CASE REPORT

Metastatic neuroendocrine tumor with metastases to the right liver in a patient with absent left portal vein

Reid C. Mahoney1,*, Kyrillos Awad1 and Gregorio Maldini1,2

1Department of Surgery, University of Hawai‘i John A. Burns School of Medicine, Honolulu, HI, USA and
2Department of Surgery, Straub Medical Center, Honolulu, HI, USA

*Correspondence address. Reid C. Mahoney, 1356 Lusitana St. #610, Honolulu, HI 96813, USA.
Tel: 630-346-6216; Fax: 808-586-3022; E-mail: rmahoney@hawaii.edu

Abstract

The patient is a 50-year-old female that underwent routine screening colonoscopy during which she was found to have a neuroendocrine tumor in the right colon. The patient underwent computed tomography and magnetic resonance imaging scans that demonstrated metastatic disease in segments 5 and 7 of the liver. Notably, the patient was found to have an absent left portal vein. The metastatic lesions abut the right portal vein; the right portal vein also supplies the left lobe of the liver in place of an absent left portal vein. She underwent a laparoscopic-assisted right hemicolectomy to remove the primary tumor. The patient recovered uneventfully from surgery and is currently being monitored by a multidisciplinary team regarding her metastatic disease. Neuroendocrine tumors can cause long-term effects on health and ultimately death if left untreated. We present a case of metastatic midgut neuroendocrine tumor that has metastasized to the liver in a patient with absent left portal vein.

INTRODUCTION

Gastroenteropancreatic neuroendocrine tumors are divided by the World Health Organization (WHO) as grades 1, 2 or 3 based on mitoses and Ki-67 index [1]. The SEER database demonstrates a non-pancreatic neuroendocrine tumor incidence of 4.7 per 100 000 patients, and recent age-adjusted data is as high as 5.2/100 000 [2]. Separately, portal vein anomalies are rare, and absence of the portal vein bifurcation is even rarer, estimated at 0.3–2% of all portal vein anomalies [3]. We present a case of metastatic neuroendocrine tumor in a patient with an absent left portal vein.

CASE DESCRIPTION

The patient is a 50-year-old female with a history of hypertension, hyperlipidemia and obesity that presented to an outpatient gastroenterologist for routine screening colonoscopy. The patient was found to have a lesion, measured at ∼2.5 centimeters in size, near the ileocecal valve. Biopsies were taken at the time of colonoscopy, which demonstrated evidence of a well-differentiated neuroendocrine tumor with invasion into the submucosa. There was no increased mitotic activity (Ki-67 <3%). The patient was referred to surgery clinic for further evaluation.

Revised: March 29, 2021. Accepted: April 24, 2021

Published by Oxford University Press and JSCR Publishing Ltd. All rights reserved. © The Author(s) 2021.
This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (http://creativecommons.org/licenses/by-nc/4.0/), which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is properly cited. For commercial re-use, please contact journals.permissions@oup.com
On assessment, the patient denied any symptoms related to her neuroendocrine tumor, including flushing, increased sweating, increased heart rate, wheezing, shortness of breath, diarrhea, weight loss or appetite changes. The only significant family history was a paternal and maternal grandfather with colon cancer. Imaging results were significant for computed tomography (CT) of chest demonstrating very small, but multiple, pulmonary nodules. A CT abdomen and pelvis demonstrated the known neuroendocrine tumor near the ileocecal valve (Fig. 1) as well as two poorly visualized liver lesions (Figs 2–4). The portal vein lacked normal left and right bifurcation; there was a circumferential right portal vein, which coursed anteriorly and superiorly and ultimately to the left lobe of the liver.

After workup was completed, the patient was consented for laparoscopic right hemicolectomy for removal of the primary tumor. The procedure was performed in a lateral to medial fashion and a hand-sewn extracorporeal anastomosis was performed after removal of the specimen. The operation was uneventful and the patient recovered appropriately in the immediate postoperative period. She was discharged home tolerating a regular diet and having normal bowel function. Final pathology demonstrated a 3 × 2 × 1.7 cm well-differentiated neuroendocrine tumor. Margins were clear and 8/19 lymph nodes were positive. There were 1/10 mitoses per high power field. Ki-67 was noted to be <1%. The final stage was T3N1M1 (stage IV) by the grading of American Joint Committee on Cancer (AJCC), and the tumor was G1 based on WHO classification.

The patient was seen postoperatively in clinic and has continued to recover well. She underwent repeat imaging that demonstrated stability in the size and location of metastatic disease. She remains asymptomatic. Oncology will continue to follow the patient with yearly bloodwork and repeat imaging. Discussions regarding the next step in management are ongoing, with the understanding that if she develops symptoms or demonstrates enlargement of the metastatic lesions on imaging, the need for intervention becomes more acute.

**DISCUSSION**

The majority (~55–65%) of metastatic neuroendocrine tumors arise in the gastrointestinal tract [4], most of which are found in the small intestine; ~7.5% of gastrointestinal neuroendocrine tumor tumors are found in the colon [5]. The rate of metastatic neuroendocrine tumor at initial presentation is ~22% with half of those having unknown primaries [4]. Treatment for metastatic
among patients with resectable disease includes resection of primary tumors in patients that are appropriate candidates for surgery [5].

Management of neuroendocrine tumor metastases can be more challenging. Current treatment modalities include metastasectomy, cytoreductive surgery, hepatic arterial embolization, radiofrequency ablation and multiple medication-based regimens including somatostatin analogues and interferon alpha [5–7]. Radiofrequency ablation can be used in hepatic lesions with good outcomes, even near portal and hepatic veins, which act as a heat sink [8]. Irreversible electroporation (IRE) is another treatment option for metastatic disease to the liver and works by inducing cell death via disruption of the cell membrane with electrical pulses, sparing the extracellular matrix and surrounding tissue [9].

There are several portal vein branching variants and congenital anomalies described in the literature. Variants in normal branching pattern occur in as many as 20% of the population, with trifurcation of the main portal vein being the most common variant [10]. Agenesis of the main portal branches is the most common congenital anatomic variant of portal vein anatomy; however, this is frequently associated with lobar agenesis [10]. True absence of the portal vein bifurcation is a very rare anomaly, estimated to represent 0.3–2% of all portal vein anomalies [3].

There have been published series in which a hepatic resection has been performed for patients with an absent left portal vein with results varying from minor complications to death [12]. Although rare, neuroendocrine tumor crisis has been described during hepatic resection of metastatic neuroendocrine tumor, and portal vein occlusion is often performed during the operation to avoid vasoactive release from the tumor [13]. Due to this patient’s aberrant portal anatomy, metastasectomy becomes exceedingly difficult. For these reasons, the decision was made to perform an upfront resection of our patient’s primary tumor and subsequently pursue a multi-disciplinary team approach to her metastatic disease. If the lesions increase in size, the operative plan is lesion resection with preoperative radiofrequency ablation. The patient’s nearby portal vein should act as a heat sink during radiofrequency ablation and would be expected to remain patent. Despite the difficulty of the operation, studies have shown increased survival for patients undergoing metastasectomy for neuroendocrine tumors [11].

This case presents a challenging clinical scenario involving management of metastatic liver disease with aberrant portal vein anatomy. It is an extremely rare anatomic anomaly that has significant clinical consequences for the patient. The lesions abut the right portal vein, and resection risks compromise to global hepatic blood flow.

**CONFLICT OF INTEREST STATEMENT**

None declared.

**REFERENCES**

1. Pasaoglu E, Dursun N, Ozyalvaci G, Hachhasanoglu E, Behzatoglu K, Calay O. Comparison of World Health Organization 2000/2004 and World Health Organization 2010 classifications for gastrointestinal and pancreatic neuroendocrine tumors. Ann Diagn Pathol 2015;19:81–7.

2. Yao JC, Hassan M, Phan A, Dagohoy C, Leary C, Mares J, et al. One hundred years after “carcinoid”: epidemiology of and prognostic factors for neuroendocrine tumors in 35,825 cases in the United States. J Clin Oncol 2008;26:3063–72.

3. Sahoo RK, Panda SK, Bahinipati P. Absent portal vein bifurcation: a rare variant and its clinical significance. Anat Cell Biol 2014;47:210–3.

4. Shuch, B, Bratslavsky, G, Linehan W.M., Srinivasan R. Sarcomatoid Renal Cell Carcinoma: A Comprehensive Review of the Biology and Current Treatment Strategies. The Oncologist 2012;17:46–54.

5. Byrne RM, Pommier RF. Small bowel and colorectal carcinoids. Clin Colon Rectal Surg 2018;31:301–8.

6. Åkerström G, Hellman P, Hessman O. Midgut carcinoid tumours: surgical treatment and prognosis. Best Pract Res Clin Gastroenterol 2005;19:717–28.

7. Diaco DS, Hajarizadeh H, Mueller CR, Fletcher WS, Pommier RF, Woltering EA. Treatment of metastatic carcinoid tumors using multimodality therapy of octreotide acetate, intra-arterial chemotherapy, and hepatic arterial chemoembolization. Am J Surg 1995;169:523–8.

8. Gambin TC, Christians K, Pappas SG. Radiofrequency ablation of neuroendocrine hepatic metastasis. Surg Oncol Clin N Am 2011;20:273–9.

9. Scheffer HJ, Melenhorst MCAM, Echenique AM, Nielsen K, van Tilborg AAM, van den Bos W, et al. Irreversible electroporation for colorectal liver metastases. Tech Vasc Interv Radiol 2015;18:159–69.

10. Gallego C, Velasco M, Marcuello P, Tejedor D, De Campo L, Friera A. Congenital and acquired anomalies of the portal venous system. Radiographics 2002;22:141–59.

11. Givi B, Pommier SEJ, Thompson AK, Diggs BS, Pommier RF. Operative resection of primary carcinoid neoplasms in patients with liver metastases yields significantly better survival. Surgery 2006;140:891–8.

12. Charny MAO, Nagorney DM, Clark DF, O’Brien TD, Turner JD, Marienau ME, et al. Partial hepatic resections for metastatic neuroendocrine tumors: perioperative outcomes. J Clin Anesth 2018;51:93–6.