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.

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 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. All the three criteria were satisfied in our case. In our case clinical presentation of this cancer. The cases with colorectal signet ring cell carcinoma 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].

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 laparatomy 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

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[Table/Fig-1]: Photomicrograph showing tumour cell having intracytoplasmic mucin pushing the nucleus to the periphery and giving a signet ring appearance (H&Ex100)
Radical resection with Palliative Colostomy, Progressive abdominal pain Persistent abdominal pain, 31 years, m

Age-Sex Survival
19 years, m
10 years, m
10 years, m
31 years, m

Hematochezia Polyp at proximal rectum/signet ring cell carcinoma Early Local resection with end to end anastomosis

Radical resection with end to end anastomosis

Rectosigmoid/signet ring cell carcinoma Advanced

Advanced
Advanced
Advanced
Advanced

Rectosigmoid/signet ring cell carcinoma

Transverse colostomy Transverse colostomy Refused treatment Surgical resection

One year 11 month
One year

Advanced

Rectosigmoid, Signet ring cell carcinoma

Right colon resection with ilocolic anastomosis

One year

Abdominal pain & distension, vomiting

Ascending colon/signet ring cell carcinoma

COnCLuSiOn
An 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.

REFERENCES


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