

# Metastatic Primary Signet Ring Cell Carcinoma of Rectum: A Case Report of 10-Year-old Male Child

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## ABSTRACT

Signet ring cell carcinomas of the colon and rectum are well documented in the adult population, but the incidence is very low in the paediatric population. Signet ring cell carcinoma has more malignant potential, mostly present as advanced stage and carries very poor prognosis. We are describing a 10-year-old male patient who presented with acute intestinal obstruction; radiology revealed large bowel obstruction and was diagnosed metastatic primary signet ring cell carcinoma of rectum on biopsy. We have discussed the diagnostic work-up and the management of this rare entity. Due to the high mortality that can be caused by a delay in making the correct diagnosis, signet ring cell carcinoma of colorectum represents a special diagnostic and surgical challenge.

**Keywords:** Mucinous Tumor, Colorectal Neoplasm, Intestinal obstruction

## CASE REPORT

A 10-year-old boy presented in emergency department with chief complaints of abdominal distension and vomiting for two days. He had history of intermittent lower abdominal pain, and altered bowel habits since two months. There was no history of bleeding per rectum. Family history was not significant. General physical examination revealed a healthy appearing male child. The blood pressure was 100/80 mm of Hg, pulse rate 110/minute and respiratory rate 22/minute. On examination abdomen was grossly distended, bowel sounds were increased. Generalised tenderness, guarding and rigidity present.

On digital rectal examination, a circumferential hard constricting growth was palpable about 5 cm from the anal verge. X-ray chest was within normal limit. X-ray abdomen was suggestive of large bowel obstruction. Routine investigations like Hb, serum Na<sup>+</sup>/K<sup>+</sup>, blood urea, liver function tests were within normal limits. With above findings, clinical diagnosis of acute intestinal obstruction due to rectal growth was made and laparotomy was planned. Laparotomy revealed gross distension of whole colon. Serosal surface showed multiple tiny nodules. A stricture was palpable in distal sigmoid colon and rest of Pelvis was frozen. Peritoneal seeding was noted and omentum was caked. Stomach, liver and spleen were normal. A diagnosis of non resectable advanced carcinoma rectum was made and considered for palliative surgery. Sigmoid colostomy with omental biopsy was done. omental biopsy shows features of metastatic deposit of adenocarcinoma having signet ring differentiation [Table/Fig-1]. The immediate postoperative course

was uneventful and FOLFOX regimen (5 Fluoruracil, calcium folinate, oxaliplatin) was started. But unfortunately patient was lost to follow up after two cycles.

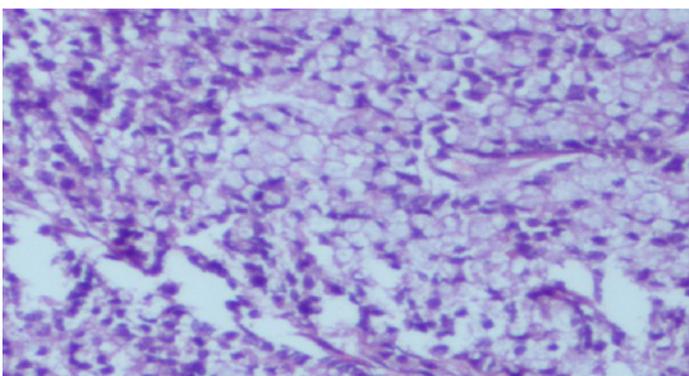
## DISCUSSION

Signet ring cell carcinoma is a rare subtype of adenocarcinoma, where abundant intracytoplasmic mucin pushes the nucleus to the periphery giving a signet ring appearance. The carcinoma can occur in many different sites of the body but more than 96% of signet-ring cell carcinomas arise in the stomach. However, it can also originate from the colon, rectum, gallbladder, pancreas, urinary bladder, and breast. Primary Signet ring cell carcinoma of the colon and rectum was first described by Laufman and Saphir [1]. Primary Signet ring cell carcinoma of colon and rectum is a distinctive malignant disease and rarer than conventional adenocarcinoma, with a reported incidence ranging from 0.01% to 2.6% [2]. Primary signet ring cell carcinoma of colorectum is diagnosed when the following criteria are satisfied. Firstly tumor must be primary, histological material is adequate and signet ring cell present more than 50% of cancer [3]. All the three criteria were satisfied in our case. In our case clinical history and laparotomy ruled out primary growth in the stomach. Immunohistochemical staining profiles for MUC1, CDX2 and MUC2 have been used to characterize and differentiate SRCC of breast, stomach and colorectum [4].

Signet ring cell carcinoma of colorectum has an aggressive clinical course and poorer prognosis. There is high incidence of peritoneal metastases and relatively low incidence of hepatic metastases, a characteristic feature distinguishing colorectum signet ring cell carcinoma from non signet colorectum carcinoma [5].

In the literature, signet ring cell carcinomas tend to affect predominately adult individuals. Median age is about 59 years when compared to the non signet cell cancer, where the median age is about 61 years [6]. Our case is rare because of very young age of presentation of this cancer. The cases with colorectal signet ring cell carcinoma in young patients in the literature are shown in [Table/Fig-2].

The most common presenting symptom for the children with colorectal signet cell carcinoma is abdominal pain and vomiting. Other symptoms include rectal bleeding, mucus in stool, change on bowel habits and weight loss. These clinical symptoms resemble those of irritable bowel disease; hence the diagnosis of colorectal signet ring cell carcinoma in children is based on a high index of



**[Table/Fig-1]:** Photomicrograph showing tumour cell having intracytoplasmic mucin pushing the nucleus to the periphery and giving a signet ring appearance (H&E x100)

Author's	Age-Sex	Symptoms	Site of tumour and histology	Stage at presentation	Treatment	Survival
Özgül et al., [7]	19years, m	Persistent abdominal pain, weight loss	Sigmoid colon/signet ring cell carcinoma	Advanced	Palliative colostomy FOLFOX-6 +Bevacizumab	ND
Sun Hyung Kang et al., [8]	21years, m	Hematochezia	Polyp at proximal rectum/signet ring cell carcinoma	Early	Local resection with end to end anastomosis	ND
Jagtap SV et al., [9]	31years, m	Persistent abdominal pain, vomiting Subacute intestinal Obstruction	Rectosigmoid/signet ring cell carcinoma	Advanced	Radical resection with end to end anastomosis	ND
Pandey A et al., [10]	10years, m 11years, m 11years, m 10years, m	All patients present with Bleeding per rectum, pain abdomen	Rectosigmoid/signet ring cell carcinoma	Advanced Advanced Advanced Advanced	Transverse colostomy Transverse colostomy Refused treatment Surgical resection	One year 11 month ND One year
Marone J et al., [11]	17years, m	Progressive abdominal pain	Ascending colon/signet ring cell carcinoma	Advanced	Right colon resection with ilioocolic anastomosis	One year
Kumkum Singh et al., (Present Study)	10years, m	Abdominal pain & distension, vomiting	Rectosigmoid, Signet ring cell carcinoma	Advanced	Palliative Colostomy, FOLFOX	ND

**Table/Fig-2:** Comparison of Colorectal signet ring cell carcinoma in young patients in the literature  
ND:Not determined

suspicion. Because of the lack of awareness at early age, vague symptoms and the aggressive characteristic of the tumour, signet ring cell carcinomas of the colon and rectum mostly present as advanced stage.

Colorectal malignancy can be confirmed by digital rectal examination (DRE), sigmoidoscopy or colonoscopy. Contrast studies, ultrasonography and computed tomography are essential procedures for detecting the extent of the disease. Surgery is the most effective treatment for colorectal cancer. It is to be stressed that the survival is dependent on the complete resection of the cancer and aggressive surgical procedures can increase the survival rates of these patients. In patients with resectable mass, complete tumor resection that includes the lymphatic basin of the affected colon and/or rectum has the greatest impact on the overall survival. The reported median survival time ranges from 15 to 32.6 months [12]. Although adjuvant chemo-radiotherapy has limited value of benefit in most of the cases, a variety of new agents such as irinotecan, oxaliplatin and leucovorin used in association with 5-FU should be considered.

## CONCLUSION

Colorectal malignancy in children are rare, hence any child presenting with pain in abdomen along with doubtful history of constipation and rectal bleeding should be evaluate properly. The present experience has shown that in the paediatric age group small bowel obstruction is common but once radiology shows large bowel obstruction, malignancy should be considered in differential diagnosis because early diagnosis and aggressive management lowers the morbidity and mortality in particular if it is a signet ring cell carcinoma of colon and rectum as in indexed case that carries extremely poor prognosis in advanced stage.

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