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Case Report

Primary signet ring carcinoma of the rectum: a rare entity

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ABSTRACT

Signet ring cell carcinoma constitute an uncommon histological type of rectal cancer with less than 1% of all rectal neoplasms. It usually behaves aggressively and has an inferior prognosis. Herein, we present a rare case in young male diagnosed by trucut biopsy.

Keywords: Signet ring cell carcinoma, Rectum, Rare

INTRODUCTION

Signet ring cell carcinoma (SRCC) is a variant of mucinous carcinoma identified microscopically by tumor cells with peripherally pushed nuclei and abundant intracytoplasmic mucin. SRCC most commonly arise in the stomach but can be seen in other organs like breast, gallbladder, pancreas, urinary bladder, and colorectum.[1] Colorectal signet ring cell carcinoma is a rare histological type with an incidence rate between 0.1% and 2.6% of all primary colorectal cases reported in literature.[2,3] Signet ring cell morphology should be present in >50% tumor cells for the diagnosis. It is a clinicopathological distinct entity with respect to colorectal adenocarcinomas in terms of onset at young age, high rate of distant metastasis, lower rate of liver metastasis, increased rate of peritoneal seeding and poor prognosis.[2] Moreover, SRCC has a high tendency for intramucosal spread with relative sparing of the mucosa. Hence, tumor cells are rarely identified in some cases in regular biopsies.[4,5]

Most case series report a predilection to males, with male to female ratio of 2:1, and is usually found in the right colon or the rectum.[6] We are reporting a case of SRCC of rectum in a young male in which diagnosis was made by trucut biopsy.

CASE REPORT

A 26 year old young male presented in surgery outpatient department (OPD) of our hospital with sudden, intermittent painless bleed per rectum following meals and significant weight loss since one year. He had no past history of similar illness. The family history was not contributory to his symptoms. General physical examination his pulse rate was 104/minute and blood pressure was 108/72 mm Hg. Patient was pale with cachexic look. Rectal examination revealed an indurated, fixed palpable growth obstructing the rectal lumen.

Investigations

Ultrasonography **(**USG) of abdomen showed a normal study. Thoracoabdominal contrast enhanced computed tomography (CECT) scan revealed few calcific granulomas in the upper and lower lobe of right lung and segment VII of liver. There was no evidence of focal mass lesion in the liver. Plain magnetic resonance imaging (MRI) of pelvis in axial and coronal planes with correlative

fast-spin echo (FSE) T2W1 in the sagittal plane was done. It revealed a growth in rectum with infiltration of internal sphincter on the left side and perirectal fat and lymph nodes. The stage given was T4b N2 M1, with extramural venous invasion I (EMVI) positive and circumferential resection margin (CRM) positive (Figure 1).

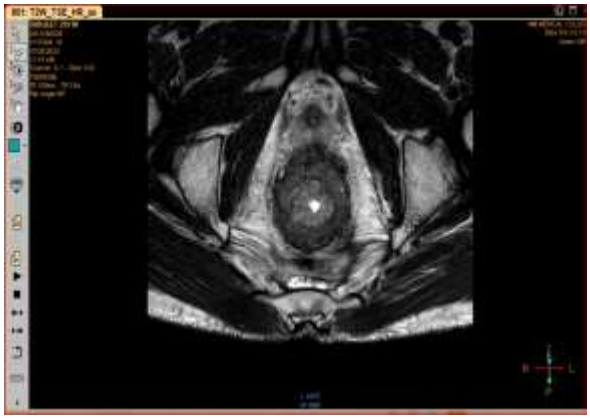

Figure 1: T2W1 MRI of pelvis revealed circumferential thickening of rectal wall due to growth.

We received a trucut biopsy from rectal mass in histopathology department. After routine processing and sectioning, it was reported as signet ring cell carcinoma rectum. On microscopy there were two cores of the tissue. Areas of coagulative necrosis (Figure 2a) and infiltration of signet ring cells into the muscle were noted (Figure 2b). The cells have crescentic nucleus with abundant vacuolated cytoplasm which showed positivity for PAS (Figure 2c).

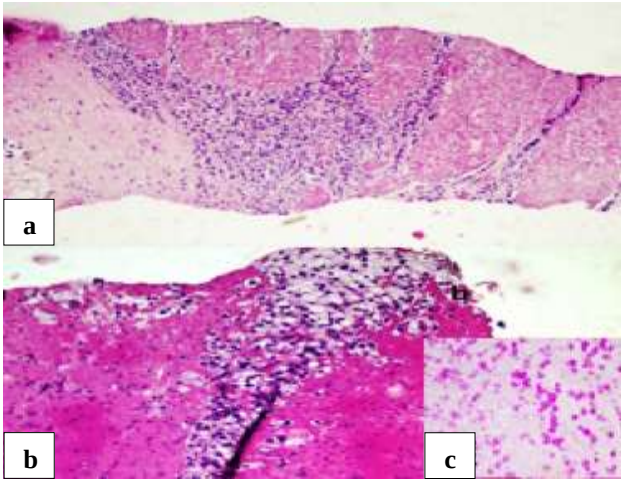

Figure 2: (a) Infiltrating signet ring cells, areas of coagulative necrosis (H&E, 100x) (b) signet ring cells with crescentic nucleus and abundant vacuolated cytoplasm infiltrating into the muscle (H&E, 400x) and (c) inset showing signet ring cells with peripherally pushed crescentic nucleus and abundant magenta colored vacuolated cytoplasm (PAS stain, 400x).

DISCUSSION

Laufman and Saphir described primary SRCC of the colon and rectum in 1951. SRCC of rectum is a rare subtype of all colorectal carcinomas. Macroscopically, SRCC exhibits thickening of the bowel wall resembling to the gastric malignancy linitis plastica, as a shrunken, rigid structure. Histologically, tumor cells resemble signet ring morphology because of abundant intracytoplasmic mucin, which pushes the nuclei to the periphery. The presence of more than 50% of signet ring cells are necessary to label signet ring cell carcinoma.[7] On the basis of gross and microscopy features, the differential diagnosis of this entity include mucinous adenocarcinoma, medullary, adeno squamous, micropapillary, spindle cell and undifferentiated carcinomas of the rectum. With the help of above-mentioned diagnostic criteria, signet ring cell carcinoma can be differentiated from these entities. One of the best parameters depicting the biologic behaviour of colorectal carcinomas is the presence of mucus secretion in microscopic examination.[6]

Besides being a rare entity, colorectal SRCC is regarded to have poor prognosis in comparison to conventional colorectal adenocarcinomas due to its high proneness for diffuse intramural infiltration, peritoneal dissemination, lymph node involvement and distant metastasis.[8] Indeed, previous age- and sex-matched controlled study has showed that the survival rate of patients with SRCC was considerably lower than that of adenocarcinomas with independent predictive factors, such as the staging of tumor at the time of diagnosis and presence of distant metastasis.[8,9]

CONCLUSION

SRCC of rectum is a rare entity with more aggressive behaviour and poor prognosis. The median age of presentation is under 40 years with male predominance. Trucut biopsy can be used as an easy tool for timely diagnosis of this uncommon tumor.

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