Extrapulmonary small cell carcinoma is a rare neoplasm. It is a highly aggressive, malignant tumor with rapid local progression and early metastasis. We report a case of small cell carcinoma arising in the rectum that presented as a bowel habit change in a 57-year-old man. Physical examination revealed a firm, fixed mass in the rectum. Computed tomography of the mass revealed a circumferential bulky mass with an irregular margin with perirectal fat infiltration and enlarged regional lymph nodes. Histologic examination of the resected mass was consistent with small cell carcinoma of the rectum.

**Index words:** Rectal neoplasms
Carcinoma, small cell
Tomography, X-ray computed

Small cell carcinomas are malignancies derived from neuroendocrine cells. Neuroendocrine cells are distributed throughout the gastrointestinal tract, pancreas, lung, thyroid, adrenal gland, and many other organs (1). The gastrointestinal tract has the largest population of neuroendocrine cells, but neuroendocrine malignancies are rare in the colon or rectum (2), accounting for less than 1 percent of all colorectal cancer (3). A PubMed search revealed about twenty case reports for small cell carcinoma of the rectum in the English literature.

Like its pulmonary counterpart, extrapulmonary small cell carcinoma is aggressive, with rapid local progression and early regional and distant spread. Therefore, the prognosis of patients with this neoplasm is generally poor, especially in extrapulmonary small cell carcinoma involving the gastrointestinal tract (4, 5). This report presents computed tomographic (CT) images, a laparoscopic biopsy, and an excisional biopsy of a case of small cell carcinoma of the rectum.

**Case Report**

A 57-year-old male patient was evaluated for complaints of bowel habit changes for the past three months, intermittent hematochezia, and tenesmus. The patient had no history of trauma or any previous surgery. A physical examination revealed the presence of a firm mass in the midrectum. The level of carcinoembryonic antigen (CEA) was normal and the other laboratory findings also were within normal ranges.

Chest radiography showed no pathological findings. An abdominal CT scan performed on a 64-channel multi-slice CT scanner revealed an approximately 7-cm segmental bulky mass with irregular margins encircling the upper rectum, with perirectal fat infiltration and massive perirectal/internal iliac lymph node enlarge-
Colonoscopy revealed a fungating mass with friable mucosa in the rectal wall about 5 cm from the anal verge. Colonoscopic biopsies were consistent with small cell carcinoma of the rectum.

A low anterior resection was performed for complete excision of the mass. Gross examination of the resected specimen showed a huge ulcerofungating mass measuring 11.5 cm × 6 cm. Macroscopically, a cross-section revealed a gray-white solid, firm, neoplastic tissue extending to the perirectal fat tissue. Histologic examination revealed the tumor to be composed of sheets of small, undifferentiated cells. A high-power view showed typical

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Fig. 1. A 57-year-old man with a 3-month history of bowel habit changes.
A. Contrast enhanced CT scan shows an irregular-marginated, bulky mass encircling the upper rectum (white arrows) with perirectal fat infiltration.
B. Contrast enhanced CT scan shows massive regional lymph node enlargement (white arrows).
C. Microphotography shows sheets of highly cellular, small, undifferentiated cells infiltrating the rectal mucosa (H/E, ×10).
D. Microphotography shows tumor cells with scanty cytoplasm, finely granular nuclear chromatin, and absent or inconspicuous nucleoli (H/E, ×100).
E. Immunostaining shows diffuse and intense membrane staining in the small cell carcinoma cells (CD56, ×100).
cytologic features of small cell carcinoma: scanty cytoplasm, finely granular nuclear chromatin, and absent or inconspicuous nucleoli (Fig. 1C, D). Immunohistochemical staining revealed these tumor cells to be diffusely positive for the neuroendocrine marker, CD56 (Fig. 1E).

A chest CT scan was performed 1 month and 6 months after surgery, and a follow-up abdominal CT scan was performed 6 months after the operation. One year has elapsed since the surgery and the patient is doing relatively well, with no evident tumor recurrence or distant metastasis.

Discussion

According to the recent World Health Organization (WHO) classification scheme (2000), neuroendocrine tumors are divided into well-differentiated endocrine tumors, well-differentiated endocrine carcinomas, and poorly differentiated endocrine carcinomas. The most poorly differentiated endocrine carcinomas are small cell carcinomas (6).

Colorectal small cell carcinoma is a relatively rare tumor, with an overall incidence of less than 1 percent among all colorectal cancers (1, 3). The most common location for this type of cancer is the rectum, followed by the cecum and sigmoid colon (1).

Extrapulmonary small cell carcinoma is a distinct disease entity with unique biological behavior and prognosis (4, 5). They frequently show neuroendocrine differentiation by immunohistochemical staining for neuron-specific enolase, chromogranins, and synaptophysin (7).

Neuroendocrine cancers of the gastrointestinal tract manifest a highly aggressive behavior (1, 2), even more than their adenocarcinoma counterpart of the same stage (1, 3). Liver and lymph-node involvement are found early in 70 to 80 percent of patients, and neither size, location, nor number of primary lesions have prognostic value in these tumors (1, 8).

The diagnosis of extrapulmonary small cell carcinoma of the colon should be made only when metastasis from a lung tumor has been excluded (9). Chest CT may be better than chest radiography for excluding primary pulmonary small cell carcinoma and for accurate tumor staging in patients with suspected extrapulmonary small cell carcinoma. Although an initial chest CT scan was not performed here, chest radiographs were normal at presentation and there was not evidence of a pulmonary lesion on follow-up chest CT. This evidence seems sufficient for diagnosing extrapulmonary small cell carcino-

Few reports on small cell carcinoma of the rectum have included radiological findings. Kim et al [9] first reported radiological findings of a poorly enhancing, bulky exophytic mass encircling the colon on CT, but only a pathological specimen was obtained from endoscopic biopsy. Here, small cell carcinoma of the rectum appeared on the CT scan image as a circumferential bulky rectal mass with irregular margins, perirectal fat infiltration, and enlarged regional lymph nodes, which matched with gross specimens.

Although small cell carcinoma of the rectum tends to present with irregular margins, bulky mass, and large lymphadenopathy, these CT findings show considerable overlap with adenocarcinoma or lymphoma. Even endorectal ultrasound findings of small cell carcinoma of the rectum resemble adenocarcinoma (10).

In summary, we present the CT findings of a very rare case of extrapulmonary small cell carcinoma of the rectum. The lesion appeared on the contrast-enhanced CT scan as an irregular-margined, bulky mass with massive perirectal/internal iliac lymph node enlargement. This tumor is a rare type of rectal cancer.

References

직장 소세포암의 CT 소견: 증례 보고

장석기・안인옥・백소야・강수민・유진영・김대봉

폐 외 소세포암은 드문 종양이며 빠른 국소 진행과 초기 전이를 보이는 매우 공격적인 악성 종양이다. 저자들은 배변 습관 변화를 주소로 내원한 57세 남자 환자의 직장에 발생한 소세포암 1예를 경험하였기에 컴퓨터단층촬영 및 조직병리소견을 함께 보고하고자 한다. 이학적 검사에서 직장에 단단하고 고정된 종괴가 만져졌다. 컴퓨터단층촬영에서 이 종괴는 불규칙한 경계의 조영증강을 보이는 환히의 거대 종괴였으며 직장주위 지방 침윤과 커진 국소 림프절이 있었다. 최종 병리 검단은 직장의 소세포암이었다.