An unusual presentation of carcinoma of the colon in a child

A case report

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Summary

A patient with carcinoma of the large bowel who presented with a subphrenic abscess is reported. This case emphasizes two important facts relating to colonic cancer in childhood: (i) premalignant disease of the large bowel is no prerequisite for the development of colonic cancer; and (ii) in childhood this disease is characterized by a fulminating course and high mortality. In about 50% of cases the tumour is of the signet ring or mucin-producing type, which explains the grave prognosis.

Although colonic cancer is rarely encountered in children, in 1880 Ahlfield published a report of carcinoma of the large bowel in a premature infant. Because of its paucity in children under 12 years of age, carcinoma of the large bowel is rarely considered in the differential diagnosis if a child presents with right upper quadrant tenderness and a mass. To illustrate this point we report the case of a 12-year-old Coloured child with carcinoma of the colon who presented, most unusually, with a subphrenic abscess. Apart from being a rare manifestation of colonic cancer, this case emphasizes: (i) the insidious onset of the disease; (ii) that colonic carcinoma can develop in a child without previous disease of the bowel; and (iii) that the short duration of symptoms does not rule out the presence of disseminated cancer of the large intestine in a child.

Case report

A 12-year-old Coloured child was referred to our Paediatric Surgery Clinic with a 2-week history of upper abdominal pain. On examination he was found to be pyrexial (39°C) with right upper quadrant tenderness and a mass, signs which the referring doctor thought to be suggestive of an amoebic abscess of the liver.

On examination he was cachectic with obvious weight loss, anaemia and visible fullness in the right upper quadrant but no cyanosis, jaundice or lymphadenopathy. The results of systemic examination were within normal limits, but the haemoglobin concentration was 7,5 g/dl and the leucocyte count 16,4 x 10^9/l with 2 - 4 pus cells/high-power field with no haematuria and no sedimentation rate remained unexplained, and a count was as follows: neutrophils 80%, lymphocytes 19% and monocytes 1%. The erythrocyte sedimentation rate was 116 mm/h and the patient's blood group was AB+.

With gentle palpation it was possible to determine the extent of tenderness and to delineate the border of the mass; the mass did not move upon inspiration, was dull when percussed and was confined to the liver and gallbladder area, while the diaphragm was found to be elevated by about 3 cm on the right.

Although bowel sounds were infrequent they were of normal intensity and no obstructive peristaltic waves could be elicited; auscultation of the mass was non-contributory. While in hospital the patient vomited some clear, semidigested food. Intravenous pyelography showed that both kidneys were normal in size and that urine excretion was satisfactory.

Ultrasonography of the mass clearly indicated that the diaphragm was elevated and that there was a large space between the liver and the diaphragm, this being suggestive of a subphrenic abscess. It was decided to resort to laparotomy and drainage of the abscess. A right subcostal incision was made and extended along the inner wall of the thoracic cage; a finger was inserted into a very large subphrenic space, from which a large quantity (± 500 ml) of malodorous pus was evacuated. A pus swab was taken for culture and sensitivity testing of the infecting organism. The subdiaphragmatic space was irrigated with normal saline and necrotic tissue and fibrin plaques were evacuated. A corrugated drain was inserted and the abdomen was closed in layers. The infrahepatic space could not be inspected at this stage because chronic infection had obliterat adw with omentum and granulation tissue.

Postoperatively, improvement in the patient's condition was dramatic. His temperature, pulse rate and breathing returned to normal within 24 hours; however, his appetite remained poor, his weight increase was disappointing (notwithstanding intravenous hyperalimentation and a high-energy, high-protein diet) and the erythrocyte sedimentation rate remained alarmingly high (75 - 85 mm/h).

The origin of the subphrenic abscess and persistently elevated erythrocyte sedimentation rate remained unexplained, and a careful search was embarked upon. A barium study of the small bowel to exclude tuberculosis demonstrated a fistula in the right colon through which barium flowed freely into the subphrenic space and out onto the skin (Fig. 1).

The rest of the large bowel was normal. In the light of these findings tuberculosis or foreign body perforation were put forward as tentative diagnoses. Drainage through the fistula/perforation increased, and it was decided to resect the affected segment of bowel after bowel sterilization.

At laparotomy it was found that the inflammatory response had improved, but enlarged lymph nodes were visible in the subhepatic region and in the distribution of the right and middle colic artery, these being suggestive of metastatic dissemination from the tissue encountered at the site of the fistula. Histological examination of biopsy specimens confirmed this suspicion and showed that the tumour consisted of poorly differentiated adenocarcinoma of the colon. Clostridium perfringens, Klebsiella pneumoniae and Escherichia coli were cultured from the subphrenic abscess.

The histological nature of the tumour and the extent to which it had disseminated to and infiltrated the surrounding structures

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Fig. 1. Arrows indicate flow of barium from hepatic flexure into the subphrenic space.

convinced our oncotherapists that further treatment with cytostatic agents or radiation therapy would be of no avail. Subsequent treatment of the patient consisted of adequate sedation and analgesia to provide the necessary comfort. The patient died 2 months later.

Discussion

In 1958 Hoener reviewed the world literature and found 189 patients with carcinoma of the rectum but only 73 with carcinoma of the colon who were under 20 years of age when the disease was diagnosed. In 1976 Anderson and Bergdahl reviewed the literature and concluded that only 10 patients with colonic carcinoma had been under 10 years of age when the disease was diagnosed. Carcinoma of the colon in children is therefore extremely rare.

We could find no history of previous episodes of ulcerative colitis, adenomatous polyps or familial polyposis in our patient; this endorses the experience of Wolloch and Dintsman and of Van Langenberg and Ong, who could find no antecedent history of intestinal disease to explain the development of colonic carcinoma in children. The experience of these authors clearly indicates that so-called premalignant lesions, such as familial polyposis or ulcerative colitis, are not a prerequisite for colonic carcinoma in children; this is contrary to the experience of Whiteside, who believes that in a small percentage of children ulcerative colitis precedes cancer of the large bowel. Middelkamp and Haffner also endorse the latter view, but in 6 cases reported by Anderson and Bergdahl no instance of colonic carcinoma in a relative could be traced.

Wolloch and Dintsman point out that opinion is divided regarding the pathogenesis of colonic carcinoma in children, and state that there is no proof that adenomatous polyps can undergo malignant transformation. In our case the barium study rendered unequivocal proof that the colon was free of disease except for the isolated area of carcinoma. None of the 68 cases of adenomatous polyposis of the large bowel studied by Turell and Maynard underwent malignant transformation. They concluded that in the majority of children presenting with colonic carcinoma there is no antecedent history of colonic disease; our findings are in agreement with this.

Factors incriminated in the causation of carcinoma of the large bowel in children include diet, chemicals, carcinogens, bile acids, clostridia, IgA deficiency, genetic factors, familial polyposis and ulcerative colitis, but in the majority of cases the cause remains enigmatic. Gross, Stowens and Holt et al. cite a patient with familial polyposis who developed adenocarcinoma. All of these children were under 16 years of age, and the patient cited by Holt et al. was 5 years old when diagnosed.

In 2 of the cases studied by Anderson and Bergdahl the tumour was multicentric in origin with a primary lesion in the transverse colon and ascending colon or the transverse colon and descending colon. In children, as in adults, the tumour occurs more frequently in the left colon, the highest incidence occurring in the sigmoid colon (23%), followed by the ascending colon (18.6%) and the splenic flexure (17.4%). The clinical picture is similar to that in adults, except that the male/female ratio is 2 : 1 in children and 1 : 1 in adults. In the series reported on by Van Langenberg and Ong the male/female ratio was 2.5 : 1.

Signs and symptoms are not dissimilar to those encountered in adults. Of the 25 cases reported by Van Langenberg and Ong, 68% had tarry stools, 56% alteration of bowel habit, 28% abdominal colic, 16% a mass and another 16% tenesmus. Two of their patients presented with a clinical picture indistinguishable from that of acute appendicitis; for the entire group the duration of symptoms ranged from 2 days to 18 months.

In children under 12 years of age tarry stools are frequently overlooked and chronic abdominal pain is often ignored; this is one reason why colonic carcinoma is diagnosed late and why the prognosis is poor. In 50% of cases the carcinoma is of the signet ring or mucin-producing type. Middelkamp and Haffner reviewed microscopic sections from 27 cases and concluded that 48% were of the mucin-producing or signet ring type, about 10 times the frequency in adults. In our case the tumour was a poorly differentiated adenocarcinoma — this contributed to the gloomy outlook since a mucin-producing adenocarcinoma is one reason why colonic carcinoma is diagnosed late and why the prognosis is poor. In 50% of cases the carcinoma is of the signet ring or mucin-producing type. Middelkamp and Haffner reviewed microscopic sections from 27 cases and concluded that 48% were of the mucin-producing or signet ring type, about 10 times the frequency in adults. In our case the tumour was a poorly differentiated adenocarcinoma — this contributed to the gloomy outlook since a mucin-producing adenocarcinoma or poorly differentiated adenocarcinoma of the colon grows rapidly and metastasizes early.

In the 6 patients reported on by Anderson and Bergdahl the longest period of survival was 45 months. Only 2 patients reported in the literature survived more than 5 years after hemicolecotomy, and the overall 5-year survival rate in 79 published cases was 2.5%, this being infinitely worse than that for adults. Of the 81 patients studied by these authors 19 had not been operated on, and 5 had undergone laparotomy only because of metastatic dissemination. Nineteen patients received palliative surgical treatment; in 2 of these survival time was not clearly documented, but the remaining 17 patients all died within 6 months of surgery. In the remaining 38 cases hemicolecotomy was offered in an endeavour to better the prognosis; 67% were alive 1 year after surgery and 20% at 2 years; but only 7% survived for longer than 5 years.

In conclusion, it must be emphasized that colonic carcinoma in a child demands early diagnosis as it carries a grave prognosis. Early detection can be accomplished by paying meticulous attention to the clinical history of loss of appetite, vomiting, weight loss and anaemia or the passing of a tarry stool. Early barium examination of the large bowel and sigmoidoscopy/colonoscopy will no doubt make a valuable contribution in bringing to the clinician's attention the presence of a tumour which would otherwise remain silent until too late to treat.

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REFERENCES


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