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Necrolytic Migratory Erythema

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A 73-year-old man with a history of diabetes mellitus, hypertension, and coronary artery disease was referred to our outpatient dermatology clinic with pruritic, thin, scaly, annular, erythematous, and eczematous (pseudovesicular) plaques with central clearing on his abdomen and lower extremities.

The patient had a 1-year history of chronic diarrhea and diabetes mellitus. He had previously been treated only with topical and intramuscular corticosteroids, which had led to temporary improvement. While the patient was off treatment, a skin biopsy was performed, the results of which revealed vacuolated keratinocytes, spongiosis, irregular psoriasiform hyperplasia, intraepidermal Langerhans cells, and scattered dermal eosinophils, findings that were interpreted as a drug hypersensitivity reaction. Direct immunofluorescence testing was performed out of concern for pemphigus foliaceus, and the results were negative for autoimmune disease, connective tissue disease, and vasculitis.

Drug-elimination protocols were attempted but led to no improvement. The patient was continued on triamcinolone cream, 0.1%, and narrowband UV-B phototherapy was initiated. The patient's lesions

improved at the involved sites, but new pruritic plaques continued to develop on his lower extremities and lower back.

Five months after presentation, the patient's signs and symptoms rapidly worsened. He developed painful, pruritic, fissured and erosive lesions, as well as crusted, scaly, arcuate plaques with a serpiginous pustular border on his neck, face, upper extremities, abdomen, lower extremities, groin, and intergluteal cleft, along with desquamating plaques on the soles (**Figure 1**).

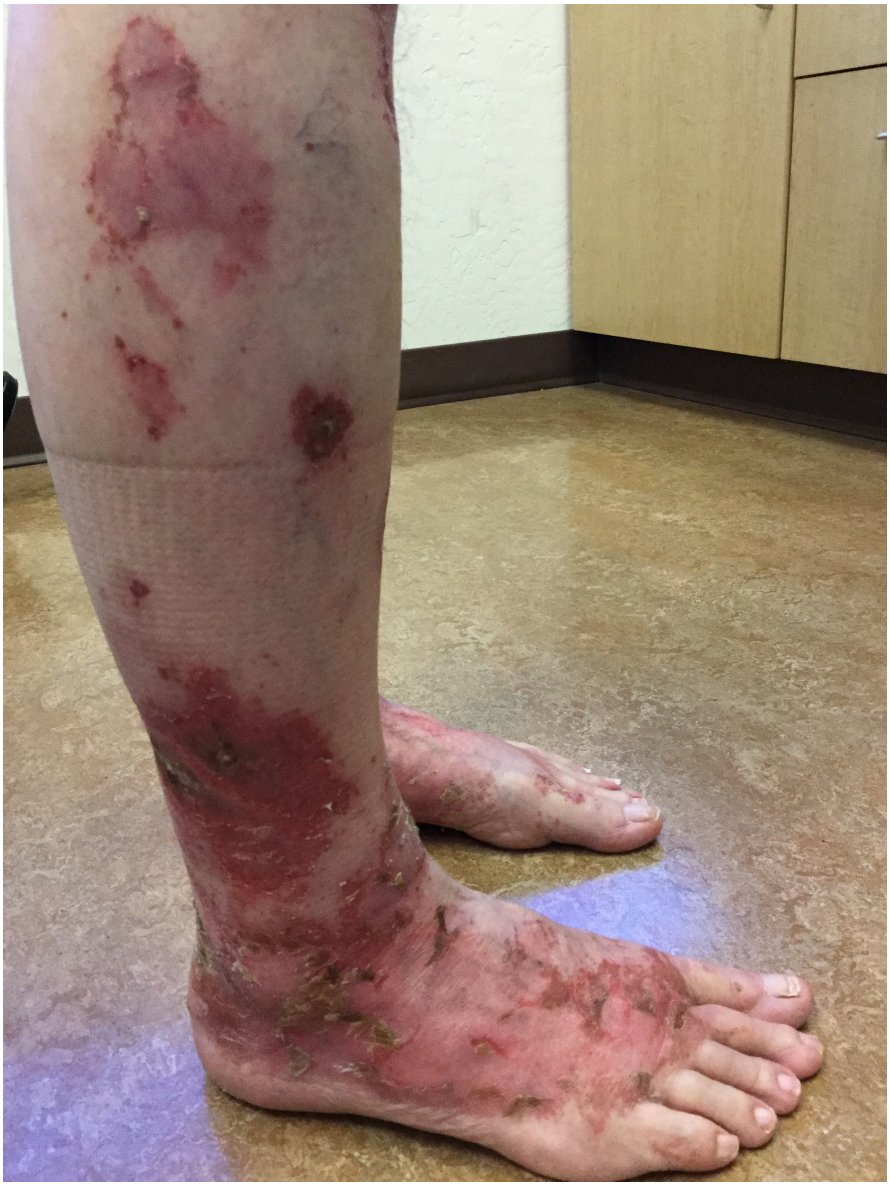

Figure 1. *The patient's lower extremities prior to diagnosis.*

Given concern for pustular psoriasis and acrodermatitis enteropathica, the patient was administered secukinumab, 300 mg, and laboratory workup and a second punch biopsy were performed. Laboratory tests revealed a glucagon level of 1968 pg/mL (reference range, 8-57 pg/mL), a zinc level of 43 μg/dL (reference range, 60-130 μg/dL), normochromic normocytic anemia, hypoalbuminemia, and elevated levels of amylase, lipase, alkaline phosphatase, and glucose.

Histopathology test results showed psoriasiform epidermal hyperplasia, spongiotic epidermal changes, and confluent parakeratosis. Superficial keratinocytes had cytoplasmic vacuolization. Increased intraepidermal Langerhans cells and scattered dyskeratotic keratinocytes were also present, consistent with necrolytic migratory erythema (NME) (**Figure 2**).

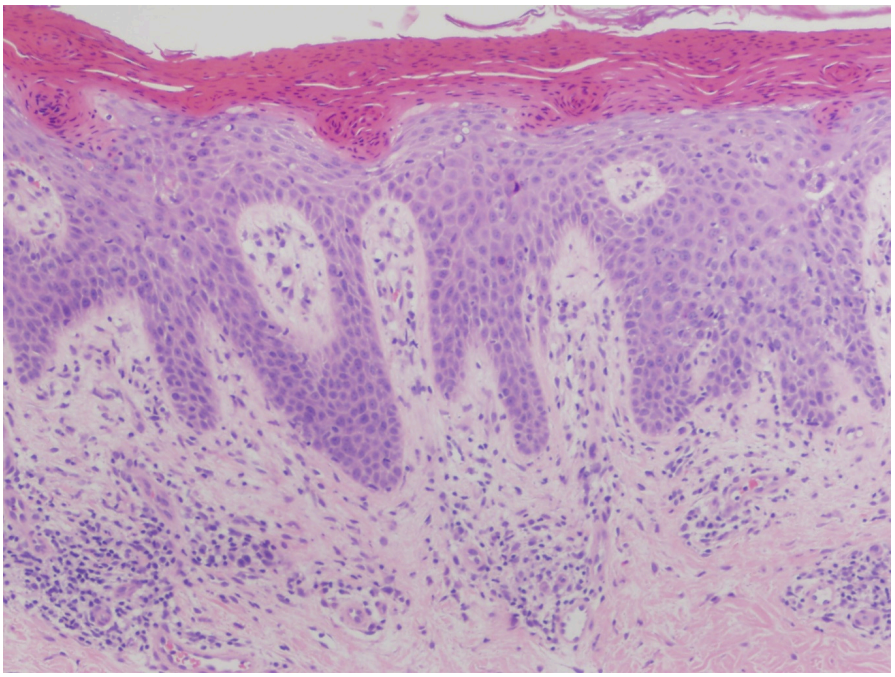

Figure 2. *A punch biopsy histology test showed cytoplasmic vacuolization of keratinocytes, increased intraepidermal Langerhans cells, and dyskeratotic keratinocytes.*

An abdominal computed tomography scan revealed a large tumor in the pancreatic body, and results of an ultrasound-guided needle biopsy was consistent with a well-differentiated neuroendocrine tumor that was positive for chromogranin and synaptophysin, confirming a diagnosis of glucagonoma.

The patient underwent distal pancreatectomy with splenectomy. Following resection of the tumor, the patient had complete resolution of his cutaneous eruption within 3 weeks.

Discussion. NME is a paraneoplastic dermatosis that is the presenting feature in approximately 70% of patients with glucagonoma syndrome, a pancreatic neuroendocrine tumor representing well less than 5% of all pancreatic neoplasms.[1] NME characteristically manifests as erythematous coalescing papules with vesiculation and well-circumscribed scaling plaques. Peripheral expansion with central clearing often creates a serpiginous configuration. Sites of predilection include the face, intertriginous areas, flexural regions, and extremities. Glossitis and angular cheilitis may also be present.[2] Patients with glucagonoma often present with diabetes mellitus, diarrhea, anemia, and weight loss.[3]

NME is difficult to diagnose, since its clinical and pathologic features mimic several dermatologic diseases, including drug hypersensitivity eruptions, pustular psoriasis (Sneddon-Wilkinson disease),

pemphigus variants, and linear immunoglobulin A bullous dermatosis. Early recognition and treatment of glucagonoma tumors prevent metastatic disease, reducing both morbidity and mortality.

A thorough history, serial examination, and repeated biopsies are critical to appropriately evaluate patients with longstanding cutaneous manifestations that are recalcitrant to standard treatment regimens, and in patients with cutaneous concerns arising with other systemic signs and symptoms.

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