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**Signet Cell Adenocarcinoma of the Rectum in 16-year-old Male Patient. (Case Report)**

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doi: 10.51505/ijmshr.2021.5221

URL: <http://dx.doi.org/10.51505/ijmshr.2021.5221>

**Abstract**

Colorectal carcinoma (CRC) is a common carcinoma of the colon and rectum, typically observed in men and women worldwide. There are many types of colorectal carcinomas, but colorectal adenocarcinomas constitute the most common type (over 90% of them). Signet Ring-Cell Carcinoma (SRCC) of Colon and Rectum is a rare histological subtype of colorectal adenocarcinoma. They constitute less than 1% of all colorectal adenocarcinomas and are known to be highly-aggressive tumors. The (SRCC) Median age is about 59 years (1).

This is a case report of a very young male with Signet Ring-Cell Carcinoma (SRCC) of rectum; this paper will help to raise physicians and pediatricians awareness about Signet Ring-Cell Carcinoma (SRCC) of Colon and Rectum.

**Keywords:** Cancer (CRC), Adenocarcinoma, Signet Ring-Cell Carcinoma (SRCC), Inflammatory Bowel Disease (IBD)

**Introduction**

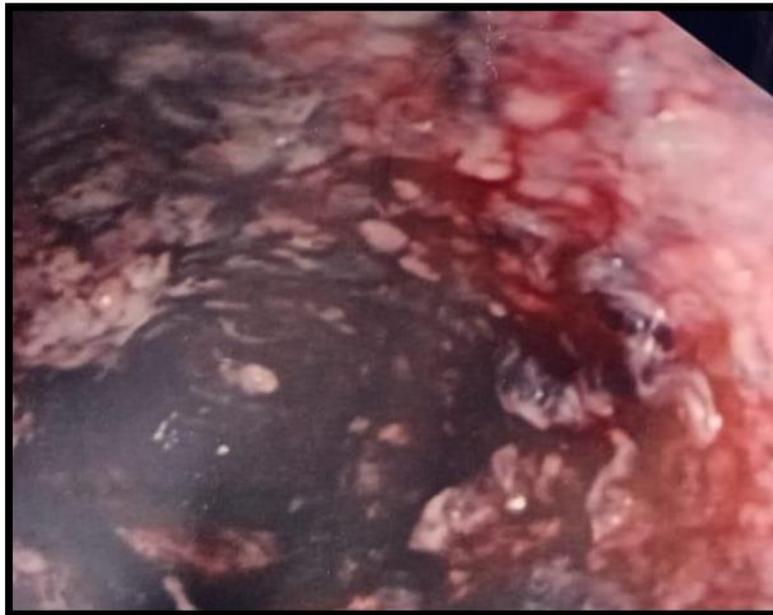
Colorectal cancer is a frequent cancer in the adult age group. It is the second-and third-most common cancer in women and men, respectively (1). The majority of patients with sporadic cancer are >50 years of age, with 75% of patients with rectal cancer and 80% of patients with colon cancer patients being  $\geq 60$  years of age at the time of diagnosis (1). Yet CRC incidence among young adults age < 50 continues to increase without a clear explanation (2). Colorectal carcinoma in patients under 40 years of age usually has a poor prognosis (3). SRCC of colon and rectal origin is very rare and comprises only 1% of colorectal cancers. Rarer still is presentation in the teenaged patient, especially in the absence of any risk factors (4). The cause of development of Signet Ring-Cell Carcinoma of Colon and Rectum is generally unknown (5). Research scientists believe that the cause of the condition is mostly due to genetic mutations, influenced by factors that include food and lifestyle habits such as high-fat, high-calorie diet and sedentary lifestyle (5). Currently, a very limited number of SRCC cases in young

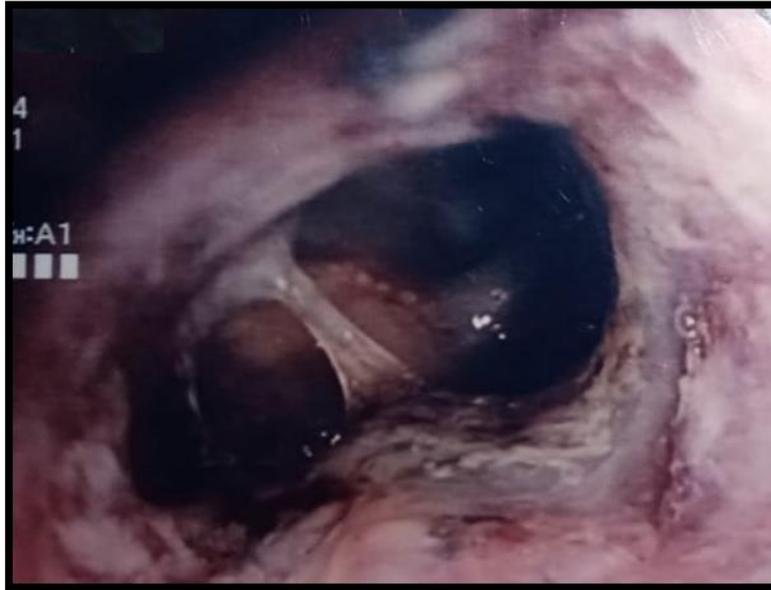
patients have been published. We presented a case of a teenaged male with stage IIIA Signet Ring-Cell Carcinoma (SRCC) of rectum

### **Case Presentation**

Sixteen-year-old male patient previously healthy presented to our department for one- week history of abdominal discomfort, several episodes of rectorrhagia, loose stool and Decrease appetite, no family history of Inflammatory Bowel Disease (IBD) or malignancy.

On physical exam, he has mild abdominal tenderness mainly in the left lower quadrant, fresh blood on the Digital rectal exam. Laboratory results were normal except for anemia 10.5g/dl. Colonoscopy was done and showed Diffuse severely ulcerated friable mucosa with necrotic tissues, extending 10 cm from the anal verge till 25cm. Biopsies showed abundant intracytoplasmic mucin pushes the nucleus to the periphery giving a Signet Ring appearance compatible with Signet cell adenocarcinoma, confirmed with another pathologist reading center. MRI abdomen and pelvis showed marked circumferential thickening of the wall at the recto-sigmoid junction 14 mm in thickness (NL 0.2-2.5 mm), extending 15 cm in length from the mid rectum to the sigmoid colon. With diffuse marked submucosal edema, multiple round nodules are present in the mesorectal fat, and the largest measures 2 cm. Patient was in stage IIIA and started on FOLFOX before operation.





### Discussion

- The median age of colon cancer is 68 for males and 72 for females (6). Early recognition of colorectal cancer (CRC) in young patients without a known genetic predisposition is a challenge, and clinic-pathologic features at time of presentation are not well described (7). Childhood CRC is a very rare disease entity. The incidence of CRC in children is 0.3 to 2 cases per 100, 000, making 0.4% of all malignant tumors with the highest mortality rate in patients below 15 years (8).
- Signet Ring-Cell Carcinoma (SRCC) of the colon and rectum is very rare and presents at a younger age compared to non-signet cell adenocarcinoma (9). The majority of publications regarding SRCC of colon and rectum demonstrate an average diagnostic age of 40. There are few cases of colonic signet-ring cell carcinoma, with fewer than 20 cases in the literature (10). Rarer still is SRCC of colon and rectum in the teenaged patient. Most tumors are located either in the right or left side of the large intestine, but rarely in the rectum. The location depends on the type of genetic defect observed.
- The symptoms may vary based on the location of tumor in the colon (whether tumor is to the right or left side).The signs and symptoms may include abdominal pain, bleeding from the rectum, fatigue, and weight loss. Complications, such as tumor metastasis to distant sites, are known to occur (5).
- Both males and females are affected, although more number of cases are observed in males. All races and ethnic groups are at risk for the condition. In general, a higher number of cases of colorectal carcinoma are reported from certain well-developed regions of the world such as Australia and New Zealand, Japan, USA and Canada, and European countries. Parts of Africa, India, Pakistan, and underdeveloped/developing geographical regions report a much lower incidence (5).

- Prognosis of the disease is influenced by factors such as aggressive histological subtypes (Signet Ring and mucinous adenocarcinoma), advanced tumor grade, and advanced stage of the disease (11).
- Colonoscopy is the gold standard of investigation during establishment of diagnosis of CRC in children and adults. Other advantages of this investigation include taking photographs and biopsy from the lesions (8).
- Surgery is performed in most cases, unless tumors are small and superficially located. In case of lymph node metastasis, surgery is followed by chemotherapy and/or radiation therapy (adjuvant therapy). In case of rectal carcinoma, chemotherapy and/or radiation therapy, to shrink tumor before surgery, may be provided (neoadjuvant therapy). The prognosis of Signet Ring-Cell Carcinoma of Colon and Rectum is generally poor, in a majority of cases, since it is a highly-malignant tumor; overall 5-year survival is around 12% or less. The prognosis is typically much worse than conventional colorectal adenocarcinomas. A poor prognosis is dictated by advanced stage, surgical resection with positive margins (indicating that the some cancer cells are left behind following surgery), and tumors obstructing of the colon (5).

### Conclusion

We report a case of signet-ring cell adenocarcinoma of rectal origin in a teenaged patient. This diagnosis is very rare, especially in this age group. Signet cell cancer of the rectum and colon is aggressive with a poor prognosis. Any delay in diagnosis is significant as this cancer is frequently associated with an advance stage at presentation.

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