Primary Squamous Cell Carcinoma of the Rectum
Case Report with Literature Review

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ABSTRACT

Background: Squamous cell carcinoma of the rectum is a distinct entity. We report a rare case of squamous cell carcinoma of the middle and low rectum. Case presentation: The patient was a 60-year-old man who presented with a history of rectal bleeding, constipation and pelvic painless. Rectoscopy and Colonoscopy revealed a polypoid tumour of the middle and low rectum. Biopsies of this mass revealed a poorly differentiated squamous cell carcinoma of the rectum. Pelvic Magnetic Resonance Imaging scan showed a polypoid, irregular and circumferential tumor in the middle and low rectum extending in length on 95mm. CT scan of the chest, abdomen and pelvis was negative for distal metastases. The patient received combined chemoradiation. Overall treatment time was 45 days. After 6 weeks of the end of treatment, the revaluation clinical tests by pelvic magnetic resonance imaging showed a poor response. After 3 months, Positron Emission Tomography with Fluorodeoxyglucose demonstrated a little hypermetabolic area with maximum standard uptake value of 6,7 in the low rectum. Patient underwent a surgical resection. The postoperative histopathological findings were non specific and chronic inflammatory changes. At the time this report was written, the patient had 18 months of follow-up. No evidence of malignancy is found. Conclusion: Squamous cell carcinoma of the rectum is a distinct entity. Chemoradiation should be chosen as the first-line therapy for squamous cell carcinoma of the rectum. However, a large study will be required to establish a safe and effective regimen.

Keywords: Squamous Cell Carcinoma, Rectal cancer, Chemoradiation, Surgery

1. INTRODUCTION

Primary Squamous Cell Carcinoma (SCC) is an unusual malignancy in rectum. Due to the low incidence of this cancer and subsequent lack of literature, the underlying pathogenesis and risk factors are not exactly known(1). The available compendium of knowledge is based on isolated case reports and case series. Historically, rectal SCC was treated like rectal adenocarcinoma, and the main treatment was surgical resection(2). Based on the experience achieved in anal SCC patients, chemoradiotherapy (CRT) has been the treatment of choice(3). We report a case of SCC of rectum which was treated successfully with CRT.
2. CASE PRESENTATION

A 60-year-old male presented to us with painless bleeding per rectum of 8 months duration. He also reported heaviness in pelvis, constipation for last 2 months and loss of appetite. No history of any co-morbid illness or substance abuse was present. Past and family history was unremarkable. General physical examination was normal except for a mild degree of pallor. Systemic examination including digital rectal examination was revealed a friable mass bleeding to the fingertip friction at 5 cm from the anal verge. A full rectoscopy and colonoscopy was planned after bowel preparation. It revealed a proliferative nodular growth starting at 5 cm from anal verge on lateral and posterior walls of middle and low rectum. Biopsies were performed and histo-pathological examination showed malignant squamous cells with keratin pearl formation suggestive of poorly differentiated SCC (figure 1A and 1B).

Immunohistochemistry samples showed Cytokeratin (CK)7 and CK20 negativity, CK5/6 positivity with overexpression of p40 (figure 2A) and p63 (figure 2B).

Figure 1A: Haematoxylin and eosin stain G x 400: widespread squamous cells carcinoma of the rectum.

Figure 1B: Haematoxylin and eosin stain G x 200: poorly differentiated carcinoma. Invasive squamous cells carcinoma of rectal mucosa.

Figure 2A: Immunohistochemistry G x 200: overexpression of p40.

Figure 2B: Immunohistochemistry G x 400: overexpression of p63.

HPV 16 research by PCR technique was not performed. A Pelvic Magnetic Resonance Imaging (MRI) scan showed a polypoid, irregular and circumferential tumor in the middle and low rectum extending in length on 95mm. It involve the mesorectum with distance to facia recti up to 5 cm. It respect the anal sphincter and no evidence of pelvic nodes metastasis were found (figure 3).
Patient underwent a surgical resection. The postoperative histopathological findings were no specific and chronic inflammatory changes. At the time this report was written, the patient had 18 months of follow-up. No evidence of malignancy is found.

3. DISCUSSION

Most of the colorectal malignancies are adenocarcinomas while rectal SCC is a rare entity. The incidence of SCC ranges from 0.1 to 0.25 per 1000 colorectal neoplasms\(^{(1)}\). Raiford reported the first case of rectal SCC in 1933\(^{(4)}\). Given the rarity of the disease, strong epidemiological data regarding patient demographics, risk factors, natural history and optimal treatment is lacking\(^{(5)}\). SCC of the rectum appears to affect individuals between the ages of 30 to 93 years old with mean age of 57 years\(^{(6)}\). A review of available reports show that 66% of cases occurred in women and 34% in men\(^{(7)}\). The rarity of this disease precludes any possibility to studying and establishing the exact etiology and pathogenesis. However, several associations have been observed. Some cases reports have been found SCC in association with inflammatory bowel diseases involving colon and rectum, ulcerative colitis\(^{(8)}\), infections including Schistosomiasis and Entamoeba Histolitica\(^{(8)}\), previous radiation exposure to the pelvis\(^{(9)}\) and immunosuppressive state\(^{(10)}\). Although a clear association between SCC of anal canal and Human Papilloma Virus (HPV) exists, similar relation with rectal SCC has not been established.

Patients with SCC of the rectum present with symptoms similar to those with adenocarcinoma of the rectum. The symptoms most frequently encountered are rectal bleeding, abdominal pain, change in bowel habits and weight loss. Patients usually experience symptoms for several weeks to months\(^{(1,2,3,4)}\).
williams et al. suggested criteria of the diagnosis of colorectal SCC in 1979. They includes absence of direct extension of tumor from canal epithelium, absence of primary SCC in the body, absence of fistulous tract to the affected bowel and histological confirmation disease\textsuperscript{(11)}. Proctoscopy can be used to retrieve a biopsy of any visible abnormalities (ulcered mass, polyps...) for definitive histological analysis. Immunohistochemistry helps to differentiate rectal from anal lesions. The most useful CK are Cell Adhesion Molecules (CAM) 5.2, 34B12 and AE1/AE3. CAM 5.2 stains rectal SCC and adenocarcinoma but not anal SCC\textsuperscript{(3)}. It was noted that Rasheed et al found SCC Antigen (Ag) to be elevated in three out of six patients\textsuperscript{(12)}. Dyson et al suggest that the SCC Ag level is not suitable for initial diagnosis of rectal SCC, but might be helpful to monitor disease response and progression\textsuperscript{(5)}. There is not concrete consensus regarding treatment of this disease entity. The primary treatment regarding colorectal SCC is surgical resection. Local excision or radical resection can be recommended depending on the characteristics of the tumor (size, location, depth of invasion and presence or not of local or distant metastasis). For most rectal SCCs, anterior or abdominoperineal resections are performed and occasionally requiring exenteration with removal pelvic of involved structures. Surgery, previously the preferred management, has been subsequently relegated to a salvage role. However, an increasing trend in the use of chemoradiation as definitive treatment is emerging. These treatment regimens use primaly 5FU in conjunction with cisplatin or mitomycin-C\textsuperscript{(3)}. They were de same drugs in treatment of anal SCC . Rasheed et al\textsuperscript{(12)} and Clark et al\textsuperscript{(13)} in two separate populations, evaluated the success of CRT in the treatment of squamous cell carcinoma of the rectum. Only three ended up with surgical resections. After histological evaluation, only one of the three resected specimens demonstrated residual tumor. Peron et al reported in his study of 11 patients treated by definitive CRT a partial response 4 to 8 weeks after finishing treatment. The clinical response to CRT was complete for 7 patients. The remaining 4 patients (36%) underwent salvage surgery. The pathologic response was incomplete for 2 of the 4 patients. For both patients, the surgery was performed 6 weeks after the end of the CRT, and the pathology report revealed a minimal residual disease\textsuperscript{(14)}. Our patient received a combined treatment with cisplatin and capecitabine along with external beam radiation therapy. The chemoradiation was followed by surgical excision. When radiation therapy was added preoperatively, Nahas showed there was an increase in sphincter preserving procedures from 67% to 71%\textsuperscript{(15)}. Guerra et al recommended that primary treatment should be CRT, with surgery reserved as a salvage option. They suggested regimen would be a total dose of 50.4 to 54 Gy external beam radiation in 1.8 Gy per fraction, given concurrently with 5FU and mitomycin C\textsuperscript{(3)}. Musio et al\textsuperscript{(16)} published a small series (8 patients) which were treated with radiotherapy (total radiation dose from 45 to 76.5 Gy) and chemotherapy (5 FU and mitomycin C most cases) combination. They reported only one recurrence that finally needed surgical resection. They concluded that high doses of chemoradiotherapy could be enough for an adequate tumor control without surgical resection. The most important predictor of survival is the stage of disease. While the majority of patients with rectal SCC present with locoregional disease (stage I-III), they are associated with a poorer overall survival (OS) when compared stage for stage with adenocarcinoma. From a review of the population study, the overall 5-year survival for rectal SCC was found to be 48.9% compared with 62.1% for adenocarcinoma. When localised, the 5 years OS was 73.7% versus 91.8% for adenocarcinoma, with 31.3% versus 65.8% for regional and 20.8% versus 8.8% for metastatic disease\textsuperscript{(17)}.

4. CONCLUSION

Rectal SCC is a rare neoplasm, with unknown tumor biology and pathogenesis. Based on our experience, we recommend definitive CRT for primary SCC of the rectum. Surgery should be reserved as a salvage option. In fact, it is necessary to individualize and approve the optimal treatment in multidisciplinary oncological team.

LIST OF ABBREVIATIONS


CONSENT SECTION

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

COMPETING INTERESTS

The authors declare that they have no competing interests.