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Glucagonoma Revealed by a Necrolytic Migratory Erythema

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Abstract

Glucagonoma is a rare pancreatic neuroendocrine tumor that usually develops in the body or tail of the pancreas. Both imaging features of a pancreatic tumor and elevated glucagon serum levels confirm the diagnosis. We report the case of a patient presenting with diffuse erythematous lesions consistent with necrolytic migratory erythema.

Text

Glucagonomas account for 1% of neuroendocrine tumors and less than 5% of primary pancreatic malignancies [1], arising from the alpha islet cells of Langerhans. Although it may appear to be a benign condition, 80% have malignant potential [2] with 50% of metastatic disease at the time of diagnosis the most common site being the liver [3,4].

We report the case of a 53-year-old male with persistent painful diffuse erythematous skin lesions of the trunk, upper and lower limbs as well as on the perineum Figure 1, and had experienced weight loss. Laboratory analysis showed hyperglycemia and hyperglucagonemia. Computed tomography (CT), revealed a well-circumscribed corporeal pancreatic lesion, homogeneously enhanced Figure 2, with tail atrophy Figure 3, no calcification, no fat stranding, and no lymphadenopathy. Further investigation showed multiple hypodense liver nodules on the arterial phase Figure 4.

Glucagonomas are typically revealed by the glucagonoma syndrome triad characterized by a Para neoplastic syndrome associating the presence of a pancreatic mass, diabetes mellitus, and necrolytic migratory erythema (NME) [5]. Sometimes, the various clinical presentations can be misleading, but the presence of a painful scaly erythematous rash (Figure 1) is one of the first signs of onset. NME is in fact the most specific and hallmark clinical sign present in 70% of cases [2].

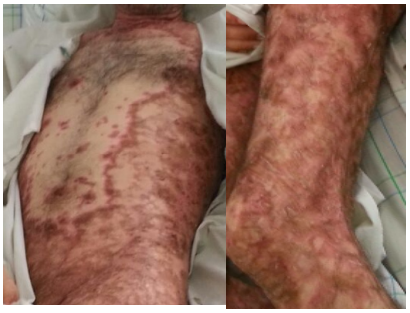

Figure 1: Body covered with purplish confluent erythematous macules with scaling and crusting lesions and active erythema on the edges. (Trunk on the left and lower limb on the right).

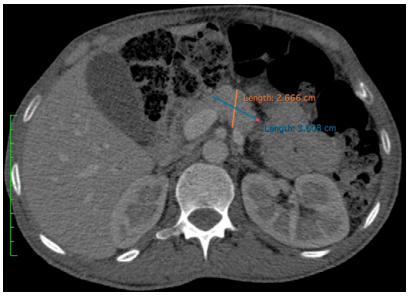

Figure 2: Axial CT scan of the abdomen on portal phase shows a large mass of the pancreas (26x30mm) homogeneously enhanced.

The diagnosis is confirmed by both laboratory tests with elevated levels of glucagon and imaging features of a pancreatic neuroendocrine tumor.

On CT scan it appears as a large (2 to 5cm of diameter), well-defined mass (Figure 2), of the body or the tail of the pancreas. After contrast injection, during the arterial phase, they tend to be homogeneous or inhomogeneous hypodense, generally well despicable with respect to the adjacent parenchyma, and hypodense or are Odense during the portal phase, due to their vascularization Figure 3. [6]

The presence of peripheral calcifications, cystic or necrotic changes along with lymphadenopathies are suggestive of the malignant potential [7].

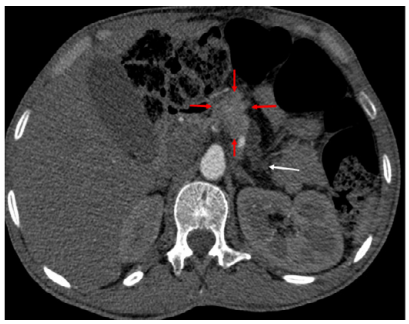

Figure 3: Axial CT scan of the abdomen on arterial phase shows homogenous mass (red arrow) enhancement of the body with tail atrophy (white arrow).

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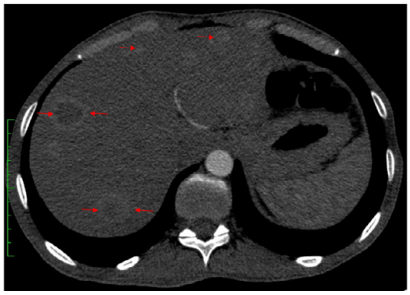

Figure 4: Axial CT scan of the abdomen on the arterial phase shows multiple liver nodules (red arrows) hypodense and heterogeneous with a hyper enhancing rim.

The CT investigation should also look for any vessel involvement and assess for liver metastasis [8]. These appear to be hypodense nodules with arterial phase enhancement and tend to overall resemble the imaging features of the primary tumor [9,10] Figure 4.

Surgical resection is the optimal strategy for the treatment of glucagonoma [9] with the disappearance of skin lesions and normalization of glucagon levels shortly after surgery. Patients who undergo resection have longer median survival than patients who did not receive surgery, even when diagnosed with later stages of the disease because of its slow growth [11].

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