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Case report

Pancreatic glucagonoma: observing the skin can lead to diagnosis

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Abstract

This is a case report of a patient with a pancreatic tumor, known as glucagonoma, whose diagnosis was suspected because of skin manifestations which led to performing a CT scan, finding the mass. She underwent surgery with satisfactory results.

Keywords

Glucagonoma; Pancreatic tumor; Necrolytic migratory erythema.

INTRODUCTION

Glucagonomas are extremely rare tumors, with an incidence of 1 case per 1 million people. Usually, these tumors are large (>3 cm) and are located mainly in the tail or body of the pancreas due to the high prevalence of α-cells in this area. More than 50%of these tumors are metastatic at the time of diagnosis (1).

The incidence of glucagonoma in men and women is similar and most patients develop them between the fifth and sixth decade of their lives. A particular characteristic of these tumors is that they are associated with skin lesions known as erythema necrolytica migrans. These lesions can lead to the suspicion of glucagonoma as its first manifestation. Patients may also have diabetes, weight loss, deep vein thrombosis (DVT) and abdominal pain (2).

OBJECTIVE

To present the clinical manifestations of a patient with glucagonoma and describe how the diagnosis was achieved.

MATERIALS AND METHODS

This is the case of a 58-year-old female patient with atrophic glossitis in the central facial region, as well as erythematous crusted plaques in the nasal cavities and angular cheilitis in the labial commissures (**Figure 1**). Similarly, the patient had crusted erythematous and erythematous-brown plaques on her limbs, presenting with centrifugal growth, in different stages (some active excoriated and others macular with post-inflammatory hyperpigmentation) (**Figure 2**).

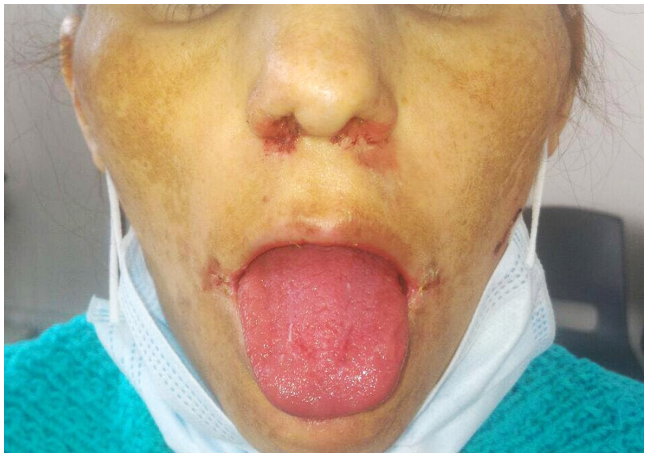

Figure 1. Angular cheilitis and crustal erythematous crusted plaques are observed.

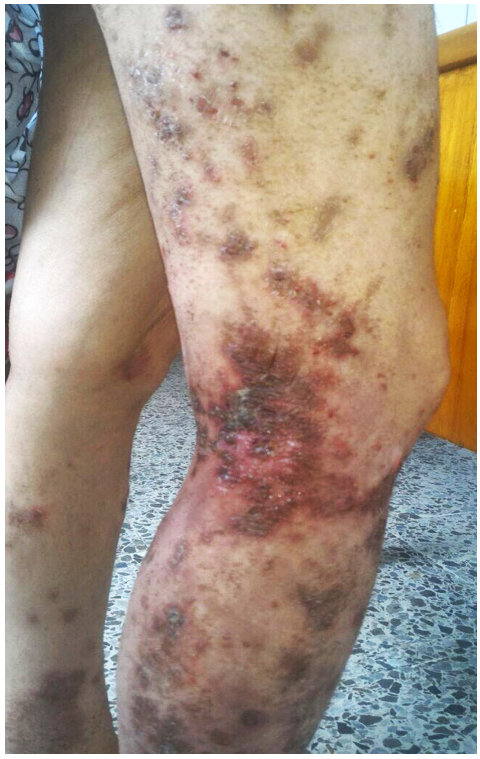

Figure 2. Crusted erythematous and erythematous-brown plaques on the limbs.

A difference in the diameter of the legs secondary to a deep thrombosis was observed. Subsequently, the patient developed anemia and diabetes. Therefore, multiple studies were performed, but it was not possible to identify the cause of the thrombosis. When the patient started to lose weight, together with abdominal pain, she was referred to our unit. During this period, she only received topical steroids to treat her lesions and oral therapy to manage her diabetes. Then, a CT scan was performed, finding a large mass in the tail of the pancreas (**Figure 3**), as well as a 2cm liver metastasis. Upon suspicion of a glucagonoma, a plasma glucagon test was performed, reporting elevated levels. For that reason, she was taken to a distal pancreatectomy (**Figure 4**), which allowed controlling her skin lesions and the diabetes. The pathology report confirmed the existence of the glucagonoma. After a year, the patient was still in cancer treatment (chemotherapy), showing a good response.

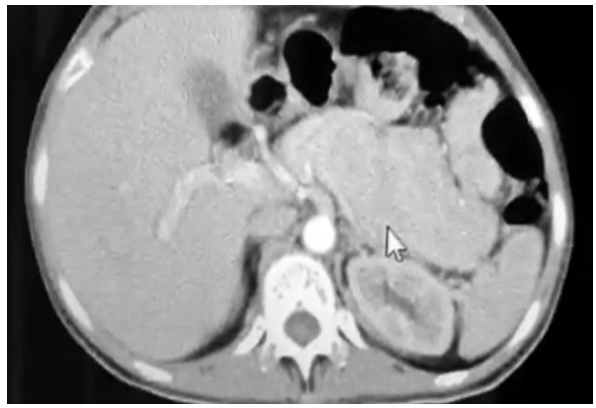

Figure 3. The arrow points to a large mass at the tail of the pancreas, which displaces the left kidney.

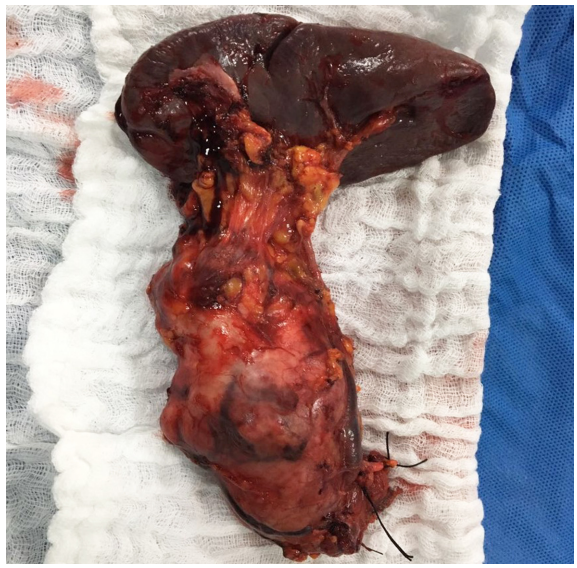

Figure 4. Surgical specimen taken from the tail of the pancreas, which included a splenectomy.

Glucagonomas were first described in 1942 by Becker *et al*. These authors reported a case, very similar to once presented here, of a 45-year-old woman with mucocutaneous lesions associated with an altered glucose tolerance test. During the patient's autopsy, a mass was found in the pancreas (3).

However, it was Dr. McGavran who, in 1966, described the typical symptoms of this condition, which are characterized by the presence of dermatitis associated with elevated glucagon levels, since glicagonomas are neuroendocrine tumors that originate in endodermal pluripotent stem cells and arise from pancreatic islets cells (4-7).

Most glucagonomas are solitary; however, less than 10% of them have been associated with the syndrome of multiple endocrine neoplasia type 1 (MEN1). In our case, this syndrome was ruled out (8).

CONCLUSION

Glucagonoma is a very rare tumor, which is why it is rarely suspected. The case reported here shows that the skin lesions of the patient could have led to an earlier diagnosis since the necrolytic erythema migration confirmed in the patient's pathology is typical of the disease. This tumor is associated with DVT, diabetes, anemia and weight loss, which are the five typical characteristics that should lead to suspect the presence of this pancreatic tumor. If detected timely, it has a very good prognosis.

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