Letter to the Editor

Robotic-assisted hepatopancreatoduodenectomy and resection of pancreatic tail for one multiple endocrine neoplasia type 1 secreting multiple hormones

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One 42-year-old female suffered from recurrent weakness and diarrhea for 2 years. She had a history of thyroidectomy and patent foramen ovale. At her local hospital, she was diagnosed and treated as transient ischemic attack, however her symptoms were not relieved and she was eventually referred to our hospital. Laboratory test showed that the fasting serum gastrin (>1,000.0 ng/L; normal: 28.1–106.5 ng/L), the insulin (7.54 μU/mL; normal: 1.9–23.0 μU/mL), C-peptide (1.92 nmol/L; normal: 0.4–1.7 nmol/L), and isolated parathyroid hormone (214.3 pg/mL; normal: 12.0–88.0 pg/mL) were elevated, while the fasting serum glucose (2.0 mmol/L; normal: 3.9–7.8 mmol/L) was decreased. The level of growth hormone, sex hormone, adreno-cortico-tropic-hormone, adrenal hormone, serum calcium and phosphorus, and tumor markers (carcinoembryonic antigen, carbohydrate antigen 199, and carbohydrate antigen 125) were normal. Magnetic resonance imaging scan showed one tumor at the pancreatic head (25 mm × 21 mm in size, *Figure 1A*) and one lesion at the tail (8 mm × 7 mm in size, *Figure 1B*), and a lesion at liver segment IV (48 mm × 43 mm in size, *Figure 1C*). Pancreatic neuroendocrine tumors (pNETs) with liver metastasis was considered. Furthermore, ^{18}F-fluorodeoxyglucose (FDG) and ^{68}Ga-DOTA, 1-Nal3-octreotide (DOTANOC) positron emission tomography-computed tomography (PET-CT) scan showed that the tumor at the pancreatic head had slight elevation of glucose metabolism with maximum standardized uptake value (SUVmax) of 2.9, but no uptake of ^{68}Ga-DOTANOC, the tumor at the pancreatic tail had no uptake of both ^{18}F-FDG and ^{68}Ga-DOTANOC, and the tumor at liver segment IV had no uptake of ^{18}F-FDG, but remarkable uptake of ^{68}Ga-DOTANOC with SUVmax of 25.5. In addition, two lesions at parathyroid gland was found with no obvious uptake of both ^{18}F-FDG and ^{68}Ga-DOTANOC. Ultrasound-guided needle biopsy and immunohistochemical staining revealed that the tumor at the pancreatic head was insulinoma, whereas the tumor at the liver was gastrinoma. Multiple endocrine neoplasia type 1 (MEN1) germline mutation analysis was performed and confirmed. Collectively, MEN1 with secretion of multiple hormones was diagnosed preoperatively.

After multidisciplinary discussion, lesions at the parathyroid gland were advised to keep regular observation due to the normal level of the blood calcium and phosphorus. For the pancreatic tumors and liver metastasis, robotic-assisted pancreatoduodenectomy (RPD) with left hemihepatectomy and spleen-preserving distal pancreatectomy (SPDP) was recommended to remove all pancreatic lesions and the liver metastasis. The operation was completed uneventfully (*Figure 1D-1F*), with intraoperative blood loss of 200 mL and no blood transfusion. Postoperative pathology demonstrated three different function neuroendocrine tumors (NETs), including insulinoma at pancreatic head (G2), glucagonoma at pancreatic tail (G1), and gastrinoma at liver (G2). The lymph nodes harvested (0/20) and surgical margins were negative. Complete relief of symptoms was achieved postoperatively. The patient recovered well and was discharged on postoperative day 15. No recurrence or metastasis was found during the follow-up after 18 months

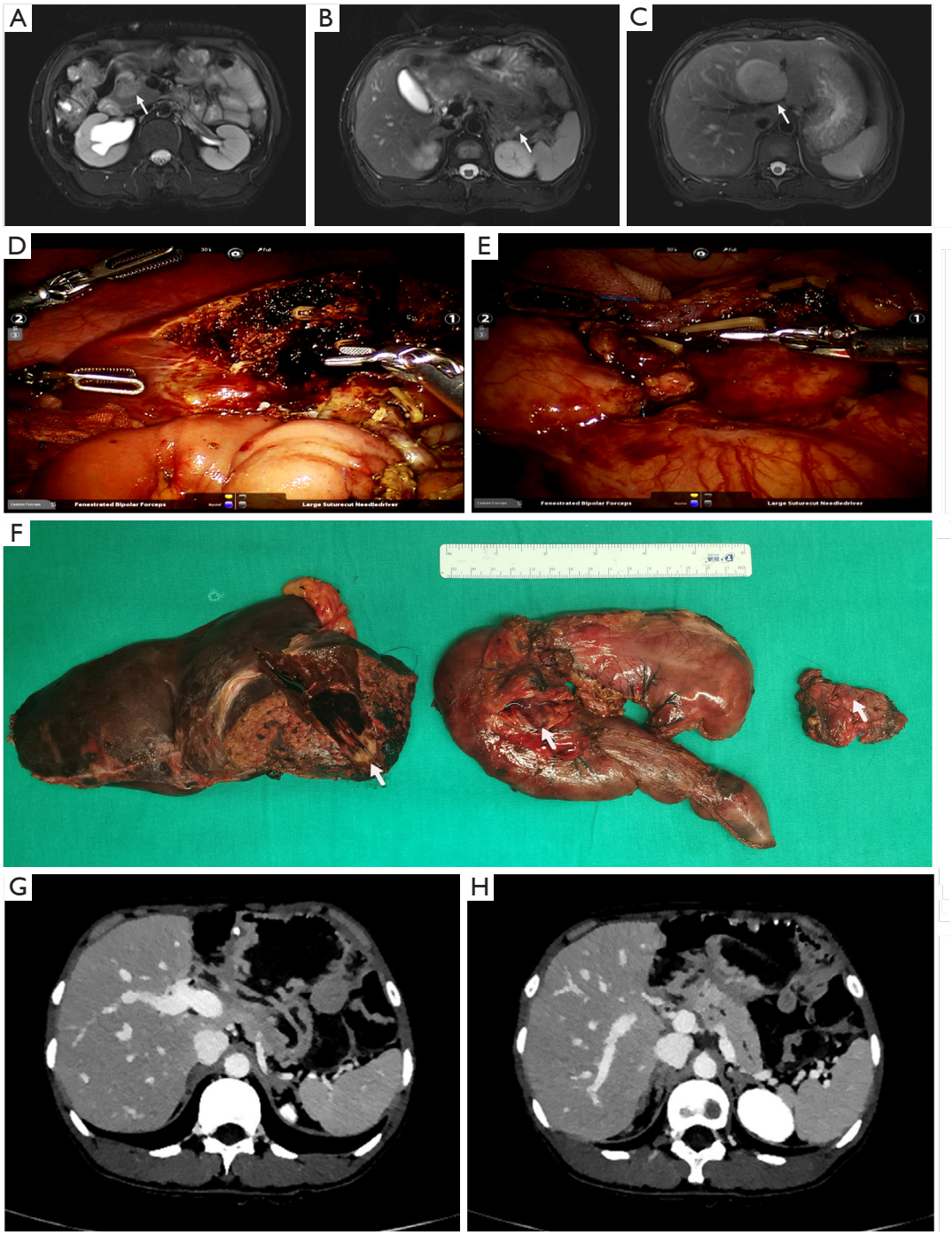

Figure 1 MRI, operation, tumor specimen, and CT of the case. (A-C) MRI showed multiple pancreatic lesions and liver metastasis, including lesion (white arrows) at the pancreatic head (A), pancreatic tail (B), and liver segment IV (C). (D) The picture of completion of pancreatoduodenectomy and left hemihepatectomy. (E) The picture of completion of splenic-preserving distal pancreatectomy. (F) The resected specimens. The left one was the liver metastasis (white arrow), the middle one was the lesion at pancreatic head (white arrow), and the right one was the lesion at pancreatic tail (white arrow). (G,H) CT scan showed no recurrence or metastasis after 18 months postoperatively. MRI, magnetic resonance imaging; CT, computed tomography.

postoperatively (*Figure 1G,1H*).

MEN1 is a rare autosomal dominant and hereditary endocrine tumor syndrome caused by mutation of the MEN1 gene with an estimated prevalence of (1–10)/100,000 (1). As the second most common manifestation in MEN1 (30–90%), duodenopancreatic neuroendocrine tumor is the second leading cause of death (2).

Resection of a MEN1-related insulinoma was considered effective in relieving hypoglycemic symptoms (3). This patient had three tumors at the pancreatic head, tail and liver segment IV, with secretion of insulin, glucagon, and gastrin, respectively. It was extremely rare. To effectively treat the patient, all three functioning tumors should be excised simultaneously. However, simultaneous radical resection of multifocal pNETs and liver tumor was technically demanding and challenging (4). With the advantages of dextrous manipulation of da Vinci surgical system (5,6), the patient was successfully treated with robotic-assisted pancreatoduodenectomy with resection of pancreatic tail and plus left hemihepatectomy, and the postoperative course was uneventful.

Most gastrinomas are localized to the duodenum (70–90%) or the pancreas (2–30%) (7). Ectopic sites of gastrinoma such as liver were rarely reported. In this case, since the expression of Gastrin was not detected in the pancreatic tumors, primary liver gastrinoma was considered. Parathyroid involvement is the most common manifestation in MEN1, affecting more than 90% of patients with overt MEN1 (8). Surgery is indicated in patients with severe hypercalcaemia. In this case, hyperparathyroidism was kept for regular observation because of the normal level of serum calcium and phosphorus.

In summary, we herein reported one rare case of MEN1 secreting three different hormones. After performing robotic-assisted pancreatoduodenectomy with left hemihepatectomy and spleen-preserving distal pancreatectomy, the symptoms were completely relived, and no recurrence was found during the postoperative follow up. For such patients with symptomatic MEN1 and liver metastasis, curative-intent surgery can be considered.

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Footnote

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Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://hbsn.amegroups.com/article/view/10.21037/hbsn-23-315/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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