Gastrointestinal

Computed tomography findings of diffuse gastrointestinal mantle cell lymphoma

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ABSTRACT

Multiple lymphomatous polyposis is an uncommon type of primary non-Hodgkin’s lymphoma, characterized by multiple lymphomatous polyps along the gastrointestinal tract. We present 2 cases of diffuse gastrointestinal involvement and illustrate radiological and pathologic findings.

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Introduction

There are several causes for intestinal polyposis, most commonly representing hereditary forms of adenomatous or hamartomatous polyps. Lymphoma presenting as multiple intestinal polyps is rather uncommon. Multiple lymphomatous polyposis (MLP) is a rare type of primary non-Hodgkin’s lymphoma, characterized by multiple lymphomatous polyps along the gastrointestinal tract. We present 2 cases of diffuse gastrointestinal involvement and illustrate radiological and pathologic findings.

Case report

Case 1

The patient is a 49-year-old man diagnosed with HIV infection 5 years ago and receiving highly active antiretroviral therapy...
(HAART) for the past 3 years. He complained of intermittent diarrhea, abdominal pain, and weight loss (10 kg) for the last 3 months. His physical examination revealed a painful abdominal mass in the right iliac fossa, and bilateral palpable lymph nodes in cervical and inguinal region. Computed tomography (CT) showed multiple polyps throughout the entire gastrointestinal tract, multiple enlarged lymph nodes in the mesentery (Fig. 1), and an exuberant thickening of the distal ileum, which represented the clinically palpable mass. Colonoscopy and upper gastrointestinal endoscopy confirmed CT findings of multiples polyps through the gastrointestinal tract. Biopsies of the lesions and bone marrow biopsy were performed.

Case 2
A 64-year-old and previously healthy male was admitted to our hospital with a 10-month history of bloody diarrhea, abdominal pain, fatigue, and weight loss (20 kg). On physical examination, there were signs of anemia and an epigastric palpable mass without tenderness. Lymph nodes were not palpable. Laboratory studies included the following: hemoglobin 7.8 g/dl (normal 13-18) and white blood cell count 1624/mm³ (4000-10,200). Abdominal CT showed multiple retroperitoneal and mesenteric lymphadenopathy and hepatosplenomegaly. Gastric and colonic wall thickening and also multiple polyps through the gastrointestinal tract were also noted (Fig. 2).

Fig. 1 – Axial post-contrast computed tomography (CT). (A) Diffuse gastric wall thickening (arrow). (B) Multiple polyps throughout the small bowel and colons (arrows).

Fig. 2 – Axial (A, B, and C) and coronal (D) post-contrast computed tomography (CT) showing multiple polyps throughout the small bowel and colons (arrows) and also lymphadenopathy (dotted circle). Notice diffuse terminal ileum thickening (C and D), palpable on physical examination (arrowhead).
Colonoscopy revealed diffuse colonic polyposis and some infiltrative and ulcerated lesions. Upper gastrointestinal endoscopy revealed a gastric thickened mucous membrane with multiple gastric and duodenal polyps (Fig. 3). Biopsies of the lesions and bone marrow biopsy were performed (Fig. 4). According to these findings, both cases were diagnosed as MLP.

Fig. 3 – Upper gastrointestinal endoscopy (A) and correspondent endoscopic ultrasound (B) show diffuse gastric thickening up to 3.0 cm. Colonoscopy (C) confirms multiple polyps.

Fig. 4 – Histologic sections of duodenum and colon showed monomorphic proliferation of small to medium-sized lymphoid cells with slight irregular nuclear contours and a predominant diffuse growth pattern involving mucosa. Immunophenotypically, cells were positive for CD20, CD5, and cyclin d1. Bone marrow biopsy showed infiltration by mantle cell lymphoma (MCL).
Discussion

The term “multiple lymphomatous polyposis” (MLP) was first coined by Cornes [1] in 1961, describing 5 patients with multiple, sessile or pedunculated polyps throughout the gastrointestinal tract, consisting of mucosal involvement by malignant lymphoma.

MLP is an uncommon type of primary non-Hodgkin’s gastrointestinal B-cell lymphoma and is regarded as the intestinal form of mantle cell lymphoma (MCL). Although there is close relationship between these 2 entities, some follicular lymphoma and mucosa-associated lymphoid tissue (MALT) lymphoma can also present as MLP [2].

MCL usually develops in elderly men with advanced stage and frequent extra nodal involvement and bone marrow infiltration, with aggressive clinical behavior and poor response to conventional therapy [3]. When associated with HIV infection, lymphomas are more likely to affect extra nodal sites and are of higher grade [4].

On imaging examinations, intestinal MCL is characterized by multiple polyps throughout the gastrointestinal tract, similar to several colonic polyposis such as hereditary polyposis. However, the diagnosis should be considered when gastrointestinal polyposis is found in elderly people, HIV infection, or when associated with bowel wall thickening without obstruction, widespread lymph node enlargements, and splenomegaly [5,6].

Histologically, MCL shows a monomorphic lymphoid proliferation composed of small- to medium-sized lymphoid cells with slightly to markedly irregular nuclear contours, with a vaguely nodular, diffuse, or mantle zone growth pattern. These cells are typically CD20 and CD5 positive, and express cyclin D1 [7]. When involving gastrointestinal tract, these can present either as a nonspecific lymphocytic infiltrate or as MLP, and may represent the exceedingly common secondary intestinal dissemination of a primary nodal MCL. The differential diagnosis includes lymphoid hyperplasia and other B-cell neoplasms, such as MALT lymphoma [8].

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