antibody that blocks interleukin (IL)-12 and IL-23. Topical tacrolimus was used only during the 1-month induction period. Two months after initiation of ustekinumab therapy, the patient had nearly total clearing that, however, did not endure to the next injection period 3 months later, and so the next dose was increased to 90 mg. By month 7, the fingers were clear (Figure 2) with a few erythematous, scaly, and well-demarcated psoriatic plaques on the scalp, legs, and 1 web space.
Discussion | 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.1 Precise pathophysiology and incidence are unknown. Case literature suggests predominance in women, but the disease affects both sexes and, rarely, children.2

Acrodermatitis continua of Hallopeau initially presents as erythema overlying the distal digits that evolves into pustules.2 The nail bed is often involved, with paronychial and subungual involvement and atrophic skin changes.3 Most patients experience a chronic, relapsing course involving the proximal digit as the condition worsens.4 Acrodermatitis continua of Hallopeau has been reported to both evolve into and stem from generalized plaque or pustular psoriasis.3 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,3 but distal phalanx osteolysis is an important comorbidity.1

The differential diagnosis includes infectious paronychia of viral, fungal, or bacterial etiology, infected contact dermatitis, and dyshidrotic eczema.4 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.4 As in pustular psoriasis, biopsy from the nail bed often reveals acanthosis and spongiform pustules.3

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 anakinra5 have been reported. However, these agents are not always efficacious and may even have the potential to incite pustular psoriasis.6

Two cases of ACH treated with concomitant ustekinumab and acitretin have been reported, one successfully,7 the other unsuccessfully.5 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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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.1 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.2,3

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,4 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...