A Granular Cell Tumor of the Rectum: A Case Report and Review of the Literature

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A granular cell tumor (GCT) is an uncommon mesenchymal lesion that rarely occurs in the colon and the rectum. We describe the case of a 51-year-old man with a 2-cm-sized rectal GCT 10 cm above the anal verge that was incidentally detected after a screening colonoscopy. Preoperative radiologic studies demonstrated a suspicious submucosal rectal mass with mesorectal fat infiltration, but without circumferential resection margin threatening, extramural vessel invasion, and regional lymph-node enlargement. The tumor was resected by using a transanal endoscopic operation (TEO) without immediate postoperative complications. The final pathology revealed that the tumor consisted of a GCT that had invaded the subserosa with clear margins. It had no other risk factors for malignancy according to Fanburg-Smith criteria. We systematically reviewed the English literature by using PubMed and Google Scholar. This report may be the first documented case in the literature to describe a TEO for a GCT that had invaded the subserosa in the rectum.

Keywords: Granular cell tumor; Colon and rectum; Gastrointestinal tract

INTRODUCTION

A granular cell tumor (GCT) is an uncommon mesenchymal cell tumor, which was first described as a myogenic tumor by Abrikossoff, a Russian pathologist, in 1926 [1]. It commonly occurs in the head and the neck, including the oral cavity, skin and soft tissue, but is rarely found in the gastrointestinal tract (GIT). Although it may be found anywhere in the GIT, the esophagus is the most common site, followed by the duodenum, anus and stomach, but its occurrence in the colon and rectum is very rare [2-6]. We systematically reviewed the English literature by using PubMed and Google Scholar to look for an appropriate treatment for a patient with a GCT that had been found incidentally and revealed invasion of the subserosal rectal wall. This report may be the first documented case in the literature to describe a transanal endoscopic operation (TEO) for treatment of a patient with a GCT in the rectum that had invaded the subserosa.

CASE REPORT

A 51-year-old Korean man with a past medical history of hypertension and hyperlipidemia was referred on Jan 7, 2014, to the Yonsei University Health System for treatment of a known GCT. The colonoscopic finding was an approximately 2.0-cm-sized, hard, rectal mass 10 cm above the anal verge (Fig. 1A). Histopathology findings indicated a GCT composed of round tumor cells having abundant granular cytoplasm with no evidence of malignancy. The patient had no other symptoms when he visited a local hospital on December 13, 2013, for a screening colonoscopy. He had no specific family history, but his sister had had breast cancer. He had a 10 pack/yr history of smoking and a body mass index of 25.7 kg/m².

On admission, the patient appeared well. The laboratory findings were all within normal limits. Although the computed tomography (CT) findings made us suspicious of T3 rectal cancer (Fig. 1B), subsequent magnetic resonance imaging (demonstrated a probable submucosal rectal mass with mesorectal fat infiltration above 8 cm from anal verge (Rb), but without circumferential re-
section margin threatening, extramural vessel invasion, or regional lymph-node enlargement (Fig. 1C). His chest x-ray showed neither active lung disease nor evidence of lung metastasis.

When we reviewed the English literature, we were able to find only a few cases involving a GCT that had extended to the level of the muscularis propria. Our colorectal team, including colorectal surgeons, oncologists, gastroenterologists, and pathologists, discussed an appropriate treatment for this rare case. Even though we first considered a low anterior resection and lymph-node dissection, we finally decided to perform a TEO, with which we had had plentiful experience for patients with early rectal cancer. The surgery was performed on January 14, 2014. The patient had an uneventful postoperative course and was discharged 6 days after surgery. Two weeks later, the patient visited our clinic, and the final pathology revealed that the tumor consisted of a 1.7 × 1.5-cm
GCT (Fig. 2), which had been positively stained for the S-100 protein (Fig. 3D) and had invaded the subserosa with clear margins (Fig. 3A–C). The patient exhibited no other risk factors for malignancy according to Fanburg-Smith criteria [7]. Because of the rarity of this disease entity and the lack of experience with a GCT with subserosal invasion, short-term follow-up was recommended. The CT scan and sigmoidoscopy performed nine months later showed no evidence of recurrence.

**DISCUSSION**

A colorectal GCT is a very rare submucosal tumor that usually follows a benign course. It is incidentally described during a screening endoscopy. In most cases, a polypectomy or an endoscopic mucosal resection (EMR) is usually sufficient to deal with this rare disease entity [8-10]. Although a malignant GCT is extremely uncommon, such cases have been reported in the literature as representing 1%-2% of all GCTs [7, 11, 12]. However, its recurrent pattern and prognosis are not fully understood. In 1998, Fanburg-Smith et al. [7] suggested 6 diagnostic criteria for malignant GCTs. Six histologic criteria were assessed: necrosis, spindling, vesicular nuclei with large nucleoli, increased mitotic activity (>2 mitoses/10 high-power fields at ×200 magnification), high nuclear-to-cytoplasmic ratio, and pleomorphism. Those authors defined a GCT as a histological malignancy when three or more of these criteria were met. They also reported prognostic factors for patients with a GCT: local recurrence, metastasis, larger tumor size, older patient age, histologic classification as malignant, presence of necrosis, increased mitotic activity, spindling of tumor cells, vesicular nuclei with large nucleoli, and Ki-67 values less

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**Fig. 3.** A submucosal tumor revealed by histological examination in resected tissue. (A) Resected lesion showing a submucosal tumor covered with normal mucosa and with subserosa invasion (H&E, ×40). (B) Higher magnification view of the tumor, which revealed nonuniform large tumor cells with slightly pleomorphic nuclei (H&E, ×200). (C) Higher magnification view of a tumor composed of large tumor cells with abundant granular cytoplasm and small round nuclei (H&E, ×200). (D) Diffuse, strong expression of S-100 protein in the tumor shown by immunohistochemistry (×200).
A colorectal GCT usually occurs within the submucosa with an intact mucosa surface, and extensions below the level of the muscularis propria are seldom reported in patients with a colorectal GCT. Cha et al. [8] reported in 2009 that GCTs were usually less than 2 cm in size and did not invade the muscularis propria. Although no guidelines for colorectal GCTs existed at that present, those authors, according to the results of their retrospective case analysis, insisted that a benign lesion less than 2 cm in diameter and separated from the muscularis propria could be removed by using a EMR. On the other hand, Znati et al. [13] suggested that patients with a colonic GCT should be managed conservatively, with a polypectomy or an EMR for tumors less than 4 cm in size and with a segmental resection for tumors with larger sizes.

Recurrence is very rare after a curative resection of a GCT, but it does occur, although seldom. In 2010, Singhi and Montgomery [14] reported the occurrence of regrowth at the prior biopsy site owing to incomplete excision. However, the recurrence pattern of GCTs is still not known, so appropriate surgical margins have yet to be clarified for patients with a colorectal GCT.

We treated our patient by using a TEO with a clear resection margin, and the final pathology revealed that the tumor had invaded the subserosa. However, because the size of the GCT was less than 2 cm and the GCT was not suitable for malignancy according to Fanburg-Smith criteria [7], in our opinion, our procedure should be sufficient in this case, even though the patient will require close follow-up.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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