Case Report

Neuroendocrine Carcinoma of the Lower Rectum: A Case Report

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Background: *Rectal neuroendocrine carcinoma is a rare disease with a prognosis worse than rectal adenocarcinoma. The authors report a case of successful ultra-low anterior resection in well-differentiated neuroendocrine carcinoma of the lower rectum.*

Case Report: *A 38-year-old man presented with chronic constipation for six years. Digital rectal examination, colonoscopy and pelvic MRI demonstrated a 5-cm. intramural mass with a smooth mucosa at the posterior aspect of the lower rectum; 4 cm above the anal verge. No liver metastasis was demonstrated by an abdominal CT scan. The patient underwent an uneventful ultra-low anterior resection. The operative time was 195 minutes and blood loss was 310 milliliters. The pathology report showed well-differentiated neuroendocrine carcinoma, presence of angio-lymphatic invasion and metastatic carcinoma in one out of twenty-six regional lymph nodes. The patient received postoperative adjuvant radiation and chemotherapy. No recurrent tumor was detected after 14 months of follow-up.*

Conclusion: *Although high recurrence rates are noted, radical oncologic resection followed by adjuvant chemo-radiation is a standard treatment in lower rectal neuroendocrine carcinoma. Long-term follow-up of this patient is still needed.*

Keywords: *Neuroendocrine carcinoma, Neuroendocrine tumor, NETs, Surgery, Rectum*

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Rectal neuroendocrine carcinoma is a rare disease but the incidence is increasing along with rectal carcinoid tumor, variants of rectal neuroendocrine tumors (NETs). According to the SEER database, rectal carcinoid tumors compromise 27 percent of all gastrointestinal NETs and 16 percent of all NETs[1]. Rectal carcinoid is primarily diagnosed in young patients with the mean age at diagnosis of 56 years old. Approximately, half of these patients are asymptomatic with incidental finding during a surveillance colonoscopy. The risk of malignant behavior is closely related to the tumor size and the depth of invasion. Small, submucosal rectal NETs are associated with a very low malignant potential. Tumors larger than two centimeters or those invading the muscularis propia are associated with a significantly higher risk of malignant transformation and metastasis. The prognosis of rectal neuroendocrine carcinoma is poorer than rectal adenocarcinoma[2]. A multidisciplinary approach is recommended for optimal treatment. Management decisions are commonly based on experience and expert recommendations due to the lack of randomized prospective clinical trials. Therefore, we report a case of successful ultra-low anterior resection in well-differentiated neuroendocrine carcinoma of the lower rectum.

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Case Report

A 38-year-old man's medical record, diagnostic imaging, endoscopic report and pathology reports were reviewed. He presented with chronic constipation for six years without rectal bleeding, pain or change in bowel habits. Digital rectal examination and colonoscopy demonstrated a 5-cm intramural mass with smooth mucosa at the posterior aspect of his lower rectum; 4 cm above the anal verge (Fig. 1). The biopsy reported a carcinoid tumor. A pelvic MRI revealed a well-defined rounded 5.3x6.2x4.6 cm^3 retrorectal mass with hypo-signal intensity on T1W, markedly high signal intensity on T2W, and intense enhancement after Gadolinium administration (Fig. 2).

His abdominal CT scan revealed no liver metastasis.

The patient underwent an uneventful ultra-low anterior resection with double stapled side to end colo-anal anastomosis (Fig. 3). The operative time was 195 minutes and blood loss was 310 milliliters. The pathology reports revealed a 5x4.5x4.5 cm^3 well-differentiated neuroendocrine carcinoma, presence of angio-lymphatic invasion and metastatic carcinoma in one out of twenty-six regional lymph nodes (Fig. 4). The patient received postoperative adjuvant radiation and chemotherapy (5-FU and Leucovorin). No recurrent tumor was detected after 14 months of follow-up.

Discussion

Rectal neuroendocrine tumor is rare and the prognosis in the malignant variant is worse than rectal adenocarcinoma. A complete resection is the curative option for a localized lesion. However, the benefits of radical surgery for advanced disease are not clear. Treatment is determined by the size of the tumor. Tumors which are small (<1 centimeter) and confined to the mucosa or submucosa can be managed with colonoscopic resection because of their low risk of malignancy and metastasis (<3 percent)[1,3]. Transanal excision is performed for wide base or intermediate sized (1 to 2 centimeter) distal rectal tumors confined to the submucosa or small tumors invading to the muscularis propia without lymph node metastases. Transanal Endoscopic Microsurgery (TEM) can be offered for tumors in the proximal rectum.

Tumors larger than two centimeters, invading the muscularis propia or with local lymph node involvement should be managed with standard rectal resection i.e. low anterior resection or abdomino-perineal resection similar to rectal adenocarcinoma because of their high risk of metastasis (60 to 80 percent)[3,4]. The role of a chemotherapeutic agent is still limited due to the lack of evidence. In ENET guidelines, chemotherapy is appropriate for 1) poorly

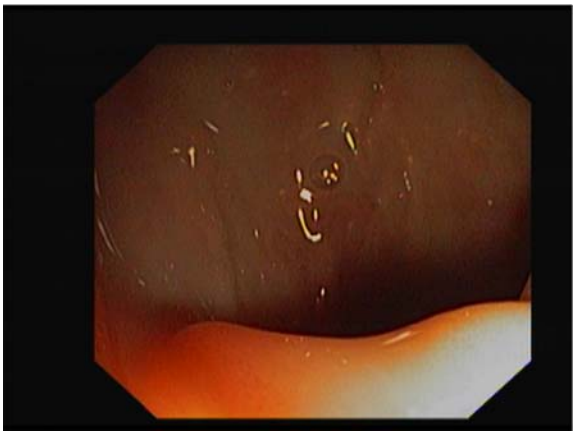

Fig. 1 Colonoscopy.

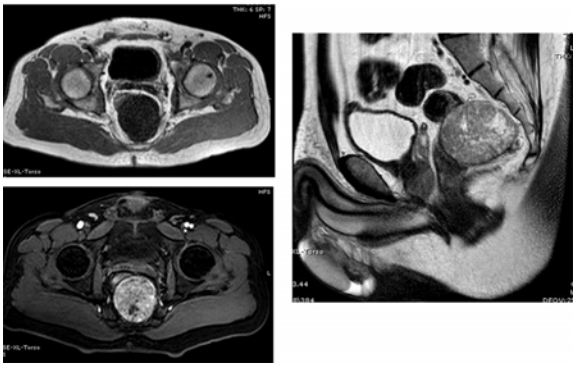

Fig. 2 Pelvic MRI.

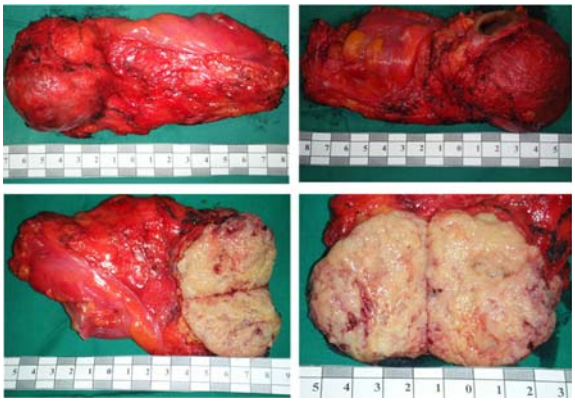

Fig. 3 Gross specimen of ultra-low anterior resection.

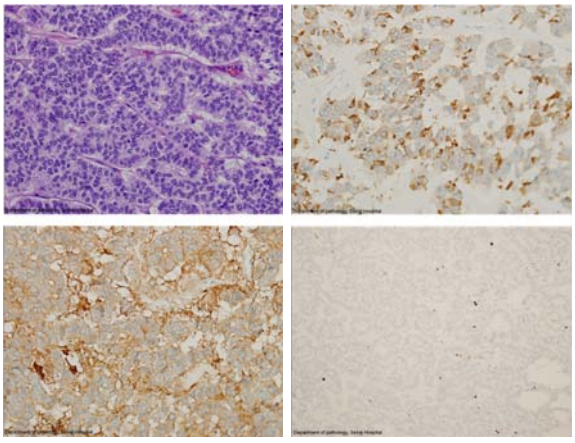

Fig. 4 Histology.

differentiated or high grade rectal NETs or rectal neuroendocrine carcinoma, 2) rectal NETs with lymph node involvement or lymphatic invasion[3]. Because of a substantially high risk of recurrence, rectal NETs patients who underwent surgery need to be followed-up in most cases especially for a tumor size of more than two centimeters, tumor invading muscularispropia or tumors with adverse features.

Conclusion

To date, radical oncologic resection followed by adjuvant chemo-radiation is standard treatment in lower rectal neuroendocrine carcinoma. However long-term follow-up of these patients is still needed.

What is already known on this topic?

High recurrence is noted after treatment of lower rectal neuroendocrine carcinoma.

What this study adds?

Radical oncologic resection followed by adjuvant chemo-radiation is contemporarily adequate treatment in lower rectal neuroendocrine carcinoma.

Acknowledgements

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Potential conflicts of interest

None.

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รายงานผู้ป่วย Neuroendocrine Carcinoma ของลำไส้ตรงส่วนล่างหนึ่งราย

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ภูมิหลัง: *Neuroendocrine carcinoma ของลำไส้ตรงเป็นโรคที่พบได้น้อยและพยากรณ์โรคไม่ดี เมื่อเปรียบเทียบกับ adenocarcinoma ของลำไส้ตรง ผู้นิพนธ์รายงานผู้ป่วย well-differentiated neuroendocrine carcinoma ของลำไส้ตรงส่วนล่างหนึ่งรายที่ได้รับการรักษาด้วยการผ่าตัด ultra-low anterior resection สำเร็จลุล่วงไปด้วยดี*

รายงานผู้ป่วย: *ชายอายุ 38 ปีมีอาการท้องผูกมา 6 ปี การตรวจทวารหนัก การส่องกล้องตรวจลำไส้ใหญ่และการตรวจเอกซเรย์คลื่นแม่เหล็กไฟฟ้าพบก้อนในผนังด้านหลังของลำไส้ตรงส่วนล่างขนาด 5 เซนติเมตรโดยก้อนอยู่เหนือจากปากทวาร 4 เซนติเมตร การตรวจเอกซเรย์คอมพิวเตอร์ไม่พบการกระจายไปยังตับ ผู้ป่วยได้รับการผ่าตัด ultra-low anterior resection สำเร็จได้ด้วยดีโดยใช้เวลาการผ่าตัด 195 นาที และเสียเลือด 310 มิลลิลิตร รายงานการตรวจพยาธิวิทยาพบว่าเป็นเนื้องอกชนิด well-differentiated neuroendocrine carcinoma และตรวจพบว่ามีการกระจายไปยังต่อมน้ำเหลือง 1 ต่อมจาก 26 ต่อม ผู้ป่วยได้รับการฉายรังสีรักษาและเคมีบำบัดหลังการผ่าตัด ไม่พบการกลับมาเป็นซ้ำเมื่อตรวจติดตามหลังการผ่าตัดไป 14 เดือน*

สรุป: *ถึงแม้ว่าอัตราการกลับเป็นซ้ำหลังการรักษาจะสูงแต่การผ่าตัดแบบ radical oncologic resection ตามด้วยการรักษาเสริมด้วยการฉายรังสีรักษาและการให้เคมีบำบัดยังคงเป็นมาตรฐานในการรักษา neuroendocrine carcinoma ที่ตำแหน่งลำไส้ตรงส่วนล่างและการติดตามผลการรักษาในระยะยาวยังมีความจำเป็น*