Case Report

A case of rectal cancers in teenager: A conundrum of genetics and clinical medicine

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ABSTRACT

Introduction: Signet cell carcinoma (SRCC) of the rectum is a rare subtype of the rectum cancer which accounts for only 0.8% of colorectal cancer in adolescents and young adults (AYAs) which spread aggressively to other organs and peritoneum.

Case presentation: We present a case of 15-year-old boy from rural area, presented with chronic diarrhea and per rectal bleeding for 3 months. The diagnosis was determined by colonoscope which revealed a fungating mass identified at 10cm from anal verge. Histological examination confirmed diagnosis of signet ring cell adenocarcinoma. CT scan of the abdomen showed thickening involving the recto-sigmoid colon and rectal mass, without evidence of distant metastatic disease. The patient’s carcinoembryonic antigen level was within the normal range. He underwent a colostomy and was subjected to neoadjuvant CCRT and surgery.

Discussion: This CASE highlights the importance and challenges in achieving early diagnosis and surgical intervention of signet-ring cell carcinoma in adolescents, as most cases are detected at an advanced stage coupled with the scarcity of information on these rarer subtypes which leads to a poor prognosis.

Conclusion: In managing Signet cell carcinoma of the colorectal, physician have to know that it has a poor prognosis in patients of any age. However, in young teenagers delayed diagnosis and treatment option are narrowed to palliative management. Genetic profiling of family members and similar environment population may be a key to early detection.

1. Introduction

Signet cell carcinoma is a rare subtype of the rectum which accounts for only 0.8% of colorectal cancer in adolescents and young adults (AYAs) which spread aggressively to other organs and peritoneum. Proportions of cases with mucinous adenocarcinoma and signet ring cell carcinoma histopathologic subtypes significantly increased with younger age at onset [1]. Signet ring cell carcinoma is characterized by the abundant intracytoplasmic mucin that pushes the nucleus to the periphery giving a signet ring cell appearance. In order to meet the WHO classifications, signet cell should account for at least 50% of the cells [1]. We report a Case of signet cell carcinoma (SRCC) in a 15 year old boy. He presented with chronic diarrhea and rectal bleeding for three months. All literature points to the delay in diagnosis as the reason for worse clinical outcome in younger patients. The scarcity of information on these rarer subtypes merits further study and investigation. This case report has been reported in line with the SCARE 2020 criteria [2].

2. Case presentation

A 15 year old boy presented with history of chronic diarrhea for 3 months aggravated with passing fresh blood and progressive suprapubic pain. He is a non-smoker. He has paternal and maternal family history of colorectal cancer. Previously an active boy, he had to stop schooling due to uncontrolled bowel output and rectal bleeding.

Laboratory examination revealed liver function, random blood glucose and renal function tests were all within normal parameters. However, mean corpuscular volume was significant at 63.3 fl. Electrolytes were normal except for sodium of 131 meq/liter, albumin 28 and urea 12.5. His tumor markers such as carcinoembryonic antigen level, Alpha feto protein and CA 125 were within the normal range.

CT scan of the abdomen showed thickening at the recto-sigmoid area...
Signet ring cell adenocarcinoma (Fig. 2). Sigmoid colostomy was done due to was unable to pass beyond the tumor. Biopsy results confirmed signet cell adenocarcinoma (Fig. 2). Sigmoid colostomy was done due to impending obstruction. He was then treated with the 6 cycles of FOLFOX regimen (folinic acid, fluorouracil, oxaliplatin) with concurrent radiotherapy. A restaging evaluation with chest and abdominal CT after completion of the treatment showed no response to treatment. The treatment was then changed to the FOLFIRI regimen (folinic acid, fluorouracil, irinotecan).

However, the patient subsequently defaulted treatment due to logistic and financial issues which prevented him from travelling from his home which was situated deep in the rural area of Borneo.

He then came back 1 year later with intestinal obstruction symptoms. Due to tumor and disease progression with metastatic features on restaging CT patient was manage as palliative. The patient’s performance status declined afterward, and he was transferred to supportive care unit. The patient was placed on patient controlled analgesia (PCA) hydromorphone and TPN. Patient subsequently succumbed to the disease and passed away.

3. Discussion

Signet cell cancer of the colon is a rare subtype of colorectal cancer, with no distant metastatic disease. Colonoscopy findings was a constricting ulcerative mass at 10 cm from anal verge which the scope was unable to pass beyond the tumor. Biopsy results confirmed signet ring cell adenocarcinoma (Fig. 2). Sigmoid colostomy was done due to impending obstruction. He was then treated with the 6 cycles of FOLFOX regimen (folinic acid, fluorouracil, oxaliplatin) with concurrent radiotherapy. A restaging evaluation with chest and abdominal CT after completion of the treatment showed no response to treatment. The treatment was then changed to the FOLFIRI regimen (folinic acid, fluorouracil, irinotecan).

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Ethical approval

No ethical approval required for the mentioned case report. However, permission was obtained from local administrative and Director General of Health Ministry, this included consent from patient family.

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Author contribution

Dr Moveendra Kumar took lead in writing the paper. Dr Nik Amin Sahid arranged the framework of manuscript with supervised correction and Dr Mahadevan Deva Tata encouraged Dr Moveendra to investigate colorectal cancer in adolescence and supervised the writing. All authors discussed the final draft and contributed to final manuscript.

Research registration number

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3. Hyperlink to your specific registration (must be publicly accessible and will be checked). Not Applicable

Guarantor

Dr Moveendra Kumar will be the guarantor and accepts full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish at this given time of submission.

Consent

Patient consented for the write up and publication.

Declarations of competing interest

Dr Moveendra Kumar, Dr Nik Amin Sahid and Dr Mahadevan Deva Tata declare that they have no conflict of interest.

Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.amsu.2021.102353.