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Case Report

Neuroendocrine tumor of the hepatic flexure: a rare colonic tumor

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ABSTRACT: Neuroendocrine tumors (NETs) are rare, particularly common in the rectum. NETs of the colon (not including the appendix and rectum) are most commonly found in the cecum, and hepatic flexure involvement is rather infrequent. The clinical presentation of colonic NETs is dependent on the primary site. Many are discovered either at the time of screening or during the investigation of abdominal pain or anemia. Here we present a challenging case of a hepatic flexure colonic NETS of 26 -year-old male treated by laparoscopic right hemicolectomy.

KEY WORDS: *Neuroendocrine tumors; Colonoscopy; Surgery; Immunohistochemical test*

INTRODUCTION

Colonic neuroendocrine tumors (NETs) are extremely rare. The incidence of colonic NETs is 1.0-2.0 per 10^6 and has remained constant over the last five decades[1]. Langhans first described an intestinal carcinoid tumor in 1867[2]. However, Lubarsch who described its histologic features classified it as carcinoma in 1888[3]. In 2000, the World Health Organization (WHO) revised an earlier classification of these tumors by Williams and Sandler to recognize their growing heterogeneity as well as neuroendocrine origin[4]. Most colonic NETs are diagnosed via colonoscopy. Correct diagnosis of NETs is as important to treat as prognosis. Due to the rarity of colonic NETs, we aimed to report the case of 26-year-old male with hepatic flexure NETs.

CASE DETAILS

A 26-year-old male patient presented to the surgical outpatient department with a chief symptom of recurrent pain in the right side of his abdomen during the previous year. The patient reported weakness, lethargy, anorexia and

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occasional black stool." On examination, a lump was palpable in the right upper abdominal quadrant. His laboratory results showed severe microcytic hypochromic anemia with hemoglobin 5.2gm %. The fecal occult blood test was positive. Ultrasonography of the abdomen suggested thickening over the hepatic flexure and ascending colon with multiple mesenteric lymph nodes. Tuberculosis of the GI tract was suspected. We performed a colonoscopy after preparation of the bowel, and found an ulceroproliferative lesion at the hepatic flexure of the colon (**Figure 1**).

Figure 1: An ulceroproliferative lesion at the hepatic flexure of the colon

The biopsy was suggestive of poorly differentiated adenocarcinoma. A computed tomography scan of the abdomen showed an asymmetric circumferential short segment of non-homogeneously enhanced wall thickness up to 20 mm at the hepatic flexure and adjacent ascending colon. The thickened wall caused a luminal narrowing with adjacent extensive fat stranding, which suggested the possibility of a malignant mass lesion (**Figure 2**).
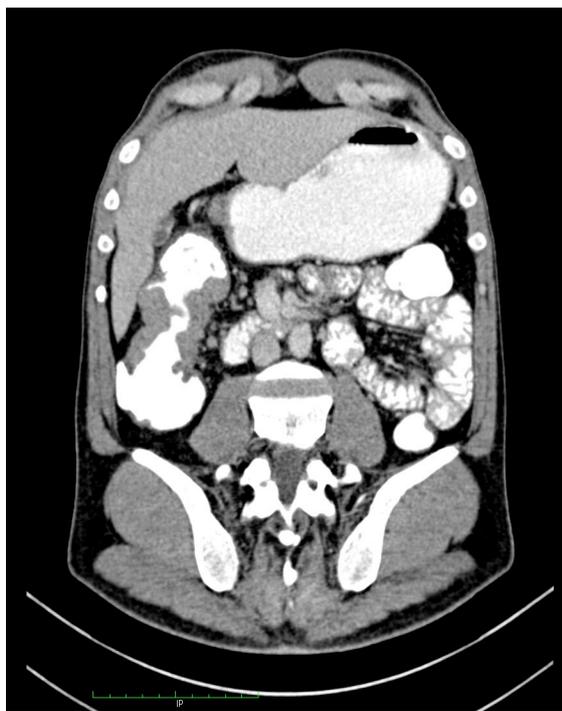
Figure 2: A computed tomography scan of the abdomen showed an asymmetric circumferential short segment of non-homogeneously enhanced wall thickness up to 20 mm at the hepatic flexure and adjacent ascending colon with luminal narrowing.

Carcinoembryonic antigen was normal. A laparoscopy-assisted right hemicolectomy was performed. Histopathological examination of the specimen revealed nuclear hyperchromasia with strips of stippled chromatin cells arranged in sheets with numerous mitoses (**Figure 3**). Therefore, diagnosis of a neuroendocrine tumor (NET) was suspected. Immunohistochemical tests were advised to confirm the diagnosis. The tumor was assessed to identify histological features suggestive of high-frequency microsatellite instability (MSI-H), which was negative in the index case. Immunohistochemical findings were positive for Synaptophysin and Chromogranin (Cg) and negative for PANCK, which confirmed the diagnosis of neuroendocrine tumor. A tumor was categorized stage IIA according to TNM staging and G2 according to grading by WHO 2010. It was well-differentiated small cell neuroendocrine

tumor. The patient was referred to the oncologist for further management. We followed a patient for 1 year without any recurrence.
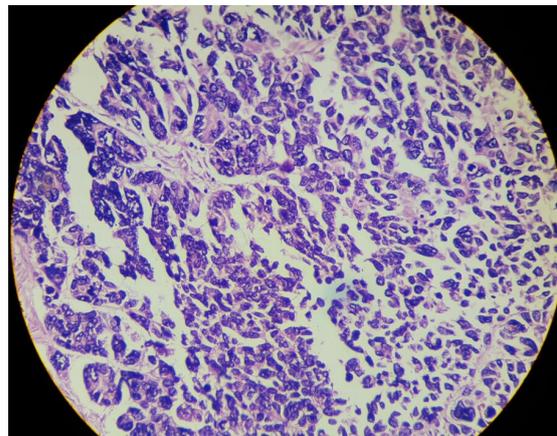

Figure 3: 40X H and E stain showed nuclear hyperchromasia with strips of stippled chromatin cells arranged in sheets with numerous mitoses.

DISCUSSION

The colon proximal to the rectum most commonly gives rise to poorly differentiated small cell carcinomas, but there are more commonly large cell variants or intermediates between the two sizes.

NETs may be mislabeled as adenocarcinoma, which affects management and the understanding of the prospects for survival[5]. The prognosis of a NET can be uncertain, partly due to the variability of their secretions. Nonfunctional NETs may present with pain, bleeding, and obstruction, but without hormone secretion, then start producing hormones, becoming syndromic[6]. NETs are classified based on their embryonic origin and vascular supply (foregut, midgut and hindgut). The substance secreted by NETs may vary; even metastases may secrete substances different from the primary tumor. NETs can also cause paraneoplastic syndrome because of the secretion of entirely different substances not related to their original cell properties[7]. NETs of the colon are the largest tumors and have the poorest prognosis among all the gastroenteropancreatic - NETs, as they commonly metastasize to the liver.

Correct diagnosis of NETs is as important to treat as to prognosis; treatment options for NETS are different from those of poorly differentiated adenocarcinoma. Colonic NETs are usually treated surgically either local resection or conventional surgery. Optimal management requires multidisciplinary treatment for which options are limited. However, everolimus plus octreotide long-acting repeatable has shown significant benefits

and improved outcomes for patients with advanced colorectal neuroendocrine tumors[8].

Most colonic NETs are diagnosed via colonoscopy. Computed tomography and magnetic resonance imaging are more sensitive and specific for improved diagnosis and follow up of neuroendocrine tumors (NETs). As in this case, histological examination and immunohistochemical tests may be essential to arrive at the correct diagnosis.

CONCLUSION

NETs can arise in different organs. Colonic NETs are exceptionally rare especially hepatic flexure of the colon. Immunohistochemical tests are necessary to differentiate NETs from poorly differentiated adenocarcinomas. This differentiation is essential to ensure accurate prognosis and correct management.

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