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Mantle cell lymphoma as a rare cause of intussusception: a report of 2 cases

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Abstract Intussusception is uncommon in adults and is only very rarely caused by malignant lymphoma. To our knowledge, there are only 2 previously reported cases of mantle cell lymphoma causing intussusception. We present 2 additional cases of intussusception at the ileocecal valve in patients being treated for mantle cell lymphoma, and a review of the pertinent literature is presented.

Keywords: Mantle cell lymphoma; Intussusception

1. Introduction

Intussusception is rare in adults and is usually associated with a pathologic mass that acts as a lead point. In the small intestine, benign neoplasms are the most common lead points, with lipomas and hamartomatous polyps accounting for most of the cases. On the other hand, adenocarcinomas are responsible for more than 50% of intussusceptions of the colon. Malignant lymphoma is an uncommon cause of intussusception. A summary of 5 large series of intussusception in adults identified malignant lymphoma as the etiologic factor in less than 1% of cases [1].

The most common lymphoma causing intussusception is diffuse large B-cell lymphoma. Although mantle cell lymphoma (MCL) often shows extensive involvement of the gastrointestinal tract, only 2 cases of intussusception attributable to MCL have been reported [2,3]. We herein describe and discuss the features of 2 additional patients with MCL who were found to have gastrointestinal involvement after presentation with intussusception.

2. Case descriptions

2.1. Case 1

A 38-year-old man presented with a 1-week history of enlarging bilateral neck masses, weight loss, fatigue, and several weeks of drenching night sweats. Computed tomography (CT) scan of the chest, abdomen, and pelvis showed diffuse lymphadenopathy. An inguinal lymph node biopsy was carried out at an outside institution, and a diagnosis of MCL was made on the formalin-fixed paraffin-embedded tissue. The diagnosis was confirmed by a fine needle aspiration biopsy that included surface marker analysis by laser scanning cytometry. The cells showed bright expression of surface κ light chain, CD20 and FMC7, and coexpressed CD19 and CD5. They were negative for CD23 and CD10. An identical immunophenotype was shown by flow cytometry analysis of a bone marrow aspiration biopsy. Fluorescence in situ hybridization analysis carried out on isolated cells showed a signal pattern consistent with a t(11;14)(q13;q32) (Fig. 1).

Combination chemotherapy was begun with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP). During the second course of the treatment cycle, the patient presented to the emergency department with severe right lower quadrant abdominal pain. No evidence of obstruction or perforation was found, and the pain resolved spontaneously. During the third cycle of chemotherapy, the patient developed a recurrence of the right lower quadrant pain, this time of intermittent, cramping quality and variable intensity. A planned restaging CT scan carried out after the third chemotherapy cycle showed a long segment of ileocolic intussusception with a 2.0-cm pathologic lead point. The lead point was interpreted to be consistent with a lipoma. The intussus-

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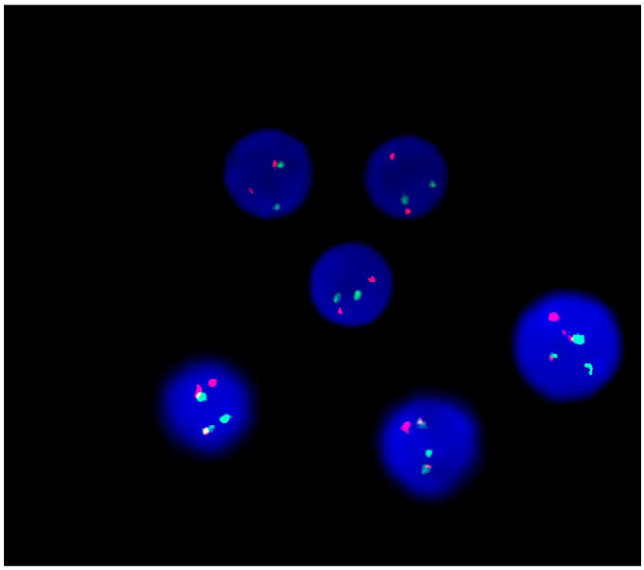

Fig. 1. Fluorescence in situ hybridization performed using the CCND1/IGH dual color, dual fusion XT translocation probe showed a fusion signal consistent with the presence of a t(11;14)(q13;q32).

ception did not resolve spontaneously, and the patient underwent a right hemicolectomy.

2.2. Case 2

A 71-year-old woman presented with masses in the right breast and right postauricular soft tissue. Six years earlier, she had been diagnosed with stage IV follicular lymphoma, for which she underwent treatment with 8 cycles of CHOP chemotherapy, with a good response. Three years before presentation, she developed a left postauricular soft tissue recurrence that was treated by radiotherapy alone. Her current disease proved to be unresponsive to treatment with rituximab, and therefore, reevaluation of the disease process was initiated.

Fine needle aspiration of the breast mass revealed a monotonous population of intermediate-sized lymphoid cells with irregular nuclear contours. Flow cytometry analysis showed a monoclonal B-cell population with the marker profile surface immunoglobulin λ+, CD19+, CD20+, CD5+, FMC7+, CD10−, CD11c−, and CD23−. Immunoperoxidase analysis carried out on a glass slide showed positive nuclear staining of the tumor cells for cyclin D1. These findings were considered to be consistent with MCL.

One month after presentation, the patient developed abdominal pain, vomiting, and rectal bleeding. A CT scan showed an ileocolic intussusception with a 5.2 × 4.8-cm lead point. The radiographic appearance was interpreted to be consistent with either a leiomyoma or lymphoma. Colonoscopic examination revealed a large mass in the cecum, which was considered to be highly suspicious for a carcinoma, and smaller, raised, smooth-surfaced lesions in the transverse colon, at the splenic flexure, and in the sigmoid colon. All of these lesions underwent biopsy, and all showed

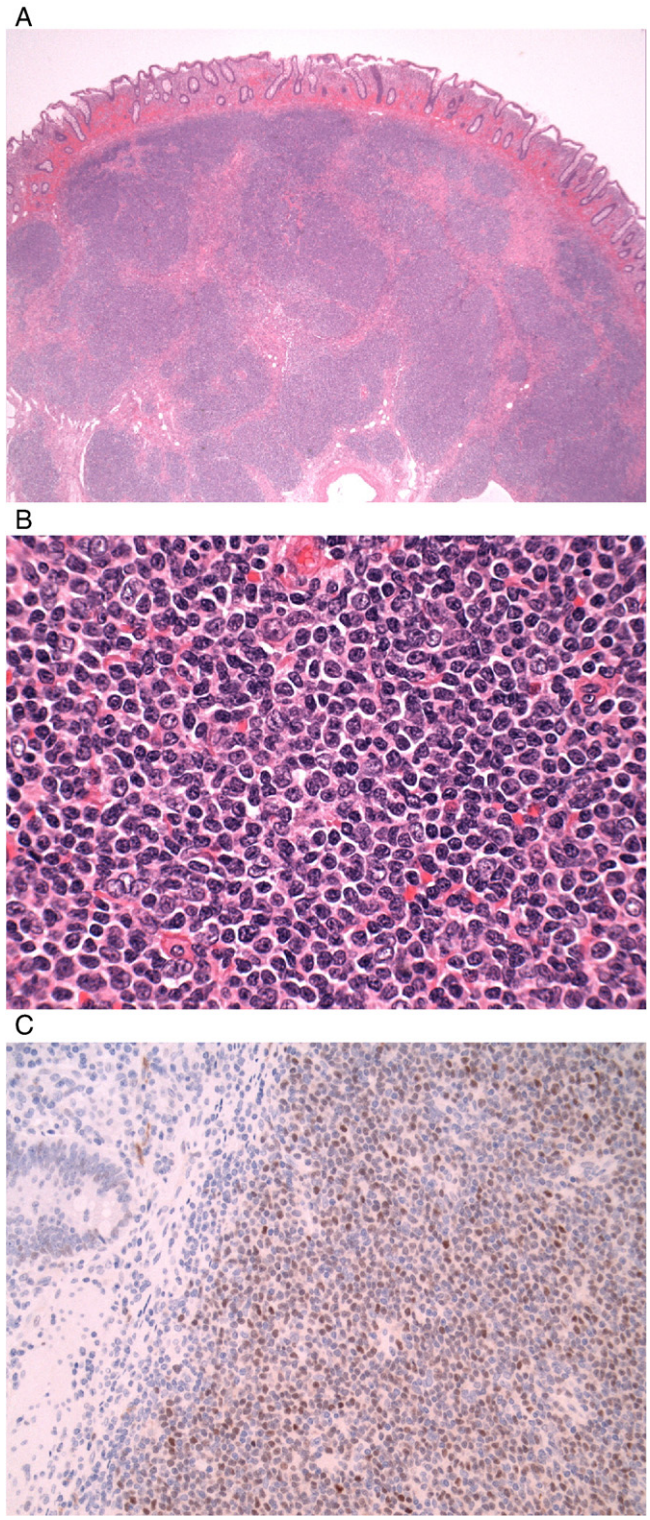

Fig. 2. Case 1. (A) The lymphoid infiltrate filled the colonic submucosa in the region of the ileocecal valve. (B) Cells were small to medium with irregular nuclei, coarse chromatin, indistinct nucleoli, and scanty pale cytoplasm. Transformed lymphocytes were absent. (C) Cyclin D1 was positive in most nuclei.