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CLINICAL CASE

A rare case of mantle cell lymphoma as lymphomatous polyposis with widespread involvement of the digestive tract

Lymphome du manteau sous la forme d'une polypose diffuse du tube digestif

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Summary Lymphomatous polyposis of the gastrointestinal tract is rare. It refers to a heterogeneous group of small B-cell lymphomas including mantle cell lymphoma, follicular lymphoma and MALT lymphoma. It is characterized by the presence of multiple lymphomatous polyps along one or more segments of the digestive tract. Clinical symptoms are non-specific. We herein report the case of a 74-year old man initially admitted for an upper and lower gastrointestinal endoscopy to explore a positive Hemoccult® test. The endoscopy revealed multiple polyps all along the gastrointestinal tract. Histopathological study showed a diffuse lymphomatous proliferation of small B-cells whose immunohistochemical features were compatible with a mantle cell lymphoma. Tumoral B-cells showed a positivity of cyclin D1 markers but negativity for CD5. Immunochemotherapy with R-CHOP (rituximab, cyclophosphamide, adriamycine, vincristine and prednisone) was initiated. Based on this case study, the pitfalls of gastrointestinal tract lymphomatous polyposis diagnosis, prognosis and treatment options are discussed.

Introduction

Digestive lymphomas represented 12.5% of all non-Hodgkin lymphoma (NHL) and 36% of extra-nodal NHL. They represent only 3% of malignant tumors in the stomach, less

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than 1% those of the colon and rectum and less than 18% those of small bowel, which are themselves very rare [1]. Digestive lymphomatous polyposis (DLP) is a rare type of digestive tract NHL. It is composed of a heterogeneous group of lymphomas including mantle cell lymphoma, follicular lymphoma, and MALT lymphoma [2]. The symptoms are non-specific, occurring most often in men around age 60. We report the case of multiple lymphomatous polyposis (MLP) corresponding to a mantle cell lymphoma with diffuse gastric, duodenal, colon and rectal localizations.

Observation

A 74-year-old man was admitted for gastrointestinal endoscopy to explore a positive Hemoccult® test. The patient had no bleeding or alteration of his general condition. The physical examination of abdomen was normal. His only past medical history consisted in an achalasia of the esophagus treated by Heller's cardiomyotomy. Upper and lower endoscopies were performed under general anesthesia. The endoscopic examinations objectified a disseminated polyposis in the stomach, duodenum, colon and rectum. The upper gastrointestinal endoscopy revealed two small ulcers in the antrum and duodenal polyps up to D2 with some greater than 1 cm and ulcerated. Colonoscopy found the same type of polyps throughout the colon and the rectum (Fig. 1). Multiple biopsies of these polyps were performed.

Histological examination showed a diffuse lymphomatous proliferation with small cells in the stroma, muscularis mucosae and extending to the submucosa. This lymphoid infiltrate repressed and stifled the epithelial structures but without lymphoepithelial lesions (Fig. 2). The tumoral B cells were of small to medium size. The immunohistochemical staining showed positivity of lymphoma cells for CD20, Bcl2, Bcl1 (cyclin D1) and CD43, and negativity for CD3, CD5, CD23 and CD10. The proliferation index assessed with the anit-Ki-67 was 15%. The same morphology and immunohistochemical staining was observed on polyps biopsies from different gastrointestinal localizations. The immunohistochemical profile was in accordance with a disseminated lymphomatous polyposis of the gastrointestinal tract corresponding to a B mantle cell lymphoma. The diagnosis was confirmed by fluorescent in situ hybridization (FISH), which showed a cyclin D1 gene rearrangement.

Physical examination revealed a left axillary lymph node and bilateral inguinal lymph nodes. Biologically, the complete blood count was normal with 8400/mm^3 leukocytes, hemoglobin of 13.1 g/dl and 187 G/l platelets. The LDH was normal but beta-2 microglobulin increased to 3.2 mg/l (normal lower than 2.2 mg/l). There was no medullar invasion on the myelogram. Thoracic and abdominopelvic CT-scan highlighted the presence of multiple lymph nodes above and below the diaphragm. Position emission tomography (PET-scan) showed an intense hypermetabolism of all the digestive structures, extending from the stomach to the rectal wall associated with multiple lymph nodes from the cervical to the inguinal regions.

In conclusion, the patient had a MLP form of mantle cell lymphoma with involvement of stomach, duodenum, colon, rectum and lymph nodes. It was decided at a multidisciplinary meeting that an immunochemotherapy with R-CHOP (rituximab, adriamycin, cyclophosphamide, vincristine and prednisone). The patient received 8 cycles of chemotherapy. The follow-up evaluation showed a partial response. There was a complete disappearance of adenopathy on CT-scan and PET-scan but a persistence of some gastrointestinal polyps with lymphoma cells. A chemotherapy consolidation with aracytine is performed.

Discussion

MLP was first described by Cornes in 1961 [3]. Several cases of a MPL form of mantle cell lymphoma have been reported [4—7] and the largest series reported the follow-up of 31 patients with MLP [8]. Its frequency is estimated at 9% of primary gastrointestinal lymphoma with a predominantly male population (65%) and a mean age of 63 years at diagnosis. The involvement of many segments of the digestive tract without clinical symptoms, as in the case reported here, is very rare [7,8]. In the largest published series, the 31 cases all had digestive problems. The main inaugural symptoms are abdominal pain, diarrhea and gastrointestinal bleeding. Obstructive tumors leading to surgery are rare. Involvement of the colon and small bowel is almost constant (90% of cases), followed by involvement of the rectum (60% of cases) and stomach (50% of cases) [8]. Involvement of the esophagus is exceptional. A mesenteric lymph node invasion is frequently found at diagnosis as in our case report. The main extra-digestive sites that may be affected are the bone marrow, peripheral lymph nodes, Waldeyer's ring and the liver.

The morphological features of digestive mantle cell lymphoma can be mistaken for those of MALT lymphoma and diffuse follicular lymphoma. An immunohistochemical study

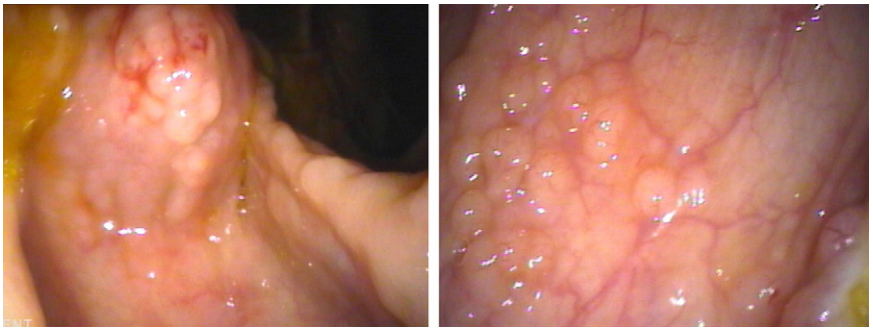

Figure 1 Lymphomatous polyposis of the colon.