Case Report

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Rectal Squamous Cell Carcinoma: A Case Report of a Rare Histology

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Abstract

The rectum is an unusual and unlikely site for the occurrence of squamous carcinoma of the gastrointestinal tract. The clinical presentation of this rare histology though, is similar to rectal adenocarcinoma. Being a rare clinical entity with limited cases, the etiology, pathogenesis and optimal treatment regimen is still unclear. In this paper, we present a case of a 55-year-old man who presented with a complaint of rectal bleeding for the previous six months. Imaging revealed mass in rectum; the biopsy was suggestive of squamous cell carcinoma. He was treated with abdominoperineal resection followed by adjuvant chemoradiotherapy.

Keywords: Squamous cell carcinoma; Hematochezia; Rectum

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Introduction

More than 90% of tumors arising in the colorectal region are adenocarcinomas. Other histologic types include melanomas, small cell carcinomas, carcinoids, sarcomas, lymphoma and squamous cell carcinomas (SCC) (1). SCC is extremely rare, contributing to between just 0.1 to 0.25% of all colorectal malignancies (2, 3). Due to its rare nature and the resultant paucity of published literature, the etiology, pathogenesis and possible risk factors of this condition remain obscure. Hence, there is significant variability in the therapeutic regimens used, and the most appropriate management protocol remains unclear. In 1919, Schmidtmannin became the first to describe a case of colorectal SCC. The patient in question was a 66-year-old man with a carcinoma of the cecum (4). William et al. described three criteria that should be met to diagnose rectal SCC: (a) metastases from SCC of any other organ to the rectum must be ruled out; (b) there should be absence of anal involvement by SCC; and (c) there should be no SCC-lined fistula reaching up to the rectum (5).

In this case report, we present a case of squamous cell carcinoma of the rectum and review the literature for the available management modalities; we also discuss the suitable treatment options for this uncommon disease.

Case

A 55-year-old male with no known comorbidities

and no history of tobacco or alcohol use presented to our hospital with a history of hematochezia associated with diffuse abdomino-pelvic pain for the previous six months. A colonoscopy was performed, which revealed a circumferential lesion located in the rectum starting at 5 cm proximal to the anal verge, about 2 cm above the anorectal ring (Figure 1). An abdominal computed tomography (CT) scan revealed an exophytic mass lesion arising from the mucosa of the lower third of the rectum. The lesion displayed heterogeneous contrast enhancement associated with perirectal and bilateral iliac lymphadenopathy. Contrast enhanced magnetic resonance imaging (MRI) of the pelvis corroborated the findings of CT scan. Whole body positron emission tomography (PET) CT was negative for distant metastases but revealed the rectal mass along with the peri-rectal and pelvic lymph nodes to be hypermetabolic.

The colonoscopic biopsy revealed a moderately differentiated squamous cell carcinoma (MDSCC) with eosinophilic, polygonal squamous cells without glandular formation. On immunohistochemistry (IHC), the cells were found to be positive for the P40 and P63 markers (Figure 2). In view of the rectal origin of the lesion, the lack of extension into the anal canal, the lack of evidence of any malignancy elsewhere in the body or any fistulous tract arising from the rectum, and the characteristic histopathological picture of a squamous carcinoma, the patient was diagnosed with rectal SCC.

The patient underwent an abdominoperineal resection (APR) of the tumor with a diversion colostomy. The histopathology of the surgical specimen also revealed a MDSCC. The tumor had arisen from the mucosa within and had reached the serosa with no perineural invasion (PNI) or lymphovascular invasion (LVI). Nine lymph nodes were dissected and all were negative for tumor cells. However, the tumor was close to circumferential resection margin. The tumor was staged pathologically as pT3N0 as per the seventh edition of the AJCC staging system. Due to the close margin, the case was discussed in a multidisciplinary tumor board, where it was decided to commence adjuvant radiotherapy to the pelvis along with concurrent capacetabine-based chemotherapy. The patient was treated with adjuvant radiotherapy to the pelvis on a Theratron 780E telecobalt machine at a dose of 5040 Grays in 28 fractions along with concurrent capecitabine and mitomycin. The planned treatment

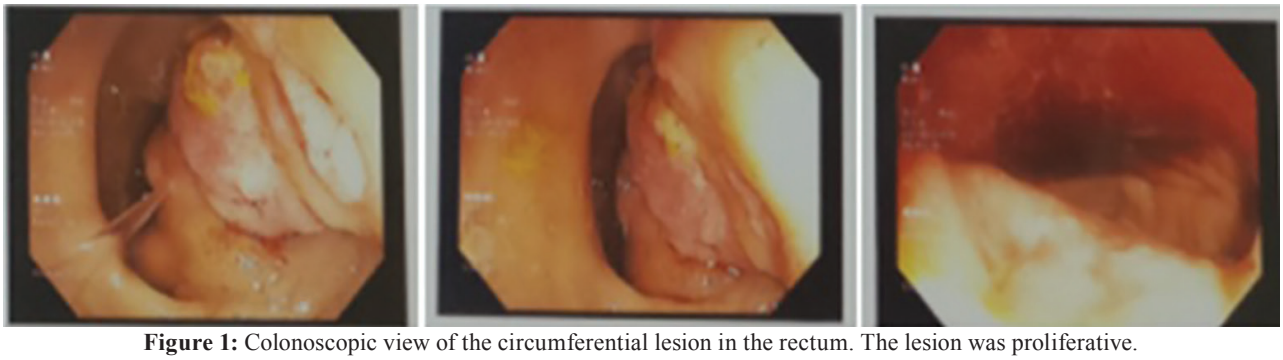

Figure 1: Colonoscopic view of the circumferential lesion in the rectum. The lesion was proliferative.

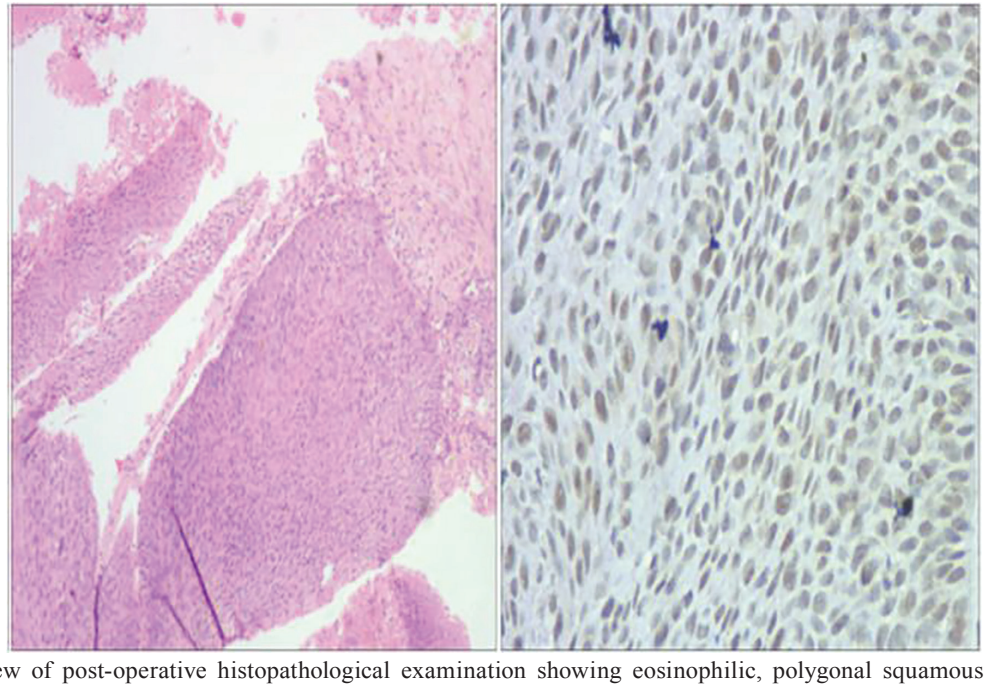

Figure 2: View of post-operative histopathological examination showing eosinophilic, polygonal squamous cells with no glandular formation as seen in the image on the left side. The image on the right shows the positive staining of cells by P40 on immunohistochemistry.

was completed without any major treatment-related toxicities. The patient has been on regular follow up since then and remains disease-free at six months after treatment completion. Informed consent was obtained from the patient for the publication of this report.

Discussion

Squamous carcinoma in the rectum is extremely uncommon, making up only 0.3% of all subtypes. Certain other histologies like gastrointesinal stromal tumors, lymphomas and neuroendocrine tumors are also infrequent, but rectal SCC is by far the rarest if sarcomas are excluded (6). Due to its rarity, the risk factors and pathogenesis of SCC are not well defined. There is an obvious paucity of literature in this regard, with the only available studies to guide us being retrospective reviews, case series and case reports.

Although the etiology and risk factors for rectal SCC have not been clearly established, one of the leading theories is that chronic inflammation can lead to squamous metaplasia and subsequent carcinoma. The disease has been known to occur in patients with pre-existing inflammatory processes such as ulcerative colitis, *Entamoeba histolytica* infestation, chronic Human Papilloma Virus (HPV) infection, and schistosomiasis (7, 8). The inflammation is believed to give rise to squamous metaplasia, which further develops into carcinoma. Another hypothesis is that pluripotent mucosal stem cells that are capable of multidirectional differentiation may give rise to malignant squamous cells in the rectum (9). A third theory suggests that these carcinomas arise from areas of squamous differentiation that are sometimes found within rectal adenomas and adenocarcinomas (10). Lastly, Michelassi et al. have proposed that epithelial damage stimulates rectal basal cells to rapidly proliferate into squamous cells, with malignant transformation rarely occurring (11). As per the report of Na-has et al., (12) the keratin profiles of both rectal SCC and adenocarcinoma are similar, indicating that both arise from a common cell line. At the same time, these keratin profiles are unlike that of squamous carcinoma of the anal canal, suggesting that SCC of the rectum and SCC of the anal canal are two distinct entities rather than an anatomical superimposition of a single disease over a contiguous neighboring organ.

Both rectal SCC and adenocarcinoma clinically present most commonly with bleeding per rectum or hematochezia. Less commonly, the patient may complain of altered bowel habits (constipation, diarrhea, tenesmus), abdominal or pelvic pain, and weight loss (13). Many presume that rectal SCCs maybe simply a continuation of anal, cervical or vaginal carcinomas. To rule out this possibility, a thorough history and clinical examination with careful attention to the anal canal and also the gynecological system in females should be undertaken. The AJCC staging system used for rectal adenocarcinoma was applied. Staging includes assessment of the primary tumor as well as of any regional or metastatic disease. Both pelvic MRI and endorectal ultrasound (ERUS) can be used for evaluation of the primary tumor, with the choice of preferred modality being dependent on individual expertise (14).

Historically, the treatment of rectal SCC has been surgical resection in the form of anterior resection (AR) or abdominoperineal resection (APR) similar to that of adenocarcinoma. Nigro's chemoradiotherapy (CRT) protocol has become the standard of care for anal carcinoma as it preserves sphincteric function while achieving local control, with the overall survival being similar to that of surgical treatment. In light of this development, this protocol has been slowly adopted for the treatment of rectal SCC. The CRT regimen used for rectal SCC is similar to the one used for the treatment of anal SCC; several studies show high rates of local control (15, 16). Due to limited published literature and absence of a standardized clinical staging system, it is not appropriate to compare different treatment modalities such as surgery and CRT. Nevertheless, chemoradiation therapy (CRT) remains an alternative to surgical resection.

In conclusion, SCC of rectum is a rare entity. As the literature on staging and treatment of squamous carcinoma of rectum lacks standardization, it is important to shed some light on this rare condition because it has a different epidemiology, pathogenesis and treatment than other colorectal carcinomas. CRT and surgical interventions have shown equal outcomes, but surgery has been the traditional form of management for patients with SCC. At the same time one cannot ignore the advantages of using CRT as the primary treatment method as it allows sphincter preservation leading to a better quality of life and, possibly, an improvement in survival. Clearly, further research is required to understand the etiology and underlying mechanism of this rare disease. Similarly, more emphasis is needed on comparing and deciding on the most preferred treatment modality for rectal SCC by carrying out larger prospective and comparative studies.

Conflict of Interests: None declared.

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