COLORECTAL CARCINOMA IN AN 11-YEAR-OLD FEMALE: A CASE REPORT

Akpa PO (MBBS, FMCpath)[1,2*], Abaniwo SA (MBBS)[1], Richard KS (MBBS)[1], Shitta AH (MBBS, FWACS)[3], Jatau IA (MBBS)[3], Kwaghe BV (MBBS, FMCpath)[1]

[1]Department of Histopathology, Jos University Teaching Hospital, P.M.B 2076, Jos Plateau State, Nigeria.
[2]Department of Histopathology, University of Jos, P.M.B 2084, Jos Plateau State Nigeria.
[3]Department of Surgery, Jos University Teaching Hospital, P.M.B 2076, Jos Plateau State, Nigeria.

Correspondence: *Akpa Philip Ojile, Department of Histopathology, Jos University Teaching Hospital, Jos plateau state Nigeria. P.M.B 2076. Email:akpaphilip@yahoo.com. Phone: +2347035128504*

ABSTRACT

Introduction: Colorectal carcinoma is a leading cancer and cause of mortality in the adult population but occurs rarely in the pediatric population. The incidence of this tumour in the pediatric population is increasing worldwide. Diagnosis is often delayed in children, with majority of their tumours being poorly differentiated. The purpose of this case report is to document the rare occurrence of this tumour in a very young child and to increase its awareness in practicing doctors.

The case: We present a case of an 11-year-old female child with a 2 month history of recurrent constipation, 3-day history of abdominal distension and pain and a 2-day history of bilious projectile vomiting. An abdominal ultrasound scan detected distended loops of bowel with an assessment of intestinal obstruction of unknown cause. Surgery was a mid-line laparotomy with segmental bowel resection of splenic flexure tumour and divided colostomy. Pathological examination revealed a 15cm length of large bowel segment with a circumferential constricting tumour in the mid portion of its length. Histological examination revealed a mucinous adenocarcinoma. Patient received adjuvant chemotherapy, had reversal of colostomy and is doing well 5 months postoperatively with no recurrence.

Conclusion: Pediatric colorectal carcinoma cases are challenging due to their tendency to be misdiagnosed in addition to other bad prognostic factors often encountered. A complete assessment is necessary in all patients who present with clinical features suggestive of this disease regardless of age.

Keywords: Colorectal carcinoma, Case report, pediatrics, plateau state, child, mucinous, Jos, Nigeria

INTRODUCTION

Colorectal carcinoma is amongst the leading cancers in adults, but is rare in the pediatric population.[1-5] It accounts for about 9.4% of cancer diagnosis and 7.9% of total cancer deaths world-wide, however only an estimated 1% of colorectal carcinomas occur in patients less than 30 years.[2,4] Familial adenomatous polyposis and lynch syndrome are associated with a substantially higher percentage of colorectal carcinomas in children than in adults.[1,4,6] Its diagnosis is not usually suspected in the pediatric population even when they manifest symptoms that will immediately raise suspicion in an adult, hence diagnosis is often delayed.[1,3]Colorectal malignancies have a poorer prognosis in children when compared to that of adults owing to late diagnosis and more aggressive tumours diagnosed in this age group.[1,4,5]

There is a paucity of published detailed literature on pediatric colorectal carcinoma in Nigeria. Musa et al reported a case of rectal carcinoma in a 9-year-old male in 2007 at a tertiary health care center in south-western Nigeria. The patient had a six-month history of abdominal pain, recurrent distension, constipation, bloody stool and progressive weight loss with a positive family history of colorectal carcinoma.[7] The diagnosis was missed on four visits to different private health facilities and was finally made on presentation to the tertiary health facility, at this point the bowel was obstructed and the tumour metastatic. The patient had laparotomy, with resection of the

tumour which was a poorly differentiated adenocarcinoma on pathological examination. Post-operative recovery was poor due to poor pre-operative state and advanced disease. The patient died 35 days' post-surgery.[7] In a 30-year (1979-2008) review by Ibrahim et al at the University of Ilorin Teaching Hospital in north-central Nigeria, 31.5% of their colorectal carcinoma cases occurred in young patients (less than 40 years). The mean age at presentation was 31 years with the youngest patient being 16 years.[8] It occurred slightly more frequently in men, a majority occurred in the rectum and most cases presented in advanced stages with poor outcome.[8] We are presenting this case because of the rarity of this tumour in the index patients age group in addition to raising awareness and index of suspicion when assessing pediatric patients with suspicious gastro-intestinal symptoms.

THE CASE

History and symptoms

An 11-year-old female patient who presented with a 2-month history of recurrent constipation, a 3-day history of progressively increasing abdominal swelling and pain, with an associated 2-day history of bilious projectile vomiting. There was no associated hematochezia and melena. No family history of colorectal malignancies.

Pre-operation findings

On examination the patient was pale and dehydrated with a pulse rate of 100 beats per min, respiratory rate of 30 cycles per min and temperature of 36.5^{o}C. Abdomen was distended and moved with respiration, the abdominal girth was 64cm, at 12cm from the xiphisternum. There were observable peristaltic movements and borborygmi. The hernia orifices were intact, there were no palpable intra-abdominal organs or masses. Percussion notes were tympanitic, bowel sounds were hyperactive, with partially distended floating bowel loops. Digital rectal examination revealed an empty rectum, the procedure was tender and the gloved finger was stained with blood. There was no ascites.

Investigations done

Serum urea, creatinine and Electrolytes were within normal range, packed cell volume was 32%. **Abdominal ultrasound scan** revealed floating gaseously distended loops of bowel demonstrating to and fro peristalsis. The liver, spleen, gall bladder, pancreas, kidneys and urinary bladder appeared normal on ultrasound scan, there was no ascites. An assessment of Intestinal obstruction of unknown cause was made.

Intraoperative findings

Operation was performed after resuscitation and obtaining an informed consent from the patient's parents. Abdominal cavity was approached via a midline laparotomy incision. There were distended loops of small bowel and a splenic flexure nodule. A 15cm segment of intestine was resected. The resected segment extended from the distal transverse colon to the proximal descending colon with unaffected intestine flanking the tumour on both sides. A single enlarged lymph node was identified in the attached mesentery and there was no ascites. A Left sided divided colostomy was applied and the abdominal cavity was closed in layers.

Pathology of specimen

On gross examination the specimen consisted of a segment of intestine that measured 15cm in length, with its mesenteric attachment and a single enlarged mesenteric lymph node. Cut open along its anti-mesenteric border revealed a circumferential constricting mass midway along the length of the bowel that narrows and completely occludes the segment of intestine. The constricted segment of bowel measured 3 cm in length and 1.5cm in wall thickness. The mucosa at the constricted portion was nodular while the mucosa adjacent to the mass on both sides appear roughened and hyperemic. Samples were taken from the constricted portion (mass), the enlarged lymph node, proximal resection margin and distal resection margin.

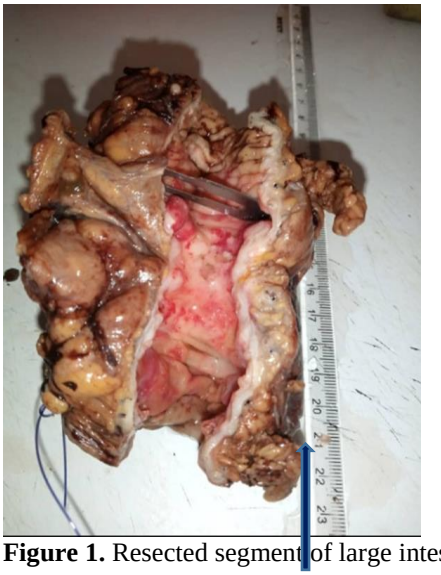

Figure 1. Resected segment of large intestine with a circumferential constricting mass (arrow) along its length.

Microscopy

Histological sections of the colonic mass showed infiltration of all layers of the intestinal wall by neoplastic epithelial cells. These neoplastic epithelial cells are a mixed population of moderately differentiated neoplastic acini and epithelial cell floating in pools of mucin. The epithelial cells lining the malignant glands exhibit nuclear pleomorphism and hyperchromasia, increased nucleo-cytoplasmic ratio and loss of polarity. The epithelial cells in the pools of mucin exhibit moderate atypia with some signet ring forms. Resection margins were free of tumour infiltration and lymph node showed sinus histiocytosis with no neoplastic cell infiltration. A diagnosis of **Mucinous adenocarcinoma** (pT3,N0, MX) (Dukes stage B) was made.

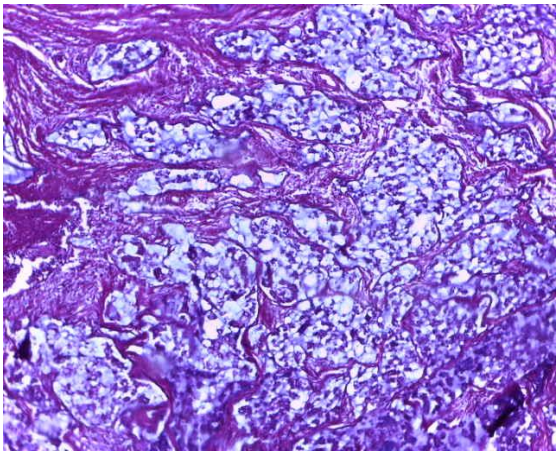

Figure 2: Photomicrograph (hematoxylin and eosin x 100 magnification) showing malignant epithelial cells floating in pools of mucin and dissecting through the intestinal wall

DISCUSSION

Colorectal carcinoma is a common tumour in the adult population peaking at about the age of 65 years, with a life time risk of five to six percent of developing this tumour.[1-6] Conversely it is a rare cancer in the pediatric population with an incidence of 1-2 cases per a million of population per year.[1-3,5] This tumour rarely occurs before the age of 20 years and accounts for less than 1% of neoplasia in the pediatric

population.[1,3] The occurrence of colorectal carcinoma appears to be an absolute exception in the pre-pubertal age group with the incidence dropping drastically in patients below the age of 10 years. [2,4] The incidence of colorectal carcinoma in the developed countries of Europe and north America is higher than in underdeveloped countries of Africa and Asia.[3] The incidence in the young appears to be slowly growing worldwide.[9] A familial predisposition does not commonly lead to an increased risk of its occurrence in individuals before the age of 20 years.[1] The common age range of diagnosis of colorectal carcinoma when it occurs in children is in the second decade, commonly between 15-19 years.[4,5] Colorectal carcinoma in the pediatric age group shows a male predominance with a male to female ratio of 2:1.[4,5,6]

The index case is the youngest documented case of diagnosed colorectal carcinoma at the Jos University Teaching Hospital (JUTH) since reliable data documentation began in the 1980s. Previous reports from this institution show that colorectal carcinoma occurs relatively commonly in young patients, but none was as young as the index case. In a 1999 publication by Sule et al at the Jos University Teaching Hospital, 35 (23.6%) out of 149 cases of colorectal carcinoma occurred in patients 30 years and below, the mean age at diagnosis was 25(STD+/-6) years with a male to female ratio of 1.2:1. The rectum was the most common site and majority presented in advanced stages with poor outcome.[10]

The risk of developing colorectal carcinoma is higher in populations that have adopted the so-called western lifestyle which entails the consumption of alcohol, red meat and low fiber foods in association with a sedentary lifestyle, being obese and smoking cigarettes.[11,12]A majority of colorectal carcinomas occur sporadically, with the rest occurring in association with genetic susceptibility syndromes.[9] The percentage of colorectal carcinomas occurring in the setting of genetic susceptibility, ranges from between 5-30% of pediatric colorectal carcinoma cases, this percentage is higher than that seen in adults.[1,4,5,6,9] The genetic syndromes associated with colorectal carcinoma include hereditary non-polyposis colon cancer, familial adenomatous polyposis, hereditary mixed polyposis syndrome, Peutz-Jeghers syndrome and familial juvenile polyposis.[1,3-6] The Bethesda guidelines state that patients diagnosed with colorectal carcinoma before the age of 50 should have genetic testing for susceptibility syndromes.[9] The index case has no known family history of colorectal carcinoma and genetic tests were not carried out to test for susceptibility syndromes.

The clinical sign and symptoms of colorectal carcinoma are similar in children and adults.[1,3] These clinical manifestations can however be non-specific and easily lead to misdiagnosis in pediatric patients.[1-3,6,9] Features such as change in bowel habits, bleeding per rectum, abdominal pain, anemia and weight loss can be mistaken for features of irritable bowel syndrome, inflammatory bowel disease, hemorrhoids, gastroenteritis and eating disorders.[1,9] The presenting symptoms of the index case would immediately raise the suspicion of colorectal carcinoma in an adult but the diagnosis wasn't suspected until during surgery and later confirmed on histopathological examination.

Colorectal carcinomas occur more commonly in the proximal colon (ascending and transverse colon) in children while majority of adult tumours occur in the distal colon within 25cm of the anus.[4,5] Published literature on colorectal carcinoma in young patients in Nigeria have however showed a predominance of rectal tumours.[8,10] The index case occurred at the splenic flexure which is relatively proximal and in keeping with the site of occurrence in children. A majority of colorectal carcinomas in adults are moderately or well differentiated adenocarcinomas, in contrast a majority of child-hood colorectal adenocarcinomas are poorly differentiated.[1,3,5] Mucinous and signet ring adenocarcinomas occur twice more frequently in children than in adults.[1] While mucinous tumours make up only 5% of adult colorectal carcinomas, it accounts for greater than 50% of the pediatric colorectal carcinomas.[3,5,6] The index case is a mucinous adenocarcinoma which is a poorly differentiated tumour. The detection of unique microsatellite instabilities in addition to the detection of more aggressive and less chemo-sensitive colorectal tumours in children suggests there may be

differences in the pathogenesis of these tumours compared to their counterparts in adults.[1,2]

The duration from onset of symptoms to diagnosis of colorectal carcinomas in children is usually long and ranges from 2 to 6 months.[1,2] Children and adolescents present with more advanced disease than their adult counterparts with about 60-80% of pediatric cases diagnosed in Dukes Stages C and D. [2-6] Vague symptoms, decreased awareness, and increased frequency of poorly differentiated types result in this advanced stages at presentation.[4,6] The index case was diagnosed in Dukes stage B which is relatively early with hope of long term survival or possible cure.

Complete surgical resection when possible is the most effective form of treatment and the only hope for cure or long term survival in pediatric colorectal carcinoma cases.[3,4] Most cases are however diagnosed late and no longer surgically resectable with locally advanced disease or metastasis.[1] In advanced disease adjuvant multi-agent chemotherapy based on a fluorouracil backbone with folinic acid, oxaliplatin, or irinotecan is commonly used.[3,4,5] The index case was completely surgically removed with tumour free resection margins. The single enlarged lymph node seen was also free of tumour cells. Patient received adjuvant chemotherapy using FOLFOX-4 regimen (oxiplatin, leucovorin and 5-fluorouracil). Colostomy was successfully reversed and patient is currently doing well 5 months' post-surgery.

The 5 year survival rate for colorectal carcinoma in children is relatively low when compared to adult survival rates, 5 year survival ranges from 5 to 28%.[1] Factors responsible for the poor outcomes include delay in diagnosis resulting in late stage disease, poor differentiation of the tumours and also limited experience of pediatric oncologists and surgeons.[2-5] A histological finding of mucinous histology, signet ring cell rate of greater than 10% and incomplete surgical resection are poor prognostic signs.[1] At presentation 60% of pediatric patients have luminal obstruction as opposed to 18% seen in adults, at surgery less than 40% of pediatric tumours can be completely resected as against 90% in adults. [6] The survival rates of pediatric colorectal carcinoma patients do not differ significantly between developed and developing countries.[4,5] The index case was detected in Dukes Stage B with a fair chance of long term survival, however the mucinous adenocarcinoma morphology is an indication for more aggressive treatment and patient monitoring. The carcino-embryonic antigen (CEA) measured at 1-month post-surgery was 2.32ng/ml which within the normal range and a good prognostic sign.

CONCLUSION

The management of pediatric colorectal carcinoma is challenging owing to its tendency to present late, the occurrence of aggressive tumours and relatively less experience among pediatric surgeons and oncologist in management of this disease. The index of suspicion should therefore be raised in assessing pediatric patients with suspicious gastro-intestinal symptoms in order to make early diagnosis and improve survival.

CONSENT

Is not applicable (no patient identifiers)

ETHICAL APPROVAL

Not applicable

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COMPETING INTERESTS

Authors of this article declare that there are no competing interests

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