Necrolytic migratory erythema

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A 66-year-old previously healthy man presented to the dermatology clinic at the hospital with a 2-month history of recurring rash, weight loss (6 kg), general weakness, burning mouth and low mood. Skin lesions had presented on his lower legs and then spread to the popliteal fossa, groin and axilla, with symmetric distribution. The patient's medical history was unremarkable, and he was not taking any medication. On examination, he appeared cachectic but had normal vital signs. He had erythematous erosive patches with advancing scaling borders (Figure 1) and a red glossy tongue. Results from laboratory testing, including calcium, parathyroid hormone, gastrin and calcitonin, were normal except for an elevated erythrocyte sedimentation rate of 45 (normal range 0–10) mm/h, and elevated levels of C-reactive protein (290 [normal range 0–5] mg/L), fasting blood glucose (12.7 [normal range 5.6–6.9] mmol/L) and hemoglobin A1c (9.1% [normal range 4.2%–6.3%]). Serum glucagon was grossly elevated at 2254 (normal range 25–250) ng/L.

We admitted the patient to hospital for investigation. Based on his highly elevated level of serum glucagon, we ordered abdominal computed tomography (CT), which showed no enteropancreatic or adrenocortical tumours. Therefore, we excluded multiple endocrine neoplasm type 1 and diagnosed necrolytic migratory erythema associated with hyperglucagonemia. His skin lesions improved with care of his hyperglycemia and fluid resuscitation with multivitamins in the hospital.

We intended to perform additional tests to detect glucagon-secreting tumours on an inpatient basis. However, the patient suddenly presented with fever and dyspnea during his hospital stay. Computed tomography of the chest showed atypical pneumonia in both lungs. His lung condition deteriorated rapidly and he died.

Necrolytic migratory erythema is observed in about 70% of patients with glucagonoma, an uncommon disease.[1] The differential diagnosis of necrolytic migratory erythema includes acrodermatitis enteropathica, essential fatty acid deficiency, drug reactions and vitamin deficiencies.[2] Pseudoglucagonoma syndrome refers to necrolytic migratory erythema in the absence of a glucagon-secreting tumour.[2] Treatment involves surgical excision of the primary tumour in glucagonoma, but symptomatic treatment with somatostatin or octreotide, or chemotherapeutic agents may be considered.[3]

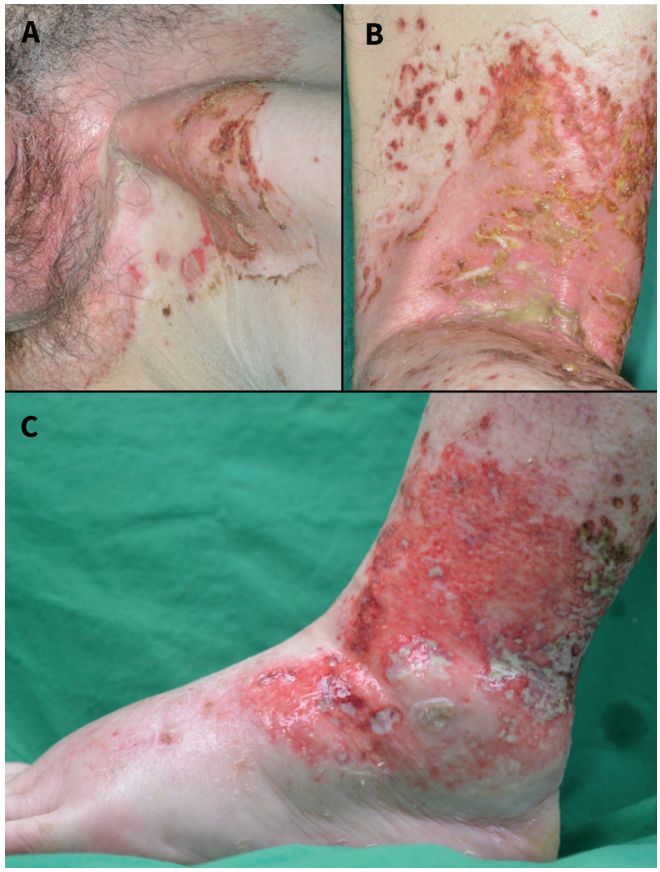

Figure 1: Erythematous erosive patches with advancing crusted borders on the (A) left groin, (B) left popliteal fossa and (C) left ankle of a 66-year-old man with necrolytic migratory erythema from glucagonoma.

References

1. van Beek AP, de Haas ER, van Vloten WA, et al. The glucagonoma syndrome and necrolytic migratory erythema: a clinical review. *Eur J Endocrinol* 2004; 151:531-7.
2. Tierney EP, Badger J. Etiology and pathogenesis of necrolytic migratory erythema: review of the literature. *MedGenMed* 2004;6:4.
3. Kovács RK, Korom I, Dobozy A, et al. Necrolytic migratory erythema. *J Cutan Pathol* 2006;33:242-5.

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The authors have obtained consent from the patient's family.

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