brief report

Adenocarcinoma of the colon in children:
Case series and mini-review of the literature

Youssef Al-Tonbary *, Ahmad Darwish, Ahmad El-Hussein, Ashraf Fouda

Hematology/Oncology Unit, Mansoura University Children Hospital, Mansoura, Egypt

* Correspondence. ytonbary@gmail.com · Accepted for publication 24 November 2012

Colon cancer is extremely rare in children. This article reports three cases of adenocarcinoma of the colon. A 12-year-old boy, a 13-year-old boy, and a 13-year-old girl presented with constipation and abdominal enlargement over a two-month duration. Abdominal ultrasound and barium enema confirmed a stenotic segment at the rectum with obvious shouldering. Adenocarcinoma was diagnosed following colonoscopic biopsy and laparotomy. We conclude that any child presenting with unexplained persistent constipation, abdominal distension or bleeding per rectum, colon cancer should be suspected and investigated with endoscopy or barium enema.

Primary gastrointestinal system malignancies constitute approximately 2% of pediatric neoplasm and, of these, colorectal carcinoma (CRC) is the second most common malignancy after primary liver tumors. Therefore, CRC remains unsuspected in children and most patients (60–80%) are in advanced stage at presentation. The overall prognosis is poor because of the delay in diagnosis, advanced stage of disease and lack of histological differentiation.

CASE 1

A 12-year-old boy complained of recurrent abdominal pain and constipation over a period of two months. There was no past history or family history of gastrointestinal problems. Examination revealed marked abdominal distension with no organomegaly. Per rectum (PR) examination revealed a large circumferential lesion obstructing the lumen, 3 cm from anal verge with limited mobility.

Abdominal ultrasound showed a long stenotic segment at the rectum with obvious shouldering at the proximal and distal ends measuring 5 cm in length and 1.6 cm in thickness, which is suggestive of a rectal tumor. The colon proximal to the lesion was markedly dilated. Barium enema confirmed shouldering and a granular mucosal pattern (Figure 1).

Colonoscopy revealed a large stenotic mass starting at 4 cm from anal verge for the long segment affecting the entire circumference of the rectum with marked luminal stenosis. Endoscope failed to pass. Multiple biopsies were taken. Pathological examination of the masses revealed firmness in consistency and a grayish whiteness in color, while microscopic examination showed grouped and scattered malignant epithelial cells with a moderate degree of atypia and pleomorphism within mucin lakes. Some showed attempts at acinar formation which is consistent with mucoid adenocarcinoma (Figure 2).

Surgical resection of the colon with colostomy was performed. There was no evidence of visceral or nodal spread. The patient received radiotherapy and adjuvant chemotherapy in the form of 5-fluorouracil and folinic acid. The patient had recurrence within six months and died within one year.

CASE 2

A 13-year-old boy presented with a two-month history of insidious onset and progressive course of abdominal enlargement, vomiting, dizziness, weakness, and dyspnea. The condition was associated with unilateral edema of left lower limb followed by severe abdominal pain and anorexia. The patient showed marked cachexia (weight loss and muscle wasting) with pallor. Abdominal examination revealed multiple abdominal masses involving mainly the left hypochondrial region with ascites (positive shifting dullness). PR examination was normal.
Blood tests showed normocytic and normochromic anemia. Serum albumin was 3.4 g/dl and CA-19-9 (Carbohydrate Antigen 19-9) was 1020 U/ml (normal up to 37 U/ml).

Computed tomography (CT) of the abdomen (Figure 3) revealed a heterogeneously enhancing soft tissue mass with calcification inside seen in the left hypochondriac region, circumferentially surrounding the descending colon, measuring about 5 × 6 cm. Evidence of diffuse enhancing peritoneal thickening and nodules, with scalloping of the posterolateral surface of the liver, extending between the urinary bladder and rectum, filling the Douglas pouch, as well as the central mesenteric mass, suggestive of pseudomyxomatosus peritoneal moderate ascites. CT chest revealed left basal pulmonary nodule, mostly metastatic.

A debulking operation and biopsy were performed on the patient and pathology revealed malignant epithelial cells arranged in glandular structures with an area of cribriform pattern and solid sheets, and groups of tumor cells floating in pools of mucin. Figure 4 shows mucoid adenocarcinoma.

The patient received chemotherapy FOLFOX 4 protocol (Oxaliplatin, Folinic Acid and 5-Fluorouracil) for three cycles every three weeks, but died from sepsis three months after diagnosis.

CASE 3

A 13-year-old girl presented with history of bleeding per rectum. The condition was associated with a mass projecting from the anal canal. PR examination revealed a mass in the lower part of the anal canal.

Blood picture showed normocytic and normochromic anemia. Bone marrow aspiration was free.
Computed tomography (CT) of the abdomen revealed a diffuse circumferential soft tissue thickening on the rectosigmoid region with haziness of the surrounding fat and multiple small perirectal lymph nodes contacting the posterior wall of the uterus.

Barium enema was performed revealing narrowing of the anal canal (Figure 5). Lower gastrointestinal endoscopy revealed a large mass (7 cm) from the anal verge with areas of hemorrhage and necrosis. Multiple biopsies were taken.

The patient underwent low anterior resection with converging ileostomy and biopsy. Pathologic examination revealed moderate differentiated adenocarcinoma reaching the serosa. There are infiltrating two out of eleven lymph nodes. TNM staging was at IIIB.

The patient received chemotherapy FOLFOX 4 protocol (Oxaliplatin, Folinic Acid and 5-Fluorouracil) for six cycles every three weeks and is still under follow up.

DISCUSSION

Although CRC is one of the most frequent tumors in adults, it rarely occurs before the age of 20 years, with an annual incidence of only one to two cases per one million people in the US, accounting for only about 80 cases per year. CRC is more common in developed countries. It is more frequent in Europe and North America than in Africa or Asia, with the exception of Japan. Well-defined CRC predisposition syndromes account for only about 3–5% of all cases of colon cancer; they include Peutz-Jeghers syndrome, familial juvenile polyposis, hereditary mixed polyposis syndrome, hereditary non-polyposis colon cancer, and familial adenomatous polyposis.

The vast majority of children reported with CRC were over 10 years of age; however, a few cases were a premature newborn with multiple congenital anomalies and a 9-month-old child reported with isolated CRC.

CRC in children tends to present with similar complaints as in adults, primarily with abdominal pain, but also hematochezia, altered bowel habits, weight loss, and anemia. Children and adolescents also often present with acute abdominal conditions, such as acute obstruction, perforation, or severe pain mimicking appendicitis. In some pediatric reports, acute presentations, including intestinal obstruction and acute pain mimicking appendicitis, account for almost 50% of presentations.

Changes in bowel habits, such as constipation or diarrhea, and in the caliber of stools may be observed before the development of tarry stools, rectal bleeding, or other changes. There may be decreased appetite and weight loss. The presentation of CRC is related to its primary site within the large bowel. Tumors involving the cecum and descending colon may become bulky before symptoms appear. Tumors of the rectum and sigmoid colon may be associated with changes in the caliber of the stool, dyschezia, hematochezia, and anemia. The diagnosis of CRC in young patients is often delayed because it is seldom suspected. Acute bowel symptoms necessitate immediate abdominal exploration, at which time perforation of the large bowel with multiple metastatic deposits may be observed. Intestinal obstruction by tumor occurs more frequently in adolescents than in adults with this cancer.

Once the diagnosis is suspected, evaluation typically includes abdominal X-rays, barium enema, CT, and eventually colonoscopy, which will show either obstruction, narrowing of the colonic lumen, or an abdominal mass. Depending on the situation, either colonoscopy with excision of a polyp or initial biopsy with delayed excision of the mass would be the next step. Biopsy is required for the diagnosis of CRC. A biopsy may be obtained during colonoscopy or laparotomy, at which time definitive surgery may or may not be feasible. Most colorectal cancers in adults are moderately differentiated or well differentiated adenocarcinomas. In contrast, more than half of reported...
Table 1. The previously reported adenocarcinoma in children and adolescents.

<table>
<thead>
<tr>
<th>Reports</th>
<th>Year of publication</th>
<th>Country</th>
<th>No. of cases</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Site of tumor</th>
<th>Pathology</th>
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<tbody>
<tr>
<td>Chana et al.</td>
<td>2001</td>
<td>India</td>
<td>1</td>
<td>12</td>
<td>Male</td>
<td>Rectum</td>
<td>Well-differentiated mucinous adenocarcinoma</td>
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<tr>
<td>Jeong et al.</td>
<td>2008</td>
<td>Korea</td>
<td>1</td>
<td>13</td>
<td>Female</td>
<td>Rectosigmoid</td>
<td>Well-differentiated adenocarcinoma</td>
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<tr>
<td>Ferrari et al.</td>
<td>2008</td>
<td>Italy</td>
<td>7</td>
<td>Median age (12)</td>
<td>4 Males</td>
<td>Right colon (2 cases)</td>
<td>Mucinous adenocarcinoma (4 cases)</td>
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<td></td>
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<td></td>
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<td>Splenic flexure (1 case)</td>
<td>Moderately differentiated adenocarcinoma (2 cases)</td>
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<td></td>
<td></td>
<td>Left colon (1 case)</td>
<td>Poorly differentiated adenocarcinoma with neuroendocrine differentiation (1 case)</td>
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<td>Sigmoid (3 cases)</td>
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<tr>
<td>Sharma et al.</td>
<td>2009</td>
<td>India</td>
<td>2</td>
<td>14</td>
<td>1 Male</td>
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<td>Mucinous adenocarcinoma</td>
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<td>Salas-Valverde et al.</td>
<td>2009</td>
<td>Costa Rica</td>
<td>11</td>
<td>Median age (11.6)</td>
<td>7 Males</td>
<td>Transverse colon (3 cases)</td>
<td>Mucinous adenocarcinoma (7 cases)</td>
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<td></td>
<td>Right colon (2 cases)</td>
<td>Adult form of non-mucinous adenocarcinoma (2 cases)</td>
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<td></td>
<td>Hepatic flexure (1 case)</td>
<td>Moderately differentiated adenocarcinoma (1 case)</td>
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<td>Splenic flexure (1 case)</td>
<td>Poorly differentiated adenocarcinoma (1 case)</td>
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<td>Left colon (1 case)</td>
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<td>Multiple segments (2 cases)</td>
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<td>Malik and Kamath</td>
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<td>India</td>
<td>1</td>
<td>11</td>
<td>Male</td>
<td>Rectum</td>
<td>Invasive poorly differentiated signet ring carcinoma grade 3</td>
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<td>Agrawal et al.</td>
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<td>India</td>
<td>1</td>
<td>11</td>
<td>Male</td>
<td>Rectosigmoid</td>
<td>Mucinous adenocarcinoma</td>
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<td>Ibrahim et al.</td>
<td>2011</td>
<td>Nigeria</td>
<td>2</td>
<td>16 and 18</td>
<td>2 Males</td>
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cases of childhood CRC are poorly differentiated mucinous adenocarcinoma, and many are of the sigmoid type.\textsuperscript{1,13} The differential diagnosis includes malignant carcinoid tumor, leiomyosarcoma, malignant fibrous histiocytoma, and metastatic tumor from other sites. All may have similar presentations; metastases may be identified only by histology or metastatic site.\textsuperscript{7}

Following adult principles of treatment, the mainstay of therapy is complete surgical resection. Without complete surgical resection, cure is not possible. Adjuvant multi-agent chemotherapy based on a fluorouracil backbone with folinic acid (leucovorin), oxaliplatin, or irinotecan, and possibly bevacizumab or cetuximab, is typically used in advanced stage disease, as well as in high-risk localized disease. There is little role for radiation therapy other than for palliation, except in rectal carcinoma. In the absence of pediatric prospective clinical trials, treatment guidelines must be extrapolated from adult trials.\textsuperscript{9}

Table 1 shows the previously reported adenocarcinoma in children and adolescents.\textsuperscript{14–23}

### CONCLUSION

Three patients over 10 years of age presented with persistent constipation and abdominal distension. The patients had a two-month history of symptoms, and were in advanced stage of the disease. The outcome of their treatment was poor. CRC, although rare, should be suspected and investigated in any child presenting with unexplained persistent constipation, abdominal distension or bleeding per rectum.

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### REFERENCES