

PICTURES IN DIGESTIVE PATHOLOGY

Glucagonoma and necrolytic migratory erythema

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A 54-year-old male with a history of necrolytic migratory erythema (NME) (Figs. 1 and 2) and glossitis. Routine blood tests were normal except for glucose: 145 mg/dL. Baseline plasma glucagon levels were 1200 pg/mL. Serum zinc was 97 mcg/dL. An abdominal CT scan showed a large mass involving the body and tail of the pancreas. A distal pancreatectomy with splenectomy was performed and no evidence of metastatic disease was observed. The skin rash cleared within a week after the operation and the patient remains free of disease at 38 months following surgery.



Fig. 1.- NME. Necrolytic migratory erythema.
ENM. Eritema necrolítico migratorio.

COMMENTS

Glucagonoma syndrome is a paraneoplastic phenomenon comprising a pancreatic glucagon-secreting insular tumor, necrolytic migratory erythema (NME), diabetes, weight loss, anemia, stomatitis, thrombo-embolism, dyspepsia, and neuropsychiatric disturbances. The occurrence of one or more of these symptoms associated with a proven pancreatic neoplasm fits this diagnosis (1). Other skin and mucosal changes such as atrophic glossitis, cheylitis, and inflammation of the oral mucosa may be found (2). Hyperglycemia may be included –multiple endocrine neoplasia syndrome, i.e., Zollinger-Ellison syndrome– or the disease may result from a glucagon-secreting tumor alone.

These tumors are of slow growth and present with non-specific symptoms in early stages. At least 50% are metastatic at the time of diagnosis, and therefore have a poor prognosis.



Fig. 2.- NME. Necrolytic migratory erythema.
ENM. Eritema necrolítico migratorio.

Five-year survival rate is unknown because of its rarity. A study has shown that tumor-related deaths occurred in 9 of 21 patients controlled within an average of 4.9 years after diagnosis. The remaining patients were alive after an average follow-up of 3.7 years (3).

CT scanning, MRI and US are currently employed for tumor localization. Positron-emission tomography has been successfully used in recent cases. Glucagonomas have receptors for somatostatin in more than 80% of cases. However, and because of their rarity, the sensitivity and specificity regarding octeotride analogues –i.e., indium-111 DTPA Nterminal Dfenyl-alanine octeotride– have not been established because of a lack of large series. These isotopes can also be employed as a therapeutic tool in recurrent or metastatic disease because of their beta-radiation emission ability (4).

Surgical resection is the only curative procedure nowadays and can be performed only when the tumor is localized and no evidence of metastatic disease is present. Even laparoscopic resection has been used in a few isolated cases. As this tumor grows slowly and has a tendency towards encapsulation, complete removal can be done in early stages when a proper diagnosis is made. Tumor debulking and mass reduction are effective for cutaneous rash relief (5), as is hepatic artery embolization for the selective necrosis of metastases. Liver resection could be considered along with pancreatectomy when isolated metastases are present, and transplantation for diffusely spread disease.

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