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Squamous cell carcinoma of middle rectum: Literature review

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ABSTRACT

INTRODUCTION: Squamous cell carcinoma SCC of the rectum is a distinct entity. We report a very rare case of squamous cell carcinoma of the middle rectum.
PRESENTATION OF CASE: The patient was a 62-year-old woman who presented with a history of rectal bleeding and discomfort. Colonoscopy revealed a polypoid tumour of the middle rectum. Biopsies of this mass revealed a poorly differentiated SCC of the rectum. CT scan of the chest, abdomen and pelvis was negative for distal metastases. The patient received combined chemo-radiation followed by surgical excision. The postoperative period was uncomplicated.
DISCUSSION: The pathogenesis of rectal SCC remains unclear and diagnosis is often delayed. Diagnostic criteria have been proposed. MRI of the rectum and trans-rectal endoscopic ultrasound R-EUS provide essential information to plan a therapeutic approach. The squamous cell carcinoma antigen level is not suitable for initial diagnosis of rectal SCC. Most authors conclude that the surgery is the gold standard treatment. Tumour stage is the most important prognostic predictor of SCC.
CONCLUSION: Squamous cell carcinoma of the rectum is a distinct entity. Before the final choice of treatment is made, digestive surgeons should bear in mind this rare tumour.

1. Introduction

Colorectal cancer is the third most common cause of cancer-related death in the world. Squamous cell cancer SCC is an unusual malignancy in rectum. Its incidence ranges from 0.1 to 0.25 per 1000 colorectal neoplasms. Its aetiology is not exactly known. Little information is available in the literature about the risk factors and natural history, prognosis and treatment of this cancer. We report a case of SCC of the middle rectum which was treated successfully with chemoradiation and surgical resection. The incidence, diagnostic criteria and management of this very rare tumour are discussed.

2. Presentation of case

A 62-year-old woman presented with a history of rectal bleeding and discomfort. The patient had neither prior cancer history nor a family history of colonic malignancy. Physical examination was unremarkable. Digital rectal examination revealed a mass about 8 cm from the anal verge on the right lateral wall of the middle rectum. Colonoscopy showed an ulcerated, polypoid tumour of middle rectum (Fig. 1). Biopsies of this mass taken at the time of the colonoscopy revealed poorly differentiated SCC of rectum: atypical SCC (Fig. 2). An abdominal CT scan showed a polypoid tumour (50 mm × 40 mm) with multiple lymph nodes in the pelvis. A CT scan of the chest, abdomen and pelvis was negative for distal metastases. The stage of the tumour was found to be T3N+M0. The endoscopic ultrasound and MRI of the rectum confirmed the results of colonoscopy and showed multiple lymph nodes in the pelvis. A positron emission tomography (PET) scan suggested the rectal tumour to be primary and was negative for distal metastases (Fig. 3).

The case was discussed by the multidisciplinary oncological team who decided that chemoradiation followed by surgical excision should be performed. The patient received a combined treatment with cisplatin and fluorouracil (5FU) along with external beam radiation therapy. She then underwent laparoscopic anterior resection. The standard technique of mesorectal excision and the concept of sphincter-preserving surgery were respected (anastomosis of the descending colon with the anus). The postoperative histopathological findings were in accordance with the diagnosis of atypical SCC of rectum that was classified as ypT3NOMO (Fig. 4). Tumor regression classification Dworak was estimated grade 3 (presence of rare tumor cells in fibrous remodeling). The margins of

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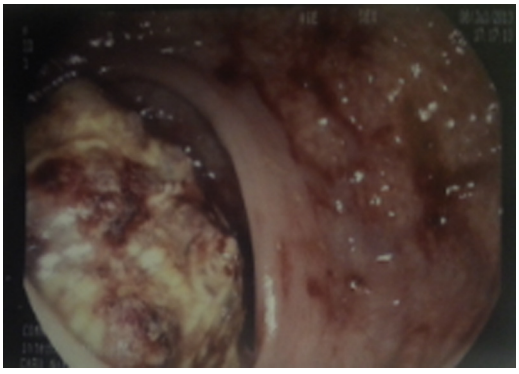

Fig. 1. Colonoscopy view showing the tumour of middle rectum.

the excised tissue were cancer-free. The lymph nodes were cancer-free. The postoperative period was uncomplicated.

3. Discussion

Malignant lesions of the rectum include secondary malignant tumours, intraepithelial neoplasm, adenocarcinoma, mucinous adenocarcinoma, adeno-squamous cell carcinoma, carcinoma in adenoma, neuroendocrine neoplasms, signet ring cell carcinoma, mixed carcinoid-adenocarcinoma, small cell carcinoma, medullary carcinoma, undifferentiated carcinoma, leiomyosarcoma, Kaposi's sarcoma, angiosarcoma, gastrointestinal stromal tumour (GIST), squamous cell carcinoma, malignant lymphomas, malignant melanoma.

The incidence of SCC ranges from 0.1 to 0.25 per 1000 colorectal neoplasms.[1] Schmidtmann reported the first case of pure SCC of the colon in a 65-year-old man in 1919.[2] Raiford reported the first case of squamous cell cancer SCC of the rectum in 1933.[3] The SCC of the rectum is more frequent than SCC of the right colon.[4] SCC of the rectum appears affect individuals with a mean age of 57 years. Based on a review of the literature, 66% of cases occurred in women and 34% in men. Table 1 summarizes some of the reported cases.

The aetiopathogenicity of SCC of the rectum are still unclear. Several hypotheses have been proposed. Some suggest that chronic irritation (radiation exposure) can cause squamous metaplasia and subsequent tumour development. Hicks et al. suggest that the pluripotent stem cells are capable of squamous differentiation.[5] Others have described the possibility of squamous differentiation of adenoma and adenocarcinoma.[6] Nahas et al. suggest a common cellular origin for rectal SCC and rectal adenocarcinoma, which is different from anal SCC.[7] The risk factors of SCC of the rectum are still elusive. There is no ethnic predilection for this tumour. The association seen between SCC of the rectum and human papilloma virus has not been established. Coexisting infections have been described including entamoeba histolytica colitis and schistosomiasis. Some case reports have found SCC in association with prostate cancer, ovarian cancer and endometrial cancer.

The diagnosis of SCC is often delayed. The symptoms can persist for several weeks to months.[8] The symptoms of the patients with SCC of the rectum are similar to those with adenocarcinoma: rectal bleeding, change in bowel habits, abdominal pain and weight loss.[9] Before labelling the diagnosis as colorectal SCC, Williams et al. have proposed reasonable criteria which must be fulfilled[10]: absence of extension of the tumour from the anal squamous epithelium; absence of evidence of squamous cell carcinoma of any other primary site; absence of squamous-lined fistula tract to the affected bowel; confirmation of SCC by histological examination (without glandular differentiation). In our patient, all of these criteria were fulfilled.

MRI of the rectum and trans-rectal endoscopic ultrasound R-EUS provide essential information on therapeutic approach. R-EUS provides better local lymph node evaluation but it has yet to be shown to be superior to endo-rectal MRI. Immunohistochemistry helps to differentiate rectal from anal lesions. The most useful cytokeratins are CAM 5.2, 34B12 and AE1/AE3.[11] CAM 5.2 stains rectal SCC and adenocarcinoma but not anal SCC.[7] It was noted that Rasheed et al. found SCC Ag to be elevated in three out of six patients.[12] Dyson et al. suggest that the squamous cell carcinoma antigen (SCC Ag) level is not suitable for initial diagnosis of rectal SCC, but might be helpful to monitor disease response and progression.[11]

Most authors conclude that the surgery is the gold standard treatment for colorectal SCC.[1] Based on a review of the literature, the treatment options in function upon tumour location include: chemotherapy alone, radiation therapy alone, surgical excision alone, chemoradiation alone, surgical excision followed by chemoradiation and chemoradiation followed by surgical

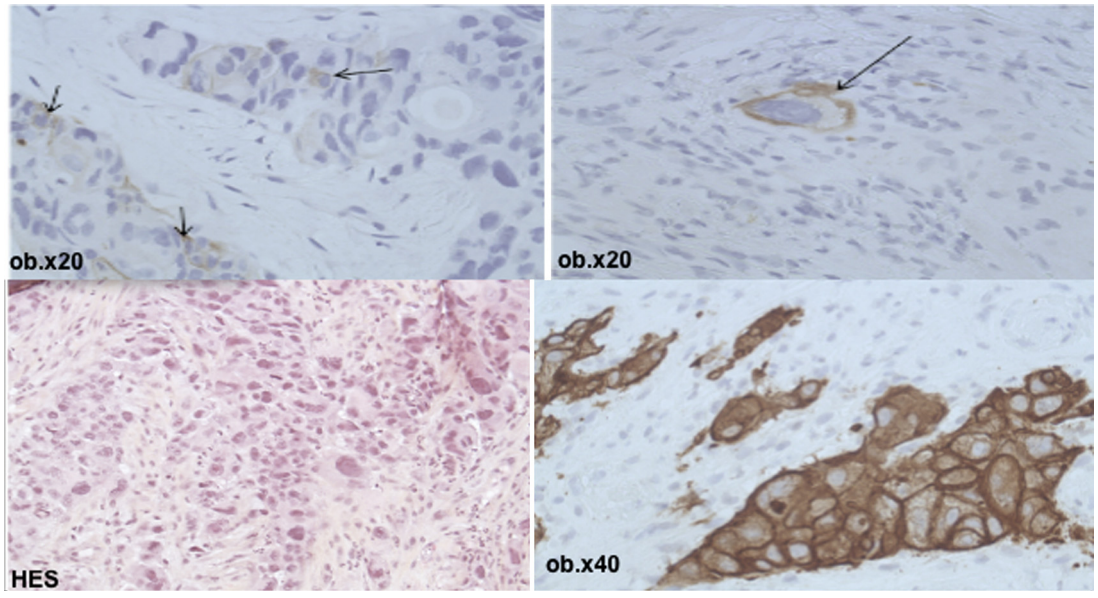

Fig. 2. Poorly differentiated carcinoma composed of large cells with large nuclei, very irregular, often hyperchromatic without keratinization phenomenon. Abundant cytoplasm (HES ob. 20×). Tumour proliferation shows a low cytoplasmic and membrane staining CK20 (ob. 20×). Cytoplasmic immunostaining CK5/6+++ (ob. 40×).