

Case Report

A Rare Case of A Primary Squamous Cell Carcinoma of the Rectum

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Abstract

Squamous cell carcinoma (SCC) of the gastrointestinal tract is uncommon and usually involves the esophagus or the anal canal. More than 90% of tumors arising the colorectal region are adenocarcinomas. SCC is exceedingly rare accounting for 0.1 to 0.25 % of all colorectal malignancies. Due to its rarity and the lack of literature in this regard, and etiology and pathogenesis also remain obscure. However, we report here a rare case of squamous cell carcinoma of the rectum in a 68-year-old woman with no known comorbidities except a history of asthenia, loss of appetite and change in bowel habits for the last three months with 10 kg of weight loss over the last two months.

Keywords: Squamous cell carcinoma, gastrointestinal tract, rectum

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Introduction

Squamous cell carcinoma (SCC) of the gastrointestinal tract is uncommon and usually involves the esophagus or the anal canal. More than 90% of tumors arising the colorectal region are adenocarcinomas.¹ SCC is exceedingly rare accounting for 0.1 to 0.25 % of all colorectal malignancies.² Due to its rarity and the lack of literature in this regard, etiology and pathogenesis remain obscure. We report here a rare case of squamous cell carcinoma of the rectum.

Case Report

A 68-year-old woman with no known comorbidities presented to our hospital with a history of asthenia, loss of appetite and change in bowel habits for the last three months with 10 kg of weight loss over the last two months. Physical exam was unremarkable and digital rectal examination did not reveal a rectal mass. Laboratory data was normal. A colonoscopy was done which revealed a semi-circumferential lesion with friable mucosa in the rectum starting at 7 cm proximal to the anal verge. Biopsy of the mass was realized.

Histopathological examination favoured a poorly differentiated basaloid squamous cell carcinoma of the rectum (Figure 1). Immunohistochemical stains were performed and showed strong cytokeratin p63 and CK5/6 immunoreactivity of malignant cells, which supported our provisional diagnosis. However, to rule out a gynecologic origin of SCC, transvaginal ultrasound was performed. Cervical and endometrial biopsies also came out negative for malignancy. Pelvic MRI was realized and showed a 6 cm semi-circumferential mass posterior to the rectum extended beyond the serosa with mesorectal invasion and perirectal lymphadenopathy. The tumor was staged T3N1Mx. Pan-scan of the body failed to show any primary source of SCC or metastatic disease. CEA and CA19-9 were within normal limits. Neoadjuvant chemoradiation was performed. Pelvic MRI following chemoradiation showed regression in size of the tumor with persistent infiltration of mesorectal fat. The patient had a surgical operation and anterior resection of the rectum was performed. The post-operative course was uneventful. The histopathology report of the surgical specimen revealed a dense fibrous

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tissue dissociating the muscularis layer, without any carcinomatous residue (Figure 2).

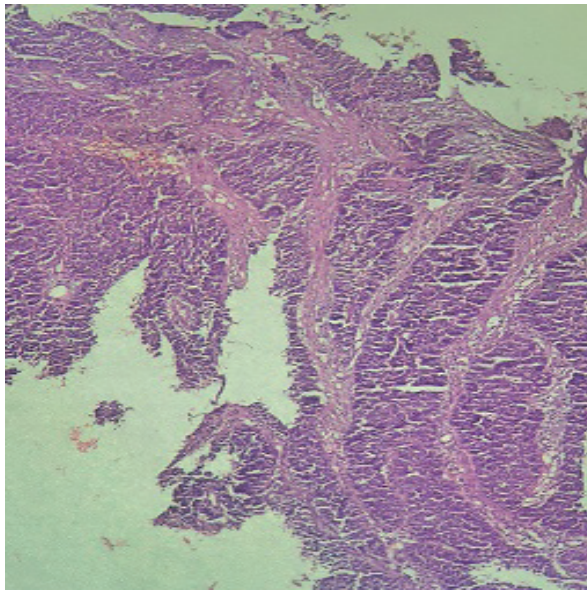


Figure1. Histopathology showing a poorly differentiated basaloid squamous cell carcinoma of the rectum (from colonoscopy specimen) (H & E staining × 250)

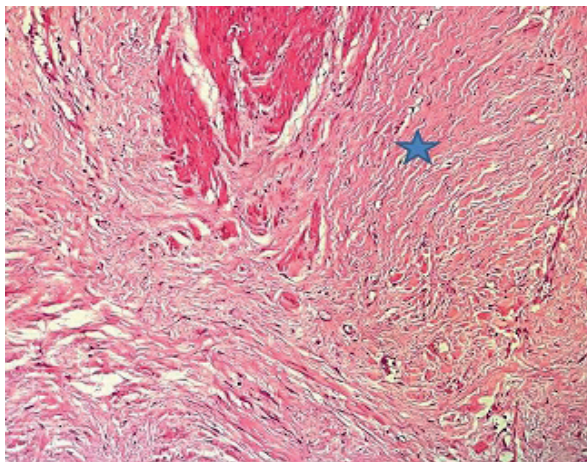


Figure2: Histopathology of the surgical specimen showing a dense fibrous tissue dissociating the muscularis layer, without any carcinomatous residue (star marked) (H & E staining × 250)

Discussion

Our case is interesting one as SCC of the rectum represents an exceptional location. With similarities to the adenocarcinoma, the SCC, in our patient, seems to have the same clinical presentation and the same evolution. Among the gastrointestinal tract, SCC commonly invades the esophagus and the anal canal. However, its localization in the rectum is extremely rare.³ There

are still many unanswered questions about this condition. Schmidtman reported the first case of SCC of the colon in 1919.⁴ Few years later, Raiford reported the first case of SCC of the rectum in 1933,⁵ and just over 200 cases of colorectal SCC have since been reported in the literature.

In 1979, Williams et al.⁶ proposed three exclusion criteria that remain essential for the diagnosis of primary SCC of the rectum. First, metastasis of the rectum from distant sites of SCC. Second, squamous cell-lined fistula involving the affected rectal region. Third, SCC of the anal canal or gynecological origin extending to the rectum. Similarly to rectal adenocarcinoma, patients with SCC usually present with change in bowel habits, loss of weight and appetite, abdominal pain and sometimes rectal bleeding.^{7,8}

The definitive diagnosis is confirmed by histological study from endoscopic biopsy. Varied endoscopic aspects have been reported, from small mucosal polyp, ulcerative lesion to large obstructing mass.⁹ In certain cases, especially when the tumor is poorly differentiated, histological findings are not decisive and immunohistochemistry can be useful to characterize the lesion. Cytokeratins AE1/AE3, CK5/6 and p63 stain positively for cells from squamous origin, helping in the differentiation from rectal adenocarcinomas. Cytokeratin CAM5.2 is helpful in differentiating between rectal SCC and anal SCC, its immunoreactivity suggests rectal tissue as the primary tumor site.¹⁰

As for rectal adenocarcinoma, pelvic MRI and CT scan of the chest, abdomen and pelvis are necessary for the evaluation of the primary tumor and assessment for loco-regional and metastatic disease.^{11,12}

Pathogenesis of rectal SCC remains unclear. One of the main theories is that chronic bowel inflammation may play a role by leading to squamous metaplasia and subsequent carcinoma. The second hypothesis suggests that squamous cell differentiation in the rectum by pluripotent mucosal stem cells may lead to dysplasia and SCC. Another hypothesis is that mucosal injury stimulates rectal basal cells to proliferate into squamous cells, with probable malignant transformation. Otherwise, some studies suggest

that associated HPV infection and pre-existing adenomas or adenocarcinomas may develop to SCC.¹²

Due to the rarity of the disease, therapeutic strategy is not well established. The treatment has traditionally involved surgery, preceded or followed by chemotherapy or radiotherapy in some cases, like in rectal adenocarcinoma.¹³ However, in last decade, the role of chemoradiotherapy is expanding. Recent analyses showed promising response to definitive chemoradiation with

improved outcomes, especially in localized rectal SCC. Surgery can be implemented for local failure or recurrence after chemoradiation as salvage therapy.¹⁴⁻¹⁶

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