Acute adult intussusception caused by primary cecal non Hodgkin lymphoma

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Abstract

Intussusception of the bowel is rather rare in adults and in about 80-90% of the cases is secondary to an underlying intraluminal pathology that serves as a lead point. In cases of colonic intussusception malignancy occurs in 63-66% of patients and it is usually adenocarcinoma and rarely lymphoma. The presenting symptoms are non specific and are in most cases of long duration, consistent with a chronic intussusception causing partial intestinal obstruction. We present a rare case of primary colorectal lymphoma in a 29 years old female that was complicated by acute ileocecal intussusception, treated with en bloc resection without reduction. (Acta gastroenterol. belg., **2011**, 74, **451-453**).

Key words : gastro-intestinal lymphoma, colon, obstruction, surgery.

Introduction

Lymphoma of the colon is a rare malignancy and represents 0.16-1.2% of all cases of colon cancer (1,2) and 5,6-20% of all cases of gastrointestinal lymphoma (1,2). It has a male predominance (1,3) and a maximum incidence in the 50 to 70 years. Lymphoma most commonly occurs in the caecum (1,2,3) and the most common histological types are diffuse large B cell lymphoma and Burkitt lymphoma (2,4). The patients usually present with unspecific symptoms such as abdominal pain, weight loss, anorexia, change in bowel habit and an abdominal palpable mass (1,3,4). Complications such as obstruction, perforation and intussusception are occasionally presented (1). We present a rare case of primary colorectal lymphoma in a 29 years old female that was complicated by acute ileocecal intussusception.

Case report

A 29 years old female was referred to our department from the hematology department for emergency surgical management of a non Hodgkin lymphoma of the cecum that was complicated by bowel obstruction due to ileocecal intussusception. The patient's medical history included polycystic ovary syndrome for 13 years.

The patient first presented approximately 5 months ago with abdominal pain and diarrhea. Laboratory examinations and abdominal X-ray were within the normal range. The patient had undergone an abdominal CT that revealed wall thickening of the terminal ileum of 5 cm in length and stenosis of its lumen that was attributed to inflammatory bowel disease, and particularly Crohn disease. A colonoscopy was performed, but because of a non-proper preparation, examination was limited to approximately 70 cm of the large bowel and the biopsies taken revealed chronic inflammation. The patient received therapy with ciprofloxacin, metronidazole, methylprednisolone and mesalazine. Because of persistent symptoms, weight loss, and a palpable abdominal mass in the right lower quadrant about four and a half months after the initial presentation, another colonoscopy was performed that showed a large ulcerated exophytic mass of the cecum. The biopsy revealed diffuse large B-cell lymphoma and immunohistochemistry showed intense positivity of the neoplasmatic cells for CD20, CD79a, Bcl-2, Bcl-6 and MUM-1 and negative for CD5, CD10, CD30 and Tdt. The Ki-67 was over 90% (Fig. 1).

The patient was admitted to the hematology department for staging of the lymphoma, but after two days she presented with acute vomiting and the CT scan revealed ileocecal intussusception, with the leading point being the mass near the ileocecal valve, causing small bowel obstruction (Fig. 2). The patient had an emergency laparotomy in which a large mass of the cecum and ileocecal intussusception was found. A right hemicolectomy, without reduction of the intussusception was performed. The lymphoma was located to the cecum, adjacent to the ileocecal valve and measured 8.5 × 6.5 × 2.5 cm (Fig. 3). The surgical margins were free of disease and 7 out of the 25 lymph nodes were infiltrated. According to the current diagnostic criteria the lymphoma was considered a primary colonic lymphoma.

The patient's postoperative course was uneventful and she was discharged after 10 days. The diagnosis of primary colonic lymphoma was established following cervical, thoracic and abdominal CT scans which did not reveal any enlarged lymph nodes and the bone marrow aspiration and biopsy were normal. Postoperatively, the patient was treated with 3 cycles of a chemotherapy regimen including rituximab, adriamycin, vincristine, cyclophosphamide and prednisone.

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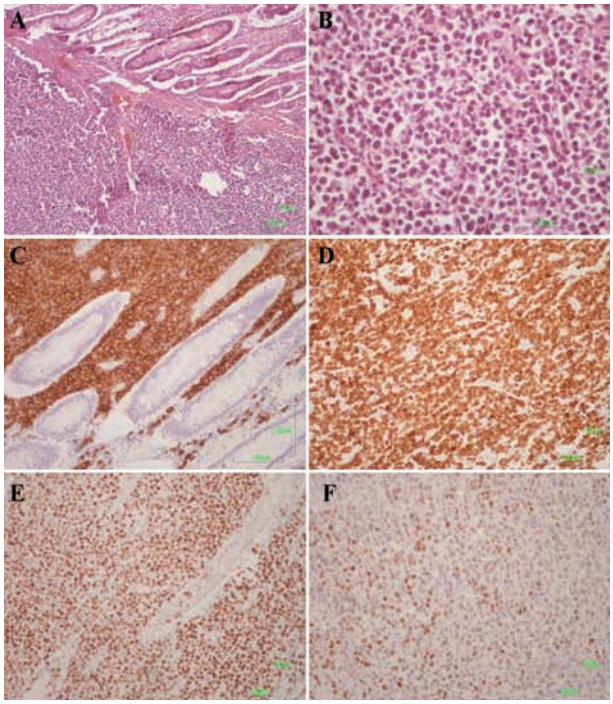

Fig. 1. — A. The neoplasmatic cell infiltration was diffuse (hematoxylin and eosin ×100). B. The neoplasmatic cells were large, with round nuclei and multiple small nucleoli (hematoxylin and eosin ×400). C. Positive immunostaining for CD20 showed that these cells were of B-cell origin (CD20 ×100). D. Extensive postitivity for bcl-2 (bcl-2 ×100). E. Extensive nuclear staining for Ki-67 was up to 90% (Ki-67 ×100). F. Moderate nuclear positivity for bcl-6 in a high proportion (bcl-6 ×100).

After four months there is no evidence of recurrent disease or systematic lymphoma.

Discussion

Intussusception of the bowel occurs when a proximal segment of the gastrointestinal tract telescopes within the lumen of the adjacent segment (5). It is rather rare in adults and accounts for 1-5% of all cases of bowel obstruction (5) and 0.003-0.02% of all hospital admissions (6). In adults about 80-90% of the cases is secondary to an underlying intraluminal pathology that serves as a lead point (5,6). In cases of colonic intussusception malignancy occurs in 63-66% of patients and it is usually adenocarcinoma and rarely lymphoma (5,7). In the present case there was ileocecal intussusception with the leading point being the cecal lymphoma that was adjacent to the ileocecal valve.

The presenting symptoms are non specific and are in most cases of long duration, consistent with a chronic intussusception causing partial obstruction (5,8). The clinical presentation usually includes abdominal pain, nausea, vomiting and less frequently constipation, change in bowel habits, gastrointestinal bleeding, weight loss and abdominal distension (5,8). Because the symptoms are non specific correct preoperative diagnosis is difficult (9).

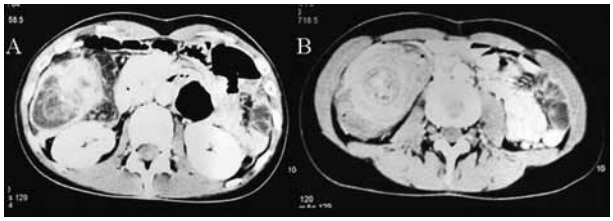

Fig. 2. — A. CT scan showing the cecal lymphoma and the intussusception. B. The pathognomonic unhomogenous "target" shaped soft tissue mass is shown.

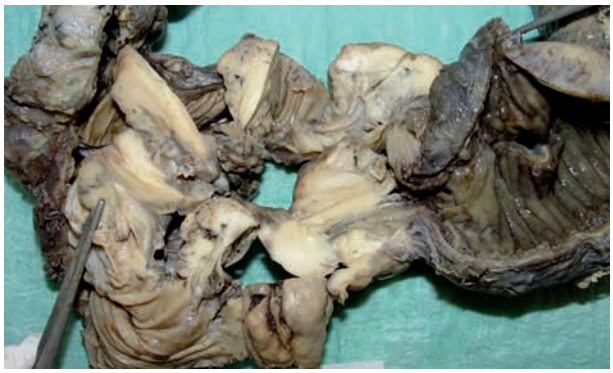

Fig. 3. — The surgical specimen showing the cecal lymphoma.

Abdominal CT is the examination of choice as it can aid the diagnosis of intussusception due to its virtually pathognomonic appearance (5,8) and is superior to ultrasonography and endoscopy (9). The characteristic findings of the CT scan include an unhomogenous "target" shaped soft tissue mass with a layering effect, when the CT beam is perpendicular to the longitudinal axis of the intussusception, as in the current case, or a "sausage" shaped soft tissue mass with a layering effect, when the beam is parallel to the longitudinal axis of the intussusception (5,8,9). Also, mesenteric vessels within the bowel lumen are typical (5,9).

Surgical intervention is considered necessary because of the large portion of structural anomalies and the high incidence of underlying malignancies (5,9), but the optimal surgical treatment remains controversial. Reduction before resection may permit a more limited resection but includes the risk of intraluminal seeding and venous tumour dissemination, of perforation and seeding of microorganisms and tumour cells in the peritoneal cavity and of anastomotic complications of the manipulated friable and edematous bowel tissue (5,9). In our case we performed an en bloc resection without reduction as the diagnosis of lymphoma was already known.

Regarding the treatment of lymphoma, surgical excision is the mainstay for treatment of primary colonic lymphoma. The diagnosis of primary lymphoma is established using the following criteria : when the

patient is first seen there is no palpable superficial lymphadenopathy, chest radiographs show no obvious enlargement of the mediastinal nodes, the white blood cell counts, total and differential are within the normal limits, at laparotomy only regional nodes are affected by disease, the liver and spleen seem free of tumour, a normal bone marrow biopsy, the absence of lymphadenopathy on CT scan of the mediastinum and a histologically confirmed colorectal lymphoma (1,4). Following surgical resection, adjuvant chemotherapy is usually administrated with a regimen including cyclophosphamide, vincristine, doxorubicin and prednisone (CHOP) (3,4) while adjuvant radiotherapy should only be used after incomplete resection (3).

Primary colorectal lymphoma carries a poor prognosis compared with primary gastric lymphoma and carcinoma of the colon (1). Prognosis is poorer in patients with an initial tumour larger than 5 cm and with tumour positivity of locoregional lymphnodes (1,4). The five year survival rate varies between 27-55% (3,4).

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