


Rapid Clearance of Necrotic Migratory Erythema Following Intravenous Administration of Amino Acids

Necrotic migratory erythema (NME) is a rare paraneoplastic skin disorder, considered the hallmark clinical sign of glucagonoma syndrome. Its recognition allows for early diagnosis of the tumor, which might lead to a better prognosis. Additional mucocutaneous features can include angular stomatitis and glossitis. We present a case of NME that cleared rapidly following intravenous administration of amino acids.

Report of a Case | A woman in her 50s was evaluated for a persistent pruritic eruption lasting for 3 months. The eruption started on the lower extremities and perianal area and rapidly expanded to involve the genital area. Her medical history included sicca syndrome for the last 20 years. In the last 2 years, the patient had developed angular cheilitis and glossitis. She also reported weakness, anorexia, weight loss (approximately 20 kg within a year), numbness and tingling sensation in the extremities, and instability during walking.

Physical examination revealed erythematous migratory plaques with irregular, erosive, scaly borders and hypopigmented centers on the perianal and genital areas. Lichenified erythematous and hyperpigmented plaques were evident on the distal shins. The patient had an erythematous tongue with angular fissures (Figure 1). Cardiovascular, abdominal, and respiratory examination findings were normal.

Laboratory tests revealed slight anemia (hemoglobin, 11.4 g/dL; reference range, 12-16 g/dL) and borderline increased hemoglobin A1c (5.8%; reference range, 4.0%-5.7%). Further evaluation revealed increased levels of gastrin (129 ng/mL; reference range, 13-115 ng/mL), chromogranin A (223 ng/mL; reference range, 19-98 ng/mL), and neuron-specific enolase (21.11 ng/mL; reference range, 0-12 ng/mL). Somatostatin receptor scintigraphy revealed high concentrations of somatostatin receptors in the pancreatic head and adjacent lymph nodes. Computed tomography demonstrated an abdominal central tumor in the pancreatic head, medial to the duodenum, causing superior mesenteric vein blockage. Endoscopic ultrasonographic examination and needle biopsy showed a neuroendocrine tumor, grade 2, and based on findings of immunohistochemical staining (MIB-1 labeling index, 3%) and markedly increased glucagon levels (>500 pmol/L; reference range, <50 to 150 pmol/L) the patient was diagnosed with glucagonoma.

Histopathologic examination of a skin biopsy specimen from the perianal area showed nonspecific subacute dermatitis with mild perivascular lymphocytic infiltrate accompanied by plasma cells and a few eosinophils. There was no evidence of necrolysis in the upper epidermis.

Two weeks after establishing the diagnosis of glucagonoma, treatment was initiated with intravenous administration of a commercial mixture of 500 mL of amino acid solution (Vamin 18 electrolyte free; Fresenius Kabi) for 12 hours, once a day, for 2 consecutive days. Marked improvement was observed after 24 hours, with reduced erythema and scaling, reepithelialization of the erosions in the genital area, and closing of the fissures on the tongue. The skin lesions continued to improve.
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 Continua 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 confluentively 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,