Journal of Enterocolitis

Case Report

Two Cases of Malignant Transformation in Crohn's Disease with Perianal Involvement

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Cite this article as: Dağcı G, Varelci S, Büyük M, Çavuş B, Çiftçibaşı Örmeci A, Akyüz F. Two cases of malignant transformation in crohn's disease with perianal Involvement. *J Enterocolitis.* 2023;2(3):66-68.

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Received: November 27, 2023 **Accepted:** December 4, 2023

DOI:10.14744/Jenterocolitis.2023.231457

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Abstract

This report includes 2 cases of Crohn's disease with perianal involvement in which adenocarcinoma developed in the setting of a fistula. The cases illustrate the importance of considering the development of neoplasia in the presence of fistula, treatment-resistant symptoms, and rectal discharge.

Keywords: Crohn's disease, malignancy, perianal disease

INTRODUCTION

The frequency of intestinal and extraintestinal malignancies in patients with Crohn's disease is higher than that in the normal population.[1] In perianal involvement, there are difficulties in the detection of malignancies due to the severity of inflammation, similarity of symptoms to those seen in infection, and limited sensitivity of imaging methods. Special attention regarding malignant transformation should be paid for patients with Crohn's disease who have long-standing anal fistulas, especially when they complain of exacerbated symptoms (i.e., increased pain, fluid discharge) from a perianal lesion.[2]

In this report, we present 2 cases of Crohn's disease with perianal involvement who subsequently developed mucinous rectal adenocarcinoma.

CASE 1

A 38-year-old male patient was admitted to our clinic with rectal pain and discharge. Eighteen years ago, he was diagnosed with fistulizing and stenosing Crohn's disease, and a right hemicolectomy was performed due to ileus at the time of diagnosis.

The patient was followed up with mesalamine and azathioprine treatments and was lost from follow-up for the next 7 years as he had no complaints. One year ago, a perianal abscess was detected in the patient due to rectal pain and discharge; drainage and seton procedures were performed; treatment was planned as 40 mg of adalimumab every other week and azathioprine 150 mg per day.

Pelvic magnetic resonance imaging (MRI) performed at his first presentation due to rectal pain and discharge revealed an appearance compatible with an abscess in the perirectal area with a size of 46 × 17 × 50 mm and intersphincteric fistula tracts located at the 4, 6, and 9 o'clock levels. Colonoscopy showed an anastomosis line and distal ileum, deep ulcers in the rectum, and perianal high-flow fistulas (Figure 1). Neoterminal ileum biopsy was consistent with mild active chronic enteritis with crypt distortion, and rectal biopsy was consistent with chronic inflammatory mucosa.

The patient underwent perianal abscess drainage and a seton procedure. After 1 month of ciprofloxacin and metronidazole antibiotherapy, the current treatment was revised as infliximab 400 mg for 8 weeks and AZA 150 mg/day. On the pathological examination of the perianal abscess drainage material, which was repeated due to the persistence of rectal pain and discharge complaints, abscesses in the fatty tissue as well as dense mucin pools and single-row mucin pools without obvious atypia within these mucin pools and epithelial cell groups arranged basally were observed, and mucinous adenocarcinoma was suspected.

Positron emission tomography–computed tomography showed focal nonhomogeneity (maximum standardized uptake value (SUV_{max}): 10.0) in the area extending from the rectum to the anal canal and skin level and pathological fluoro 2-deoxyglucose (FDG) uptake (SUV_{max}: 7.8) in several foci in the right lung. It was reported that Miles operation was performed after neoadjuvant FOLFOX and radiotherapy.

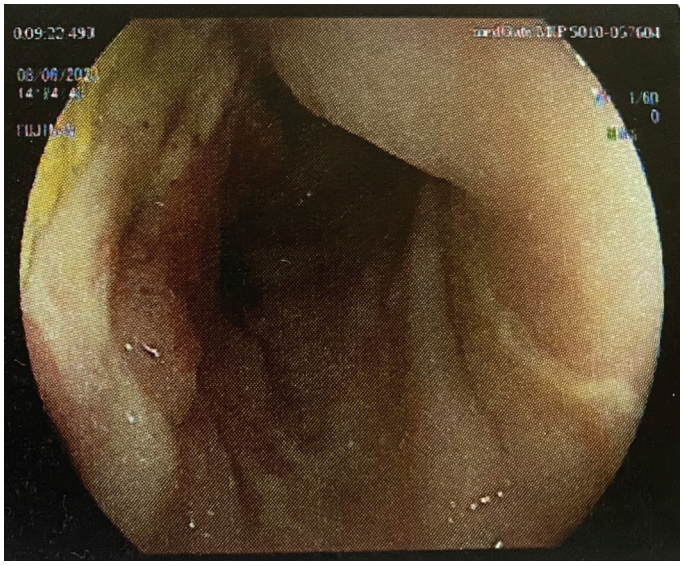

Figure 1. Deep ulcers in the rectum of case 1.

CASE 2

A 48-year-old male patient presented to our clinic with pain in the rectum and a palpable mass. It was learned that he had a history of anxiety disorder and a diagnosis of fistulizing and stenosing Chron's disease for 21 years, 3 times perianal abscess drainage, and a seton procedure in the center to which he was referred. Due to ileal stenosis, a right hemicolectomy was performed 11 years ago. The patient underwent treatments with AZA+meselamin for 11 years, infliximab for 8 years, and adalimumab for 2 years, respectively.

A year ago, a sigmoid loop colostomy was performed due to a perianal abscess and fistula refractory to medical treatment. Pelvic MRI revealed an 8 × 7 cm mass lesion originating 2.5 cm superior to the anal canal and extending to the prostate and subcutaneous adipose tissue in a 5-cm segment towards the superior part of the rectum. endoscopic ultrasound revealed transsphincteric fistula and a 4-cm heterogeneous

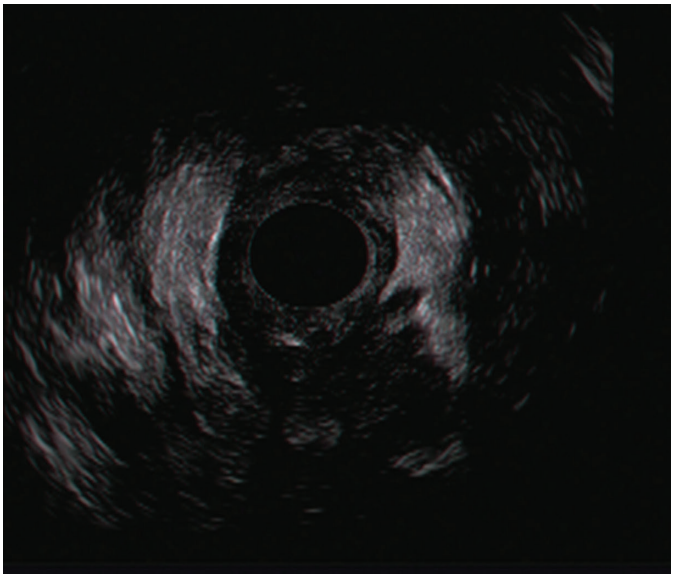

Figure 2. Transsphincteric fistula and a 4-cm heterogeneous mass in case 2.

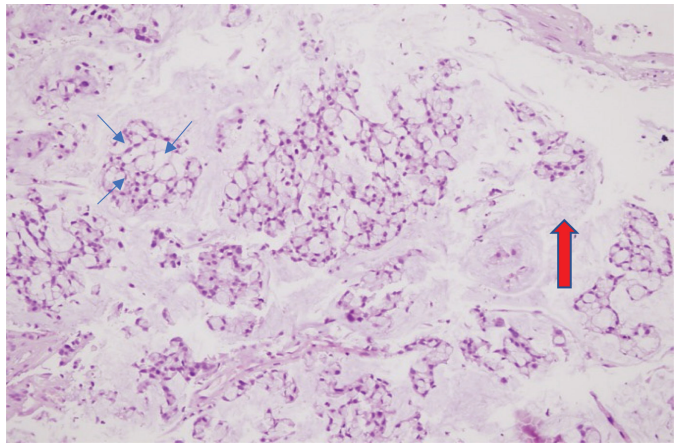

Figure 3. Tumor cells (blue arrows) in groups in extracellular mucin (red arrow) of case 2: mucinous adenocarcinoma.

mass at the 5 o'clock position (Figure 2). The sample taken from the tumor revealed mucinous adenocarcinoma upon pathologic investigation (Figure 3). Positron emission tomography–computed tomography showed a mass lesion (SUDmax: 8.6) with heterogeneous increased FDG uptake originating from the proximal rectum and extending to the perianal region, extending to 9.8 × 6.8 cm in the widest part of the lesion. The patient was staged with locally advanced rectal cancer, and abdominoperineal resection was performed after neoadjuvant 28 sessions of radiotherapy+capecitabine and bevacizumab treatment. Liver metastasis was detected in postoperative 6-month control imaging.

DISCUSSION

Patients with Chron's disease have a 2-fold to 3-fold increased risk for colorectal cancer compared to the general population.[3] The pathogenetic pathway leading from inflammation to dysplasia and cancer in Inflammatory bowel disease differs from that of sporadic cancers in terms of the sequence and rate of genetic alterations triggered by specific cytokines and transcriptional factors upregulated in the inflammatory microenvironment.[4] The presence of perianal inflammation is significantly associated with a high risk of both colorectal and anal cancers.[5,6] Patients who develop perianal fistula-related or rectal adenocarcinoma are typically young patients with long-standing anorectal inflammation.[7] The mucinous type accounts for 15%-20% of rectal cancers, is detected at more advanced stages than other subtypes, and has a lower response to chemotherapy.[8]

In addition to having had several surgical procedures, our 2 patients also had fistulizing disease, treatment-refractory perianal involvement, advanced disease age, and were on anti-Tumor Necrosis Factor-alfa Inhibitor therapy. To rule out malignancy in Crohn's disease cases with perianal involvement, prolonged disease age, recurrent abscess drainage, and fistula history, multiple lesion biopsies and pathologic investigations should be performed.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – G.D., F.A.; Design – G.D., S.V., F.A., A.Ç.Ö.; Supervision – B.Ç., S.K., F.A., A.Ç.Ö. Materials – G.D., B.Ç., F.A., M.B.; Data Collection and/or Processing – G.D., S.V.; Analysis and/or Interpretation – G.D., S.V., F.A.; Literature Search – G.D.; Writing Manuscript – G.D., S.K., F.A., A.Ç.Ö.; Critical Review – B.Ç., S.K., F.A., A.Ç.Ö.

Declaration of Interests: The authors declare that they have no competing interest.

Funding: The authors declare that this study has received no financial support.

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