Visual Vignette

Samantha Sathyakumar, MD[1]; Kripa Elizabeth Cherian, MD[1]; Felix Jebasingh, MD[1]; Julie Hepzhibah, MD[2]; Nitin Kapoor, MD, DM (Endo)[1]; Thomas Vizhalil Paul, MD, DNB (Endo), PhD (Endo)[1]

From the [1]Department of Endocrinology, Diabetes & Metabolism, and [2]Department of Nuclear Medicine, Christian Medical College, Vellore, India.

Case Presentation: A 56-year-old woman presented with history of waxing and waning scaly erythematous skin rashes, weight loss, and recent onset diabetes mellitus. Recurrent erythematous skin lesions associated with ulceration and crusting were present, predominantly over lower limbs for the previous 3 years (Fig. 1). There was progressive weight loss of about 12 kg, with episodes of abdominal discomfort of 2-years duration. She was diagnosed with diabetes mellitus a few months previous and had been on insulin since then. Clinical examination revealed anemia, angular cheilitis, and glossitis. The rest of the examination was unremarkable. Laboratory evaluation showed normocytic normochromic anemia (hemoglobin, 8.1 g/dL; normal, 12 to 15 g/dL) and hypoalbuminemia of 2.9 g/dL (normal, 3.5 to 5.5 g/dL). A skin biopsy done showed features of lamellar hyperkeratosis, confluent parakeratosis, irregular acanthosis, lymphohistiocytic infiltrate with neutrophilic crusting, and dermal edema. A ^{68}Ga-DOTA-octreotate (DOTATATE) positron emission tomography–computed tomography (CT) scan was performed (Fig. 2). **What is the diagnosis?**

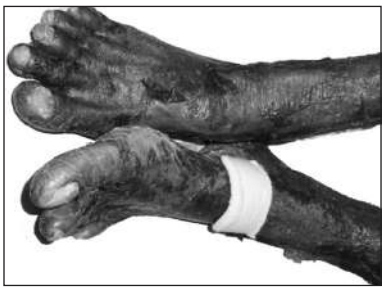

Fig. 1

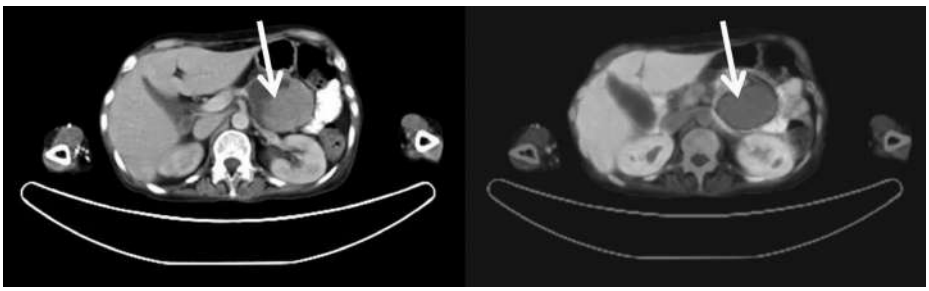

Fig. 2

Answer: Glucagonoma syndrome. This patient had features of necrolytic migratory erythema (NME), and CT scan revealed a large, well-defined, intensely enhancing lesion of 6.2 × 5.4 cm in the pancreatic tail, which was DOTATATE-avid on functional imaging. Her serum glucagon level was 4,375 pg/mL (normal, 20 to 250 pg/mL). In the background of recent onset diabetes, weight loss, cheilosis, normocytic anemia with NME, and a pancreatic neuroendocrine tumor, a diagnosis of glucagonoma syndrome was made. There was no evidence of metastatic disease, and she was scheduled for surgical excision of the tumor. Although this syndrome was first described in 1942, very few cases have been reported in the literature to date (1). NME is present in about 80% of these individuals but may also be seen in conditions such as pellagra and zinc deficiency. About 75% are malignant, and half of cases involve metastasis at the time of diagnosis. Surgery remains the treatment of choice if the tumor is localized at diagnosis (2). In case of metastatic disease, a multimodality approach combining surgery, transarterial chemoembolization, percutaneous radiofrequency thermal ablation, long-acting octreotide, and antineoplastic agents such as evirolimus and sunitinib may be effective (3).

DISCLOSURE

The authors have no multiplicity of interest to disclose.

REFERENCES

1. **McGavran MH, Unger RH, Recant L, Polk HC, Kilo C, Levin ME.** A glucagon-secreting alpha-cell carcinoma of the pancreas. *N Engl J Med*. 1966;274:1408-1413.
2. **Kapoor N, Fleming JJ, Ramakrishna B, et al.** An unusual cause for weight loss and diabetes. *Hong Kong Med J*. 2011;17:505-506.
3. **van Beek AP, de Haas ER, van Vloten WA, Lips CJ, Roijers JF, Canninga-van Dijk MR.** The glucagonoma syndrome and necrolytic migratory erythema: a clinical review. *Eur J Endocrinol*. 2004;151:531-537.

Editor's Note: Submissions to "Visual Vignettes" are welcomed. Please send to Endocrine Practice, 245 Riverside Avenue, Suite 200, Jacksonville, FL 32202.

Address correspondence to Dr. Thomas V. Paul, Department of Endocrinology, Diabetes and Metabolism, Christian Medical College, Vellore-632004, India. E-mail: thomasvpaul@yahoo.com.

Published as a Rapid Electronic Article in Press at http://www.endocrinepractice.org on September 15, 2016. DOI: 10.4158/EP161307.VV