Turk J Gastroenterol *2015; 26: 435-7*

Squamous cell carcinoma of the cecum developing in a patient with long-standing ulcerative colitis and a coexistent carcinoid tumor in the appendix: A case report

Saqib Ahmad, Nicholas Watson, Kiran Shamim, Achyuth Menon, Khorrum Abdullah
Department of Gastroenterology, Kings Mill Hospital, Mansfield, United Kingdom

ABSTRACT

Squamous cell carcinoma (SCC) of the colon is a rare tumor that accounts for 0.1%–0.2% of colonic malignancies. However, pure SCC of the cecum is extremely rare. Although SCC of the colon is a rare complication of idiopathic inflammatory bowel disease (IBD), SCC of the cecum in the context of IBD has not been previously reported in the English literature. We report a case of the coexistence of SCC of the cecum and a carcinoid tumor in a 46-year-old female with long-standing pan-ulcerative colitis.

Keywords: Squamous cell cancer, cecum, IBD

Case Report

INTRODUCTION

Squamous cell carcinoma (SCC) of the colon is a rare tumor that accounts for 0.1%–0.2% of colonic malignancies (1). However, pure SCC of the cecum is extremely rare. Although SCC of the colon is a rare complication of idiopathic inflammatory bowel disease (IBD), SCC of the cecum in the context of IBD has not been previously reported in the English literature. We report a case of the coexistence of SCC of the cecum and a carcinoid tumor in a 46-year-old female with long-standing pan-ulcerative colitis (UC).

CASE PRESENTATION

A 46-year-old Caucasian female with a 30-year history of pan-UC in long-term remission presented to our emergency department with a 6-week history of discomfort in the right iliac fossa, weight loss, malaise, and exertional dyspnea. A colonoscopy performed 5 years previously showed no active disease. A second-degree relative of the patient had died from colorectal cancer. The patient had no extraintestinal manifestations of UC, no significant gynecological history or symptoms, and no history of smoking as well as no history of skin cancer or other malignancies.

Clinical examination revealed anemia and a palpable mass in the right iliac fossa. Initial investigations showed the following: hemoglobin, 5.0 g/dL; platelet count, 601×10^9/L; albumin, 28 g/L; and ferritin, 5 ng/mL, despite oral iron supplementation. An attempt to perform an urgent colonoscopy was abandoned because of the patient's discomfort. Subsequent contrast-enhanced computed tomography of the chest, abdomen, and pelvis revealed a 7-cm tumor involving the cecal pole with adjacent lymph nodes having a diameter of 10 mm (radiological staging, T3/T4). Thickening of the rectal wall and sigmoid colon consistent with colitis was also noted.

Proctocolectomy with mesorectal excision and intersphincteric dissection were performed on obtaining informed consent from the patient. A large mobile cecal tumor with multiple enlarged ileocolic lymph nodes was found. Macroscopically complete resection was achieved. There were no postoperative complications, and the patient remained in good condition at the 12-month follow-up.

Histological examination of the specimen (Figure 1) showed completely excised, moderately differentiated

Address for Correspondence: Saqib Ahmad, Department of Gastroenterology, Kings Mill Hospital, Mansfield, United Kingdom
E-mail: ahmad_saqib@hotmail.com

Received: July 17, 2013 **Accepted:** November 28, 2013 **Available Online Date:** July 24, 2015

SCC of the cecum invading beyond the muscularis propria, with positive immunostaining for CK5/6 and P63 and negative immunostaining for CK20 and CDX-2. All 35 examined lymph nodes were tumor-free, with no vascular or perineural invasion. Examination of the specimen and adjacent mucosa revealed negative P16 immunostaining for the presence of human papillomavirus (HPV). However, there was histological evidence of surface glandular dysplasia with markedly villiform architecture, and this merged into areas that seemed to have surface squamous metaplasia/dysplasia. There was a background of quiescent UC, and gross thickening of the muscularis mucosa and minor distortion of the crypt architecture were noted on conducting histopathological analysis.

In 2006, the patient underwent biopsies of the ascending, transverse, and sigmoid colon, which were reported as mildly active UC indefinite for dysplasia. There was also a small neuroendocrine tumor (carcinoid) in the appendix.

DISCUSSION

Long-standing UC confers an increased risk of adenomatous dysplasia and colorectal adenocarcinoma, influenced by the extent, severity, and duration of colitis (1). In contrast, pure SCC of the colon is rare (0.1–0.25 per 1000 neoplasms), and tend to occur in the lower rectum and anal canal because of inflammation-induced metaplasia at the squamo–columnar junction (1-3). The first report of pure SCC of the colon was by Schmidtman in 1919 (4). Since then, less than 50 cases of SCC in the colon or rectum have been reported. In 1979, Williams et al. (5) proposed the following three criteria that should be met before establishing a diagnosis of SCC: (a) metastasis from other sites to the colon must be excluded; (b) a squamous-lined fistula tract must not involve the affected bowel because it may be a source of SCC; and (c) SCC of the anus with proximal extension must be excluded.

Although epidemiological evidence suggests that the incidence of anal SCC has not increased in long-standing IBD (4), UC has been most frequently reported with colonic SCC, and the symptoms are comparable with adenocarcinoma (4). Both squamous metaplasia and SCC have been reported as possible complications of IBD, but the number of cases is very limited. Kulaylat et al. (6) reviewed 16 reported cases of SCC complicating IBD, which is the largest series in the literature to date. Only one case occurred in the setting of chronic Crohn's colitis, and the rest were associated with UC. Of the 16 cases, 20% were adeno-SCC and 80% were purely SCC. None of them involved the cecum (6).

The pathogenesis of these tumors is poorly understood but may involve squamous de-differentiation of a pre-existing adenocarcinoma or squamous metaplasia progressing to dysplasia and carcinoma as a result of a chronic inflammatory stimulus (1-5). In the present case, the presence of squamous metaplasia in the adjacent mucosa suggests the latter. As with SCC of the anus and other sites, HPV infection has also been implicated; however, this did not appear to be a factor in our case (7,8).

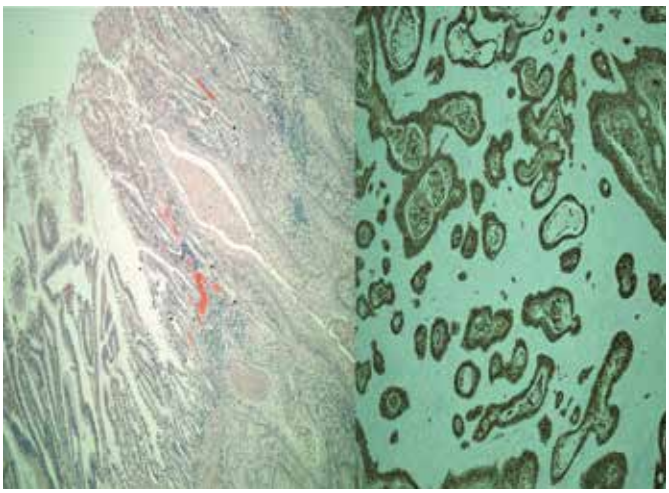

Figure 1. Surface glandular dysplasia merging into areas appearing to have surface squamous dysplasia with infiltrating squamous cell carcinoma (Left; H/E stain, 4×). CK 5/6 positive immunohistochemistry test result (Right).

Compared with sporadic colorectal carcinoma, colorectal carcinoma developing in patients with IBD has several distinguishing clinical features. Colitis-associated colorectal cancer affects individuals at a younger age compared with the general population, and it more often has mucinous or signet ring cell histology; there is a higher rate of two or more synchronous primary colorectal cancers. Some studies have demonstrated a more proximal distribution in the colon (9).

Patients with IBD are at a risk of adenocarcinoma and SCC development; although the risk factors and clinicopathological features are identical in both carcinomas, SCC develops less frequently than adenocarcinoma (10). This is the first reported case of SCC in the cecum of a patient with long-standing ulcerative colitis. Surgical resection remains the mainstay of treatment.

Ethics Committee Approval: NA.

Informed Consent: NA.

Peer-review: Externally peer-reviewed.

Author contributions: Concept - K.A.; Design - N.W.; Supervision - K.A., A.M.; Resource - N.A.; Materials - N.A.; Data Collection &/or Processing - N.A.; Analysis &/or Interpretation - N.A.; Literature Search - S.A., K.S.; Writing - S.A., N.W.; Critical Reviews - K.A.

Acknowledgements: We are grateful to Professor Neil Anthony Shepherd of Cheltenham General Hospital for independently reviewing the original histopathology specimens and report.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Pikarsky AJ, Belin B, Efron J, et al. Squamous cell carcinoma of the rectum in ulcerative colitis: case report and review of the literature. Int J Colorectal Dis 2007; 22: 445-7. **[CrossRef]**
2. Juturi JV, Francis B, Koontz PW, Wilkes JD. Squamous-cell carcinoma of the colon responsive to combination chemotherapy. Dis Colon Rectum 1999; 42: 102-9. **[CrossRef]**

3. Cheng H, Sitrin MD, Satchidanand SK, Novak JM. Colonic squamous cell carcinoma in ulcerative colitis: Report of a case and review of the literature. Can J Gastroenterol 2007; 21: 47-50.
4. Schmidtman M. Zcor Kenntis Settener Krebs-formen Virchows. Arch Path Anat 1919; 226: 100-18.
5. Williams GT, Blackshaw AJ, Morson BC. Squamous carcinoma of the colorectum and its genesis. J Pathol 1979; 129: 139-47. **[CrossRef]**
6. Kulaylat MN, Doerr R, Butler B, Satchidanand SK, Singh A. Squamous cell carcinoma complicating idiopathic inflammatory bowel disease. J Surg Oncol 1995; 59: 48-55. **[CrossRef]**
7. Steven H, Itzkowitz, Xianyang Yio. Inflammation and Cancer IV. Colorectal cancer in inflammatory bowel disease: the role of inflammation. American Journal of Physiology 2004; 287: G7-G17.
8. Frisch M, Johansen C. Anal carcinoma in inflammatory bowel disease. Br J Cancer 2000; 83: 89-90.
9. Comer TP, Beahrs O, Dockerty MD. Primary Squamous Cell Carcinoma and adenocarcinoma of colon. Cancer 1971; 28: 1111-7. **[CrossRef]**
10. Balsano NA. Squamous cell carcinoma of the caecum. Arch Surg 1985; 120: 1176-7. **[CrossRef]**