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Case Report

Squamous Cell Rectal Carcinoma: About A Rare and Unusual Case: A Case report

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Abstract

Primary squamous cell rectal carcinoma is a rare malignancy that accounts for 0.3% of rectal tumors. To this date, no sure risk factors have been determined. The etiopathogeny is still unknown, despite the different suggested hypotheses. Specific diagnosis criteria have been set, to identify rectal squamous cell carcinoma from another entity. Moreover, due to its rarity, only few data exist on its management, and no standardized therapeutic regimen was set. We report a very rare case of primary squamous cell rectal carcinoma found in the middle rectum, in a 73-year-old woman. Rectos copy was used to visualize the tumor mass and to take biopsies; their histological study revealed a rectal location of a squamous cell carcinoma. After discussion of the case in a multidisciplinary consultation meeting, management consisted of definitive chemo-radiation.

Keywords: ***Squamous cell carcinoma, Rectum, Radiotherapy, Chemotherapy.***

Introduction

The most common histological type for colorectal cancers is adenocarcinoma accounting for 90% of colorectal cancers (CRCs), while for anal canal cancers it is squamous cell carcinoma. Primary rectal squamous cell carcinoma (RSCC) is exceedingly rare [1]. Several theories have been put forward to explain the pathophysiology of RSCC ranging from to the presence or pluripotent stem cells of the endodermal origin in the colorectal mucosa to the proliferation of basal cells into squamous cells due to damage to the colorectal mucosa [2]. While diagnosis is based on the colonoscopy findings, the confirmation will rely on the histological and immuno-histochemical examination. Different therapeutic strategies have been used, and the discussion about whether to go for definitive chemo radiation or curative surgery is still ongoing [2]. The interest of evoking this entity is to share data concerning this rare form of rectal tumor, improving the prognosis of patients presenting with it.

Case presentation

Patient Information

This is a case of a non-smoker 73-year-old woman, presenting with a year history of mucinous diarrhea at a rate of 3 to 4 diurnal and nocturnal stools/day, associated with intermittent rectal bleeding, an intense anorectal pain and tenesmus, evolving in a context of a pyrexia and marked weight loss. There were no significant medical comorbidities, no previous abdominal surgery and no family history of gastrointestinal malignancy.

Clinical findings

Physical examination was unremarkable, apart from an altered overall general condition and mucocutaneous pallor. Digital rectal examination revealed no mass but brought back bloody stools. The rest of the somatic examination, notably the examination of the lymph nodes and the examination of the respiratory system, was without anomalies.

Diagnostic assessments

Colonoscopy showed an ulcerated tumor located 8 cm away from the anal margin extended over about 4 cm, hemi-circumferential, friable and hemorrhagic at biopsy, narrowing the lumen but still crossable with the endoscope. Biopsies of this mass taken at the time of the colonoscopy corresponded to a largely ulcerated squamous membrane, made up of large cells, with anisokaryotic nuclei, sometimes voluminous, hyperchromic, seat of abnormal mitosis (**Figure 1**). The cytoplasms are abundant eosinophilic. These atypia occupy the entire epithelial height. Presence of suspected micro-invasion of the underlying chorion. The presence of foci of tumor necrosis is also noted. The histological study concluded to a squamous cell carcinoma with suspected foci of microinvasion, in the widely ulcerated material received. (**Figure 2**)

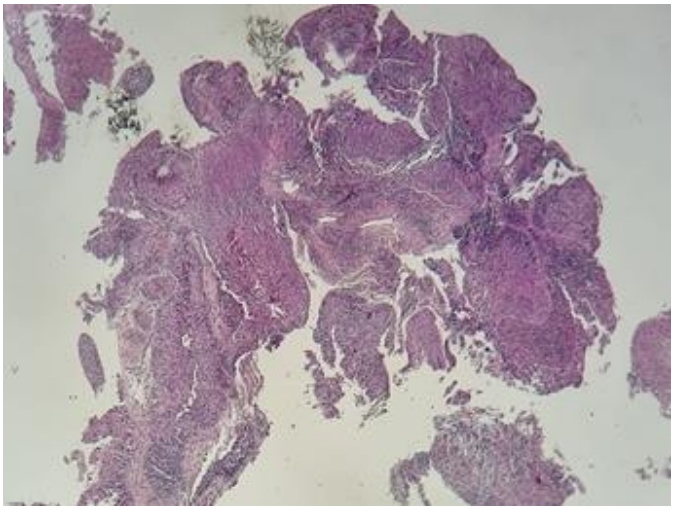
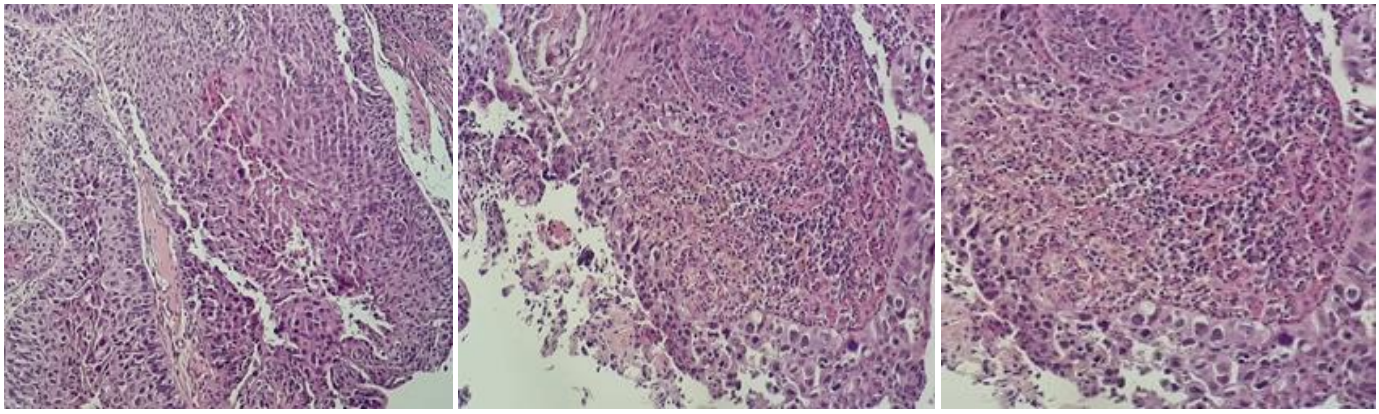


Figure 1: Magnification ×4. One fragment corresponds to a widely ulcerated squamous mucosa without analysable colorectal mucosa.



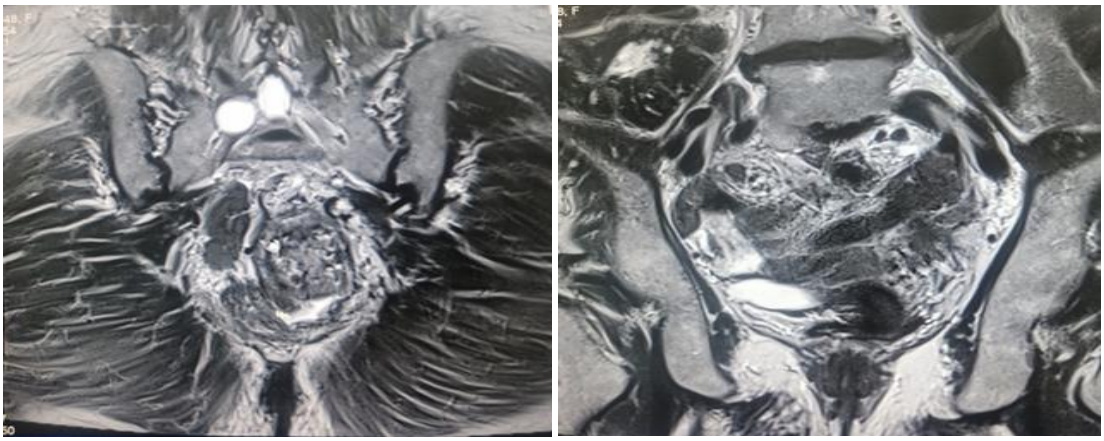
(a) **(b)** **(c)**

Figure 2: Magnification ×40: (A) The tumor cells are medium to large, with anisokaryotic, sometimes enlarged, hyperchromatic nuclei, with irregular outlines and abnormal mitoses. These atypical cells occupy the entire epithelial height. (B) The basement membrane is mostly continuous. There are localized foci of suspected microinvasion of the underlying chorion (Red arrows). (C) Abnormal mitosis (Blue arrow)

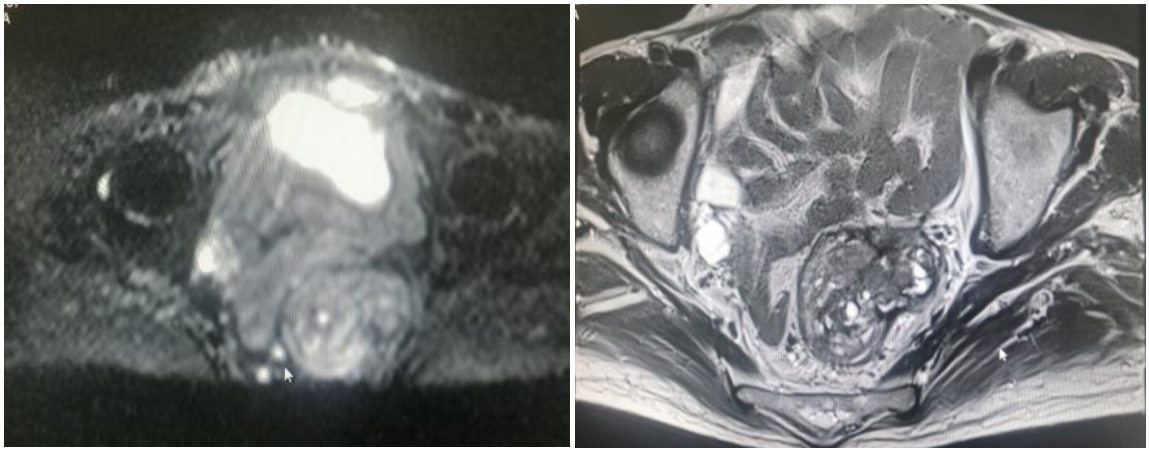
A pelvic MRI, showed a tumor of the upper and middle rectum, 8 cm from the anal margin, measuring 57*42*63 mm, budding at the level of the mesorectal fat and arriving in intimate contact with the fascia recti and close to the uterus and the uterine cervix with preservation of the fatty interface of separation, with multiple lymph nodes in the pelvis (**Figure 3**). A CT scan of the chest, abdomen and pelvis was negative for distal metastases. The stage of the tumor was found to be T3N+M0. Due to the squamous cell origin of her rectal mass, she underwent subsequent gynecologic evaluation, with cervical biopsies which were negative for malignancy.

Therapeutic Intervention

After discussion of the patient's file in a multidisciplinary consultation meeting (RCP), who decided to proceed with chemoradiation and to skip surgery. The patient is still receiving her treatment, consisting of a combination of cisplatin and fluorouracil (5FU) and external beam of radiotherapy. However, secondary surgery can be considered in case of local failure or recurrence as salvage therapy.



(a) **(b)**



(c) (d)

Figure 3: Abdominal imaging demonstrates a mass evolving in the upper and middle rectum, 8 cm from the anal margin, measuring 57x42x63 mm, budding at the level of the mesorectal fat (A,D) with conservation of the anal canal (B), with multiple nodules of the mesorectum (as shown in the diffusion sequence of the pelvic MRI) (C).

Discussion

The most frequent histological type for colorectal cancers is adenocarcinoma, representing 90% of colorectal cancers (CRC), while for anal canal cancers it is squamous cell carcinoma. Primary rectal squamous cell carcinoma is exceptionally rare, accounting for 0.10-0.25 per 1000 CRCs. Other histological types that can be observed in CRC are squamous cell carcinoma, neuroendocrine tumors, lymphomas, carcinoid tumors or leimyosacromas [1].

The first case of RSCC dates back to 1933 and was first reported by Raiford [3]. The vast majority of suspected cases of SCC of the rectum are actually SCC of the anal canal with proximal extension into the rectum, which is different from primary SCC of the rectum [3]. It is indeed a different entity, answering to the criteria set by Williams [4] to distinguish the two entities:

(a) Absence of SCC in another primary site that might spread directly into the bowel or that might provide a source for an intestinal metastasis;
(b) The tumor must not have a squamous-lined fistula tract to the affected bowel;
(c) The tumor cannot represent proximal extension of SCC of the anus and a lack of continuity between tumour and the anal epithelium has to be demonstrated, and finally
(d) Histological SCC confirmation.

We have progressed from questioning the mere existence of RSCC to accepting squamous metaplasia, either due to stress or chronic inflammation, as their origin. While multiple theories to explain their aetiology have been developed, the most accepted ones are chronic inflammation due to inflammatory bowel infections such as Human Immunodeficiency virus (HIV), Human Papilloma virus (HPV), amebiasis, chronic mucosal injury or radiation exposure as triggers for squamous metaplasia [5]. Tobacco use has also been incriminated in the pathogenesis of the RSCC [1]. RSCC appears to affect Caucasian individuals with an average age of 60 years, and in contrast to rectal adenocarcinoma, it affects most likely women [1,6].

The diagnosis is usually delayed, for patients can be asymptomatic for weeks. Patients generally present with one or more of the following symptoms: change in bowel habits such as constipation or diarrhea, rectal bleeding, lower abdominal pain, anorectal pain, tenesmus, weight loss and anorexia [7]. The diagnosis is based on a colonoscopy showing the tumor process with biopsy showing evidence of squamous cell carcinoma.

Clinical staging requires investigations including MRI of the rectum and trans-rectal endoscopic ultrasound (R-EUS). R-EUS provides information which can help the therapeutic approach, allowing for a better local lymph node evaluation. Yet it still hasn't shown superior results to endo-rectal MRI. Immunohistochemistry helps to differentiate rectal from anal lesions [8]. While this specific IHC stain was not used in our case due to its limited availability, the immunoreactivity of cytokeratin CAM5.2, an epithelial marker, suggests that the primary tumour site is the rectal tissue, rather than the anus [1]. Thus ruling out the tumor to be a proximal extension of an ASCC. While some authors, have used the TNM classification of the rectal adenocarcinoma, others haven chosen the ASCC staging system [9]. Due to the lymphatic dissemination of RCC, the most common metastatic sites are the liver, lungs and bones [5].

Colorectal SCC is considered to have a more aggressive behavior and worse prognosis than adenocarcinoma [2].

The RSCC is a rare entity. Therefore, no treatment guidelines have been established. The therapeutic regimens are usually derived from either rectal adenocarcinoma, due to the anatomical similarity or from ASCC because of the histological resemblance. While radical surgery was the most frequently used therapeutic approach for RSCC in analogy with rectal adenocarcinoma. A switch from curative surgery to definitive radiotherapy has been witnessed over the past half-decade [10]. Several studies actually recommend treating patients with RSCC like ASCC. Therefore, definitive chemo-radiation should be the primary treatment regimen, while surgery can be performed for local failure or recurrence as salvage therapy [4,11].

Radiotherapy volumes should include, apart from the tumor, the following regions: mesorectum, presacral nodes and internal iliac nodes. External iliac nodes should be added for T4 tumors involving anterior structures, while the inguinal iliac nodes should be considered for tumors located in the lower third of the rectum [11].

On the contrary, a recent large population based study with regional disease of rectal SCC shows an better results from the group undergoing multimodal therapy including surgery compared to those receiving a treatment without surgery, in terms of overall survival and disease-specific survival [10]. Thus re-opening the current discussion on the role of surgery in the treatment of RSCC.

Conclusion

It should be noted that primary RSCC is a very distinct entity that is markedly distinguishable from rectal adenocarcinoma and ASCC, and does not behave similarly to either one of them. The underlying etiology and the treatment strategy for RSCC remains controversial. Surgery as the primary treatment is now surpassed but still maintains its position in the treatment of patients with incomplete tumor response or local recurrence.

Authors Contributions

All authors participated in the conception, drafting the work, critically revised the manuscript, approved the final version to be published, and agree to be accountable for all aspects of the work.

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Declaration of conflicting interests

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