

Mucinous Adenocarcinoma Colon in a 14 Year Old Boy-A Rare Case Report

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Abstract: Colorectal region is the most common site of gastrointestinal malignancy and represents about 15% of all cancer-related deaths. The incidence peaks around seventh decade of life and less than 20% presents before the age of 50. But colorectal carcinoma in children is very rare, with an incidence of only 1.3 per million.^[1] Because of this rarity, clinical management and treatment strategies are generally extrapolated from experience with adults.^[8] However, many small series and case reports suggest that children are more likely than adults to have an advanced-stage disease at presentation, unfavourable (mucinous) tumour histology, and a poor outcome.^[9]

A boy was admitted with acute intestinal obstruction and underwent laparotomy and which showed that patient had colonic mass and was proven to have adenocarcinoma colon on histopathology and was planned for chemotherapy.

Key Words: Acute intestinal obstruction, children, mucinous adenocarcinoma colon

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I. Introduction

Colorectal region is the most common site of gastrointestinal malignancy and represents about 15% of all cancer-related deaths. The incidence peaks around seventh decade of life and less than 20% presents before the age of 50. But colorectal carcinoma in children is very rare, with an incidence of only 1.3 per million.^[1] Limited numbers of colorectal carcinoma in children have been reported, mostly presenting in teens, though, the youngest one reported so far was a 9-month-old baby^[2]

After primary liver tumours, colorectal carcinoma is the most common primary gastrointestinal system malignancies in children and adolescents, and constitute approximately 1% of paediatric neoplasms^[3,4]. Although predisposing factors have been associated with the disease, it usually occurs sporadically^[5,6,7].

A recent study of the Italian TREP project on rare paediatric tumours (A national comprehensive project on Rare Tumours in Pediatric Age (the TREP project) was launched in 2000^[3], using data from the Italian network of cancer registries, estimated a colorectal cancer incidence rate of 0.09 and 0.72 per million person-years for children ages 10 to 14 years and 15 to 17 years, respectively, but recorded no cases in children aged <10 years.⁴ In the TREP project, only 6 cases (all aged >10 years) were registered between 2000 and 2006, out of 336 patients with rare paediatric tumours.

Because of this rarity, clinical management and treatment strategies are generally extrapolated from experience with adults.^[8] However, many small series and case reports suggest that children are more likely than adults to have an advanced-stage disease at presentation, unfavourable (mucinous) tumour histology, and a poor outcome.^[9]

The purpose of this report is to further enhance the clinician's body of knowledge about this disease as well as compare and contrast the paediatric forms of this disease to the adult forms. Paediatricians and paediatric imagers, alike, should become familiar with this rare tumour, its presentation, and imaging features to enable early diagnosis and treatment.

II. Case Report

On 27th February 2017, a 14 year old boy presented with features of acute intestinal obstruction and had a history of vague abdominal pain for the last one month which was relieved temporarily by antispasmodic medications, the patient also gives history of not passing flatus and stool for past 2 days and severe colicky type of abdomen in right iliac fossa and right lumbar regions. The routine haematological investigations were within normal limits. The chest x ray was normal but the X-ray abdomen FPA showed dilated bowel loops in the left hypochondrium and left lumbar regions.



Figure 2 x-ray abdomen FPA air fluid levels

The boy did not improve on conservative management and had to undergo an exploratory laparotomy, on exploration a large growth in ascending colon involving the hepatic flexure of colon is noted causing the

complete obstruction of lumen and dilation of ascending colon, caecum and appendix. There is also loss of tone of IC junction with backward flow of contents into the small bowel. Multiple enlarged mesenteric lymph nodes were noted.

Right hemi-colectomy was performed including the removal of caecum,



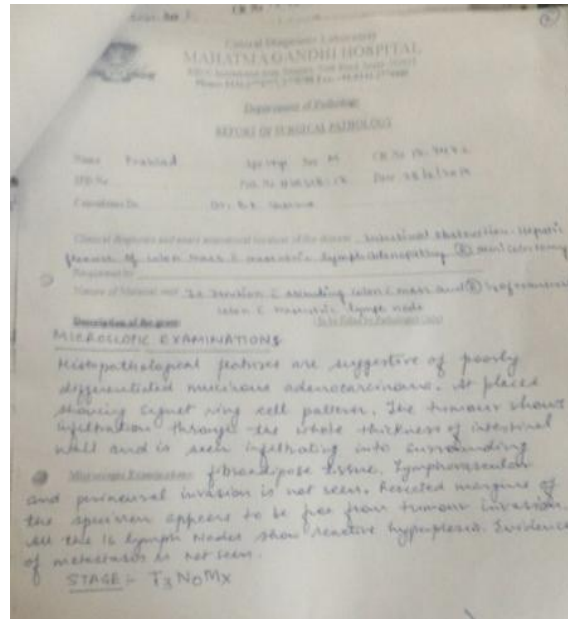
Figure 2 intraoperative picture



Figure 3 intraoperative picture after resection

appendix, and terminal 15 cm of terminal ileum.

The resected specimen consisted of 15cm of terminal ileum, appendix, caecum, ascending colon and proliferative circumferential growth (grey white, hard in consistency) at hepatic flexure, 5 cm of transverse colon and mesenteric lymph nodes which are 16 in number.



Histopathological examination is suggestive of poorly differentiated mucinous adenocarcinoma and at places showing signet ring cell pattern. The tumour shows infiltration of whole thickness of intestinal wall and also into surrounding fibroadipose tissue. Lympho-vascular and perineural infiltration is not seen. Resected margins of the specimen are free from tumour invasion. All the 16 lymphnodes resected showed reactive hyperplasia. Evidence of metastasis is not seen (STAGE:T₃N₀X₀). No family history of colorectal carcinoma in first or second degree relative was found. The patient is planned to undergo chemotherapy with 5FU/leucovorin and oxplatinin (FOLFOX)



Figure 3 gross specimen showing mass lesion of colon

III. Discussion

Colon cancer is very rare in paediatric population. Paediatric oncologists are often unfamiliar with the management of patients with adult-type cancers because they rarely or never encounter them in their daily activities. They generally include carcinoma of the gastrointestinal tract, cutaneous melanoma, renal cell carcinoma and others, in the heterogeneous assortment of “rare paediatric tumours”, orphan diseases for which the cooperative paediatric oncology groups have made few efforts to develop clinical studies in the past. A national comprehensive project on Rare Tumours in Paediatric Age (the TREP project) was launched in 2000,

aiming to collect clinical data, develop diagnostic and therapeutic recommendations for each rare tumor, establish co-operative networks with other specialists involved in the management of these tumors (i.e. adult oncologists, surgeons), and organize pathological and biological studies^[8]. Colonic carcinoma in children has no predilection for gender^[10]. This is in contrast to adults where there is a 3:2 preponderance in men^[11]. The reported peak incidence is approximately 15 years of age^[12]. Another series reported the median age to be 15 years of age.^[13] Hereditary non-polyposis colon cancer and other familial syndromes account for a few, but most childhood cases appear sporadically. Predisposing factors like familial polyposis of colon, other polyposis syndromes, ulcerative colitis, familial multiple cancer syndromes are noted in about 10% of colorectal cancers in paediatric population, which is far higher than in adults.^[14] Salas-Valverde *et al.*,^[14] reported presence of predisposing factors in three out of 11 cases but in many others, no predisposing factor can be found.^[15,6] The present case was also a sporadic one. In cases with predisposing factors also, carcinoma usually develops in adulthood. In paediatric population colorectal carcinoma shows male preponderance (M: F= 2:1).^[15] Presenting symptoms of colorectal carcinoma in children vary from bloody stools, pain of abdomen, and altered bowel pattern to unexplained weight loss. Since all the symptoms and signs are vague, early diagnosis is often missed and the disease presents at an advanced stage, often with intestinal obstruction. In the present case, the child presented with acute intestinal obstruction. Because of limited number of cases being reported in the literature, there is controversy about tumour distribution in the colon in children. In the present case, the growth was situated at the hepatic flexure of colon. In adults, colorectal adenocarcinoma usually shows tubular differentiation. In mucinous carcinoma, lakes of extracellular mucin are formed in which relatively bland looking tumour cells float. The mucin absorbs water, swells and invades tissues, thereby promoting spread of malignant cells. This tumour may grow to large size because of the pooled mucin. It interferes with immune recognition of carcinoma cells caused by mucopolysaccharide coating.^[17] Signet ring cell carcinoma causes early metastasis to lymph nodes, peritoneal surfaces and ovary, rather than liver and has extremely poor prognosis. This variant accounts for only 1% of primary colon carcinoma in adults. In the present case, histology showed predominating signet ring histology. In children with colon carcinoma, the histologic type found in more than 50% is an aggressive, mucin producing, poorly differentiated adenocarcinoma, which is found in only 5% to 8% of adults.^[18] In addition, the low index of suspicion for this tumor in children results in an advanced disease at diagnosis and subsequently a poor prognosis.^[19] Rao *et al.*^[20] reported in 1985 that 67% of colon carcinoma in children presented as Duke stage D, 23% as stage C, and only 10% as stage B. The prognosis in children with carcinoma of the colon is therefore worse than in older patients with a 51% versus 75% overall 5-year survival.^[18] Because these tumours are found in an advanced stage in children at the time of diagnosis, they are often not resectable. The tumour may spread throughout the peritoneal cavity to involve the omentum, peritoneum, mesenteric lymph nodes, liver, and ovaries and they may spread through the bloodstream to the lungs and eventually the brain, bones, or both.^[21] The present case was at stage B of Duke's staging. All the studies published so far, described a poor outcome, for childhood colorectal carcinoma. The five-year-survival rate for children ranges from 7% to 12%, and is similar for both developed and developing countries.^[5] Advanced stage at presentation and mucinous/signet ring cell histology contribute to this outcome. Among all prognostic parameters, tumour stage, lymph node metastasis and vascular and perineural invasions are most important. All of them are usually present when the histology is a signet ring type, as in the present case, making the prognosis extremely poor. Because the disease remains unsuspected, presentation is delayed with up to 60% having luminal obstruction as opposed to 18% in adults. At operation, complete resection is possible in less than 40% of cases as against 90% possible resection in adults.^[5] This is especially true for the signet-ring subtype, which grows so rapidly that regional lymph node involvement and diffuse peritoneal seeding are common findings at presentation.

The management of colorectal cancer in children is similar to adults. Complete surgical excision is the most important prognostic factor and the primary aim of the surgeon, but in most instances this is not possible. Most patients with microscopic metastatic disease usually develop gross metastatic disease and few individuals with metastatic disease at diagnosis become long-term survivors.

IV. Conclusion

Despite its aggressive nature, colorectal carcinoma in children need not be a hopeless illness; cure with long-term survival is possible. Early diagnosis and a

Multidisciplinary therapeutic approach are important to improve prognosis. Prognosis in colorectal cancer is usually determined by the degree of intestinal wall invasion, lymph node involvement, and haematogenous metastases. For this reason, early detection increases the chance of cure. Enhanced clinician awareness of the possibility of colon cancer in youngsters, especially in those with a known genetic predisposition or a positive family history, is a key factor.

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