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AGlucagonomaAccompanied with Chronic NecrolyticMigratoryErythema and Multiple Liver Metastasis: A Case Report

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1.Abstract

GlucagonomaisanextremelyrarepNETs,usuallypresentingwith glucagonomasyndrome,NecrolyticMigratoryErythema(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 livermetastasisina23-year-oldfemalepatient.Shecomplainedof aprogressive,pruriticandpainfulskinlesionsfora14-monthduration. The skin lesions, including erythematous, brownish plaques and crusted erosions, were appeared in the lower extremities and herface.Shealsohadcheilitisandglossitis.Laboratoryinvestigationsrevealedmarkedlyelevatedbloodglucagonconcentration.A synchronousresectionofpancreatictumor(pancreaticoduodenectomy) and liver metastasis (enucleation) were then performedand pathological examination of excised tissue showed a Grade3 pancreatic neuroendocrine tumor. Postoperative immunohistochemical staining examination confirmed the final diagnosis of glucagonoma.Theskinlesionsimprovedgradually3daysafterthe surgeryandpostoperativeplasmaglucagonlevelsdecreasedobviously. She received an Octreotide Acetate Microspheres per 28d. Thepatientrecovereduneventfullywithouttumorrecurrenceat a 2-month follow-up visit. The diagnosis of necrolytic migratory erythemaisamatterofgreatimportance,sinceitmightbeanaux- iliary tool for the early detection of glucagonoma.

A 23-year-old girl was presented to hepatobiliary surgery departmentinJuly,2020,inordertoelucidatetheetiologyofapancreatic 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, pruriticandpainfulskinlesionsaffectingoffourteenmonths'duration.Theskinlesions,includingerythematous,brownishplaques 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 bloodglucagonconcentration(>800pg/mL,normalrange: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 antigen19-9wereunremarkable.Abiopsyofoneofthelivernodulesprovedametastaticgrade2neuroendocrinetumor(Ki-67indexof15%).Forfurtherstaging,68Ga-DOTATATEPET/CTwas performed,whichrevealedanintenseGa-avidpancreaticmassreplacing 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(Figure3)andpathologicalexaminationofexcisedtissue

showedaGrade3pancreaticneuroendocrinetumor,withamitotic countof3per10high-powerfields,andKi-67proliferationindex of30%(WorldHealthOrganization2017criteria).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 315pg/mL(Figure4).ShereceivedanOctreotideAcetateMicrospheresper28d.Thepatientrecovereduneventfullywithouttumor recurrence at a 2-month follow-up visit.

GlucagonomaisanextremelyrarepNETs,withanestimatedglobal incidence of one in 20 million people [1]. The average age at diagnosisforglucagonomais53.5years,affectingmenandwomen in almost equal proportions. It usually presents with glucagono- ma syndrome, including dermatosis named Necrolytic Migratory Erythema(NME),diabetesmellitus,deepveinthrombosisand depression. NME is mostly the first clinical manifestation of glucagonomasyndrome,whichoftenstartsaspruriticandpainfulerythemaandgraduallyenlargeandcoalescetoformbullouslesions [2].Surgicalremovalisconsideredtheonlydefinitiveandcurative treatmentforpancreaticglucagonomaandNME[3].Optionaloperations include simple enucleation (< 2 cm) with peripancreatic lymph dissection, pancreaticoduodenectomy with peripancre-atic lymph dissection, distal pancreatectomy with peripancreatic lymphdissectionandsplenectomy.However,morethanhalfofall glucagonomas present with a metastatic disease, most commonly liver metastasis. It has reported that extended surgical resectionof 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 couldalsoreducehormonelevelsandimproveclinicalsymptoms as well as prognosis.

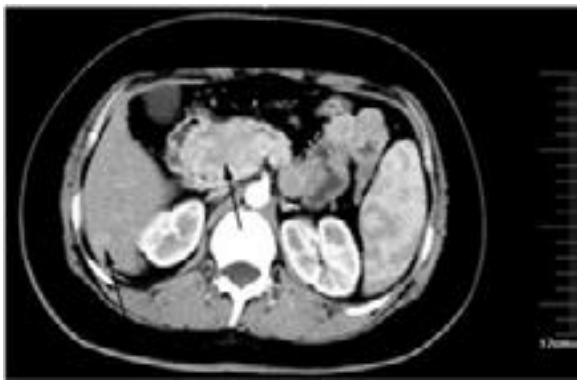

Figure1: Multiplehepaticnodulesdetectedabdominalenhancedcomputedtomography(CT)showedenhancementinthearterialphasebuthypodense in the portal phase.

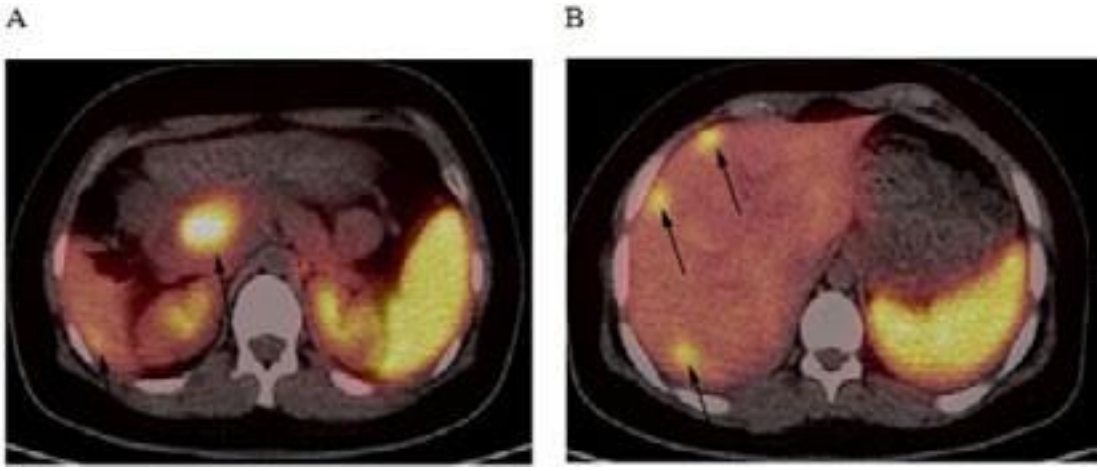

Figure 2: 68Ga-DOTATATE PET/CT was performed to reveal an intense Ga-avid pancreatic mass replacing the head of pancreas (A) and multiple Ga-avid lesions in the liver (B), with no additional distant metastases.

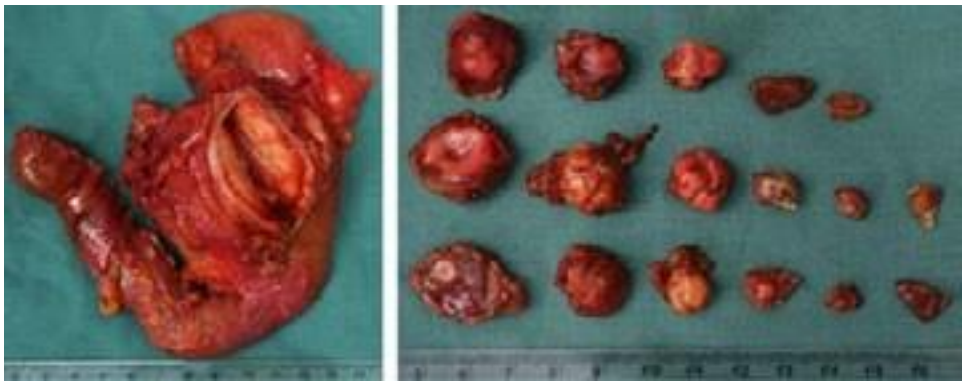

Figure3: Theexcisedtissuefromasynchronousresectionofpancreatictumor(pancreaticoduodenectomy,A)andlivermetastasis(enucleation,B).

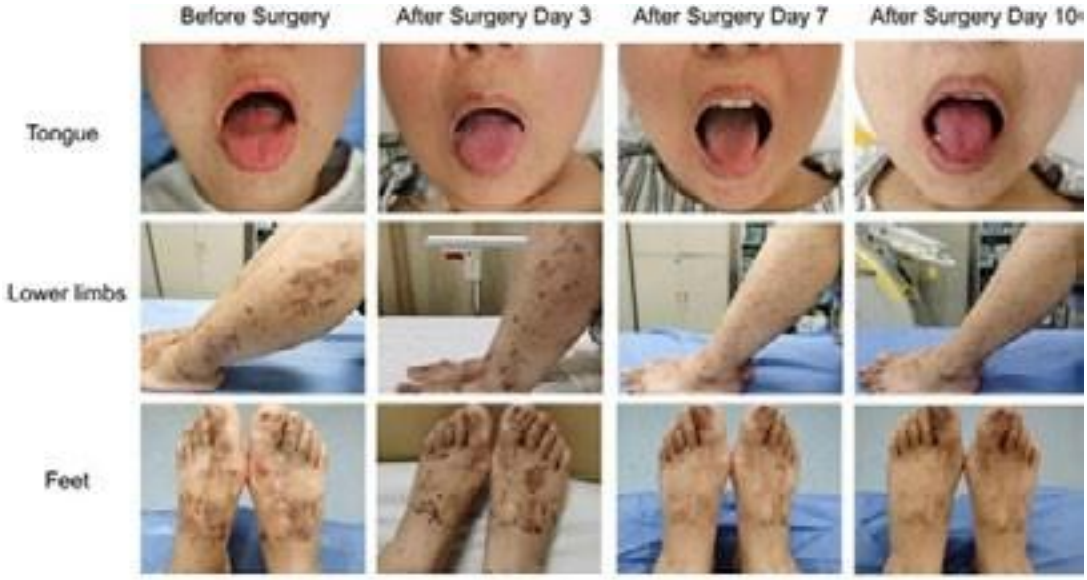

Figure4: The skinlesions (tongue,lowerlimbs andfeet) improvedgraduallyafter thesurgery.

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