

A Glucagonoma Accompanied with Chronic Necrolytic Migratory Erythema and Multiple Liver Metastasis: A Case Report

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1. Abstract

Glucagonoma is an extremely rare pNETs, usually presenting with glucagonoma syndrome, Necrolytic Migratory Erythema (NME) is mostly the first clinical manifestation of glucagonoma syndrome. Here we describe a rare case of glucagonoma syndrome accompanied with chronic necrolytic migratory erythema and multiple liver metastasis in a 23-year-old female patient. She complained of a progressive, pruritic and painful skin lesions for a 14-month duration. The skin lesions, including erythematous, brownish plaques and crusted erosions, were appeared in the lower extremities and her face. She also had cheilitis and glossitis. Laboratory investigations revealed markedly elevated blood glucagon concentration. A synchronous resection of pancreatic tumor (pancreaticoduodenectomy) and liver metastasis (enucleation) were then performed and pathological examination of excised tissue showed a Grade 3 pancreatic neuroendocrine tumor. Postoperative immunohistochemical staining examination confirmed the final diagnosis of glucagonoma. The skin lesions improved gradually 3 days after the surgery and postoperative plasma glucagon levels decreased obviously. She received an Octreotide Acetate Microspheres per 28d. The patient recovered uneventfully without tumor recurrence at a 2-month follow-up visit. The diagnosis of necrolytic migratory erythema is a matter of great importance, since it might be an auxiliary tool for the early detection of glucagonoma.

A 23-year-old girl was presented to hepatobiliary surgery department in July, 2020, in order to elucidate the etiology of a pancre-

atic mass (42mm×60mm×50mm) and multiple hepatic nodules detected by abdominal enhanced computed tomography (CT), which showed enhancement in the arterial phase (Figure 1) but hypodense in the portal phase. She complained of a progressive, pruritic and painful skin lesions affecting of fourteen months' duration. The skin lesions, including erythematous, brownish plaques and crusted erosions, were appeared in the lower extremities and her face. She also had cheilitis and glossitis. The topical steroids were used without clinical improvement. She denied abdominal pain, diarrhea, weight loss and the history of diabetes mellitus.

Laboratory investigations evidenced a relevant mild anemia with hemoglobin 95 g/L (normal range: 115-150 g/L), low serum albumin 31.2 g/L (normal range: 40-55), and a markedly elevated blood glucagon concentration (>800 pg/mL, normal range: 0-200 pg/mL). Glycosylated hemoglobin (HbA1c), C-reactive protein (CRP), liver and kidney parameters, blood coagulation test, neuron-specific enolase, carcinoembryonic antigen and carbohydrate antigen 19-9 were unremarkable. A biopsy of one of the liver nodules proved a metastatic grade 2 neuroendocrine tumor (Ki-67 index of 15%). For further staging, 68Ga-DOTATATE PET/CT was performed, which revealed an intense Ga-avid pancreatic mass replacing the head of pancreas and multiple Ga-avid lesions in the liver, with no additional distant metastases (Figure 2A, B).

A synchronous resection of pancreatic tumor (pancreaticoduodenectomy) and liver metastasis (enucleation) were then performed (Figure 3) and pathological examination of excised tissue

showed a Grade 3 pancreatic neuroendocrine tumor, with a mitotic count of 3 per 10 high-power fields, and Ki-67 proliferation index of 30 % (World Health Organization 2017 criteria). Postoperative immunohistochemical staining examination confirmed the diagnosis of glucagonoma and revealed positive staining for chromogranin A (CgA), synaptophysin (Syn), Somatostatin Receptor 2 (SSTR2). The skin lesions improved gradually 3 days after the surgery and postoperative plasma glucagon levels decreased to 315 pg/mL (Figure 4). She received an Octreotide Acetate Microspheres per 28d. The patient recovered uneventfully without tumor recurrence at a 2-month follow-up visit.

Glucagonoma is an extremely rare pNETs, with an estimated global incidence of one in 20 million people [1]. The average age at diagnosis for glucagonoma is 53.5 years, affecting men and women in almost equal proportions. It usually presents with glucagonoma syndrome, including dermatosis named Necrolytic Migratory Erythema (NME), diabetes mellitus, deep vein thrombosis and

depression. NME is mostly the first clinical manifestation of glucagonoma syndrome, which often starts as pruritic and painful erythema and gradually enlarge and coalesce to form bullous lesions [2]. Surgical removal is considered the only definitive and curative treatment for pancreatic glucagonoma and NME [3]. Optional operations include simple enucleation (< 2 cm) with peripancreatic lymph dissection, pancreaticoduodenectomy with peripancreatic lymph dissection, distal pancreatectomy with peripancreatic lymph dissection and splenectomy. However, more than half of all glucagonomas present with a metastatic disease, most commonly liver metastasis. It has reported that extended surgical resection of pancreatic neuroendocrine tumor and liver metastasis (more than 30% of the liver tissue retained) provides a more favorable outcome, because the tumor is slow-growing and the survival is improved. In addition, cytoreductive surgery for liver metastases could also reduce hormone levels and improve clinical symptoms as well as prognosis.



Figure 1: Multiple hepatic nodules detected abdominal enhanced computed tomography (CT) showed enhancement in the arterial phase but hypodense in the portal phase.

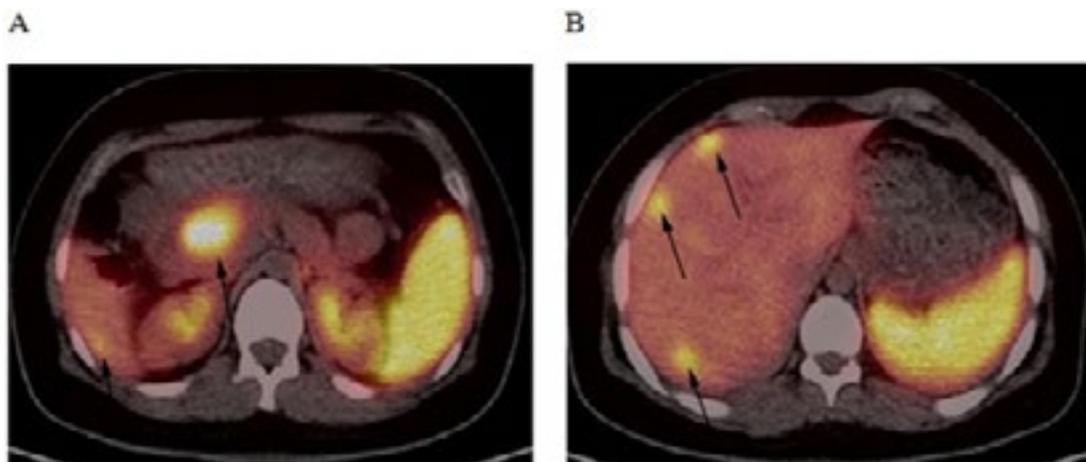


Figure 2: ^{68}Ga -DOTATATE PET/CT was performed to reveal an intense Ga-avid pancreatic mass replacing the head of pancreas (A) and multiple Ga-avid lesions in the liver (B), with no additional distant metastases.

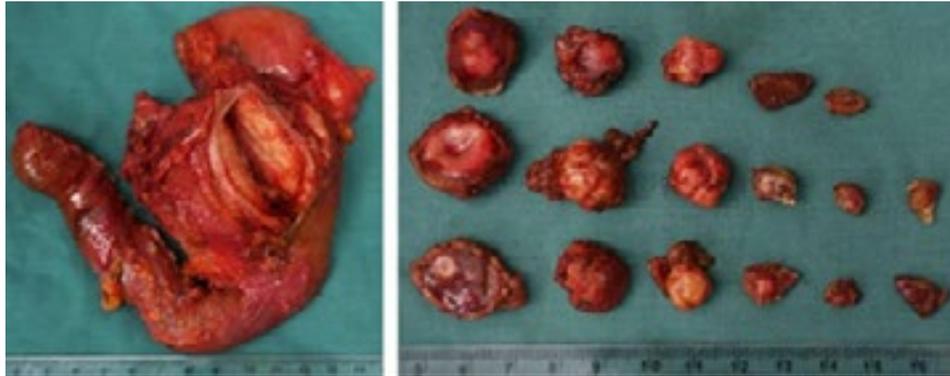


Figure 3: The excised tissue from a synchronous resection of pancreatic tumor (pancreaticoduodenectomy, A) and liver metastasis (enucleation, B).



Figure 4: The skin lesions (tongue, lower limbs and feet) improved gradually after the surgery.

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