Childhood colorectal carcinoma: A case series

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ABSTRACT

Colorectal carcinoma (CRC) is rare in children, except for a few sporadic reports there is not much information about it in the literature. It is important for pediatricians and paediatric surgeons to be aware that CRC does occur in children, and it should not be excluded only on the basis of the patient's age. Its rarity in children has meant a poor understanding of its biological nature and the treatment modalities to be followed.

Key words: Childhood, colorectal carcinoma

INTRODUCTION

Colorectal carcinoma (CRC) is a rare occurrence in children. Our knowledge of CRC in children is based on a handful of case series and case reports. CRC does occur in children as in adults, although the prevalence is more in the latter. CRC accounts for 2% of malignancies in adolescents and has been reported in children as young as 9 months of age.[1] The possibility of a CRC should be borne in mind for any child with signs and symptoms of intestinal obstruction, intractable abdominal pain, alteration in bowel habits and gastrointestinal bleeding to refer these suspected cases to a surgeon at an early stage. In this report we present two cases of childhood CRC to discuss their presentation, treatment and prognosis.

CASE REPORTS

Case 1

A 14-year-old boy presented to our surgery emergency department with a history of abdominal pain, failure to pass stools/flatus for 3 days and abdominal distension of one day duration. There was no history of vomiting, altered bowel habits, bleeding per rectum or passage of mucus in stools.

Examination revealed a clinically dehydrated but haemodynamically stable boy of average build and mild pallor. The abdomen was distended, with visible peristalsis and increased bowel sounds. Digital rectal examination did not reveal any significant finding. He was diagnosed as a case of acute small intestinal obstruction and was planned for exploratory laparotomy.

Blood chemistry revealed dyselectrolytemia. He was prepared and had emergency lapatotomy. At surgery, the small intestine and colon were distended. There was a large hard mass in the recto-sigmoid junction which was adherent to the presacral fascia and the anterior surface of the sacrum. The loops of the terminal ileum were firmly adherent to this growth.

Due to inadequate information about preoperative staging, unsure nature of tissue planes with vital structures (ureters, iliac vessels), and suboptimal general condition, we took incisional biopsy from the tumour and fashioned a loop ileostomy for fecal diversion. The postoperative period was uneventful.

The histopathological report confirmed mucinous adenocarcinoma. In the postoperative period, staging investigations and optimization for major resection was performed. A detailed evaluation was done, including an abdominal CT scan which revealed the presence of a growth in the recto-sigmoid junction, no lymphadenopathy, no hepatic metastasis but there was a loculated collection in the pelvis and right subphrenic space.

A month after the first surgery, the patient had a re-exploration. The findings included extensive inter-bowel adhesions, growth in the recto-sigmoid junction, malignant infiltration of a terminal loop of ileum, but there was no hepatic metastases. Adhesiolysis, resection of the tumour-bearing recto-sigmoid (followed by stapled colorectal anastomosis) region with segmental resection of the infiltrated loop of ileum was done [Figures 1 and 2]. The ileostomy was not taken down. The histopathological report reconfirmed the presence of the mucinous adenocarcinoma. The one month delay before the second operation was partly due to the need for detailed pathological examination of the specimen; the special stains (NSE, chromogranin etc.) used to specify the growth took sometime.

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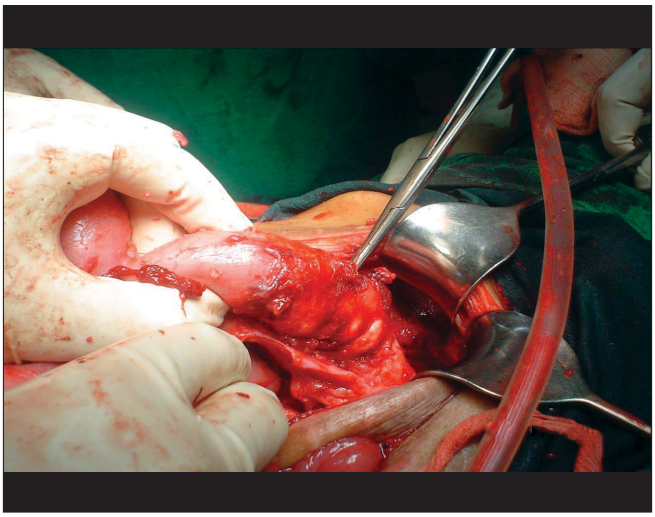


Figure 1: Intra-operative photograph in the first patient showing the growth in the recto-sigmoid junction

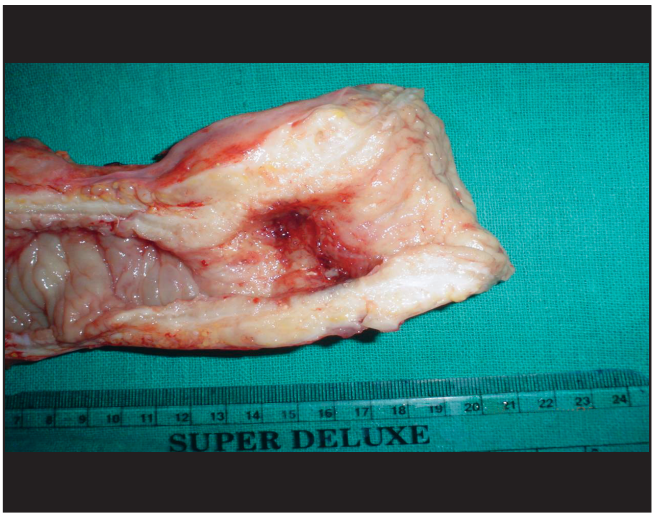


Figure 2: Resected specimen showing the growth

The patient received 6 cycles of chemotherapy (cisplatin, 5fluorouracil, leucoverin, etoposide, adriamycin) in the post operative period. A PET scan done recently did not reveal any viable residual/recurrent disease. The patient is disease-free two years on follow-up.

Case 2

A 14-year-old girl presented to the surgery emergency with complaints of constipation, abdominal distention of 7 days' duration along with several bouts of vomiting for 2 days.

Examination revealed a dehydrated and cachectic girl in shock with severe pallor, bilateral coarse crepitations in the chest and a grossly distended abdomen. No significant finding was elicited on the digital rectal examination.

An erect x-ray of the abdomen revealed multiple air-fluid levels. The patient underwent an emergency laparotomy. The findings were normal small bowel loops, grossly distended large bowel loops, extensive mesenteric lymphadenopathy and a hard stenosing growth in the recto-sigmoid.

Incisional biopsy was taken from the growth and a diverting transverse loop colostomy was performed.

The patient's condition continued to deteriorate in the post operative period despite ventillatory and ionotropic support until she died on the 3rd post-operative day. The histopathological report revealed mucin secreting adenocarcinoma.

DISCUSSION

CRC is generally thought to be a disease of adult and extremely rare in younger age group. CRC has a reported incidence of 1.3 to 2 per million in children;[2,3] unlike the equal sex distribution in adults,[4] CRC affects male children twice as much as in girls. CRC in the young population appears to be more aggressive, to present with later stage, and to have poorer pathologic findings.[5] The poor prognosis in this report can be attributed to delay in diagnosis, advanced stage of the disease at presentation and poorly differentiated histology.[6]

Most children with CRC present in an advanced stage with evidence of metastatic disease, either as gross tumour or as microscopic deposits in lymph nodes, on the surface of the bowel or on intra abdominal organs.[7] Peritoneum is the most common site of extensive intra abdominal disease followed by the greater omentum and the liver.[8]

Mucinous CRC occurs approximately four times as many other tumours in the younger age group;[9] both patients in this report had mucinous CRC. The strikingly higher frequency of mucinous histology suggests that the biology of CRC differs in paediatric from adult patients.[10]

Review of the literature shows contradictory data regarding location of the childhood colorectal cancer. Whereas they are evenly distributed in some reports,[10] they affect entirely the left colon (recto-sigmoid region is the most

common site for the primary tumour) in others,[8,11] and in others still the right side (and the transverse colon) is mostly involved.[12,13] In both our cases, the tumours were located in the recto-sigmoid region.

As a result of its rarity in children and the lack of prospective paediatric studies, recommendations for therapy are primarily extrapolated from adult clinical trials and advocated therapeutic recommendations are the same as for adults.[14]

Curative surgical excision is the primary treatment of childhood colorectal cancer.[1] However, in most cases this is not possible due to advanced disease at presentation.

The role of palliative chemotherapy is very controversial. Active agents in adults with CRC include fluorouracil, folinic acid (leucovorin), oxaliplatin, and irinotecan. Furthermore, newer targeted therapeutic agents, such as bevacizumab and cetuximab, have added additional efficacy to the standard chemotherapy.[1]

This report emphasizes that colorectal malignancy should not be excluded only on the basis of the patient's age; health care providers should have a heightened awareness for this disease condition, particularly because excellent modalities exist to diagnose and treat colorectal cancer. Survival rates for young patients with colorectal cancer are comparable to those of older patients, when matched for Dukes stages.[9]

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