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Nodular lymphoid hyperplasia complicated with ileal Burkitt's lymphoma in an adult patient with selective IgA deficiency

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ABSTRACT

INTRODUCTION: Primary lymphomas of the small intestine are rare. Burkitt's lymphoma (BL) occurs sporadically in adults. Nodular lymphoid hyperplasia (NLH) is a rare disorder characterized by diffuse nodular lesions, which represent hyperplastic lymphoid follicles, and it is often associated with immunodeficiency syndromes.
PRESENTATION OF CASE: We present a 38-year-old male patient in a state of surgical emergency, suspected of Crohn's disease, who had an unusual combination of NLH and BL of the proximal ileum. Furthermore, retrospectively analyzed documentation revealed selective IgA deficiency.
DISCUSSION: Association between NLH and intestinal lymphomas in patients with immunodeficiency syndromes was indicated before. This case report supports the notion on NLH as a transition state between immunodeficiency and intestinal lymphomas.
CONCLUSION: This is one of the first case reports which presents the combination of NHL and BL. The awareness of the existence of this rare combination, especially in young adult males, can improve the diagnostic accuracy and the treatment management.

1. Introduction

Primary lymphomas of the small intestine are rare and mostly of B-cell and non-Hodgkin types [1]. The problem is that they are hardly recognized unless intestinal complications occur. Their accurate diagnosis is often difficult, because the immune system of the gut can respond on many immunologic stimuli with nonspecific hyperplastic reaction [2]. The most common sites include ileum and terminal ileum, followed by the jejunum and duodenum [1]. Diffuse large B-cell lymphoma (DLBCL) is the most common histological subtype in the GI tract, in general. Other histological subtypes in the small intestine include mucosa-associated lymphoid tissue (MALT) lymphoma, enteropathy-associated T-cell lymphoma (EATL), mantle cell lymphoma (MCL) and follicular lymphoma (FL). BL occurs sporadically, mostly in boys and adolescent males, representing with symptoms of small bowel obstruction [3,4].

This case report is one of the first presenting a rare combination of BL of the ileum and nodular lymphoid hyperplasia (NLH), probably due to selective IgA deficiency [5]. NLH is a condition characterized with numerous small nodules diffusely distributed along the segments of the GI tract, histologically evaluated as reactive follicular hyperplasia [6]. In children, NLH has a benign course and is often associated with viral infections and food allergies [7]. In adults, associations were found with immunodeficiency disorders, Giardia infection, celiac disease and HP infection [8]. In some cases, lymphomatous transformation has been reported [9].

2. The presentation of the case

A 38-year-old male was admitted to the Department of Surgery for abdominal colic, vomitus, stomach bloating and distention. He had a six-month history of episodes of abdominal colicky pain localized in the periumbilical area. Upon physical examination, the abdomen was distended, with the signs of the wall tenderness and of reduced bowel movements; radiologic assessment indicated the signs of ileus. Laboratory checkup did not indicate significant results, except for mildly increased C-reactive protein (CRP), 33.0 mg/L, and fibrinogen activity level, 3.8 g/L. Gastroscopy indicated multiple tiny polyps in the whole duodenum, histopathologically confirmed as inflammatory infiltrates. Colonoscopy with terminal ileoscopy was also performed to confirm or reject the diagnosis of Crohn's disease (CD). This examination revealed numerous

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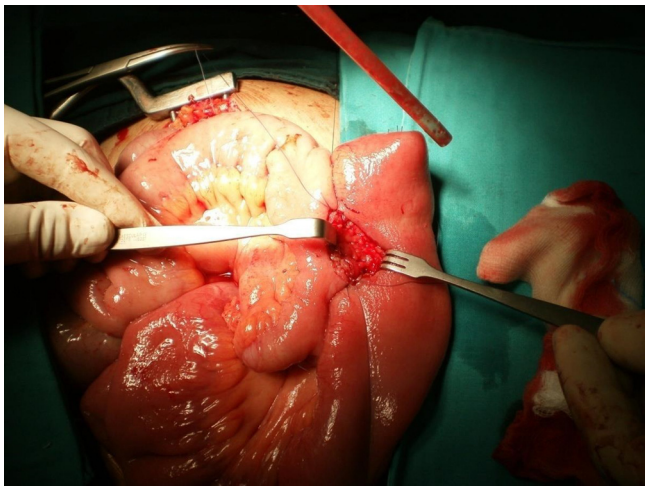

Fig. 1. Numerous tiny polyps diffusely distributed throughout the small intestine mucosa.

polyps, measuring 2–3 mm in diameter, in the cecum. Mucosa in the terminal ileum was hyperemic and edematous, with creases and wrinkles and cobblestone-like appearance. Histopathological examination of the bioptic specimens revealed abundant chronic inflammatory infiltrates, Paneth cell metaplasia and isolated lymphatic follicles with reactive germinal centres, but these findings were not deemed sufficient to confirm the diagnosis of CD. Abdominal ultrasound revealed two small polypoid tumors in the gallbladder, increased small intestine wall thickness, in the segment of 8 cm in length, as well as lumen stenosis, in the parts projected behind the posterior urinary bladder wall. Magnetic resonance (MR) enterography confirmed increased wall thickness of the terminal ileum, visible along the segment of 5 cm in length, and luminal narrowing, in the parts located above the urinary bladder. The evident intraabdominal lymph node or other organs' enlargements were not found. Radiographs of the small intestine indicated delayed contrast discharge at the level of the jejunal loops. The results of other tests performed, including testing for HIV and hepatitis B and C, PPD (purified protein derivative) skin test and parasites in stool, were all negative. Immunoglobulin blood test revealed selective IgA deficiency (IgA values less than 0.05 g/L). Anamnesis indicated a long-term presence of atopic dermatitis of hands and legs as well as chronic rhinosinusitis.

These results, taken together with the clinical findings, were highly indicative of CD. Adequate treatment had already been initiated earlier on. Due to the symptoms and signs of the acute small intestine obstruction at the admittance, the patient underwent the emergency operation. The tumor mass found intraoperative, ranging 3 cm in diameter, caused the complete obstruction of the lumen of the proximal ileum. Surrounding mucosa was densely populated with small polyps, measuring 2–3 mm in diameter. Segmental resection of 11 cm long part of the intestine, with the tumor herein, was performed, and the intestinal continuity was reestablished by latero-lateral anastomosis. There were no further gross pathological findings, except for dilated jejunal loops. Mucosa in the rest of the small intestine also indicated polypoid appearance (Fig. 1).

Histopathological analysis of polypoid intestinal specimens was indicative of reactive follicular lymphoid hyperplasia, showing large lymphoid follicles with prominent germinal centres (Fig. 2). Microscopic examination of the solitary cross-sections of the tumor tissue revealed diffuse cellular infiltrations with medium size to large atypical lymphoid cells and slight nuclear polymorphism. Apoptotic bodies and "tingle body" macrophages were scattered here and there, altogether giving the "starry-sky" appearance (Fig. 3).

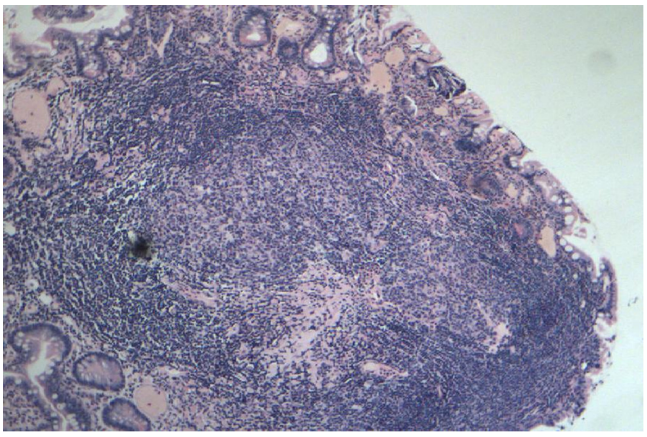

Fig. 2. Microscopic appearance of the lymphoid polyp found in mucosa of the small intestine (H&E, x100).

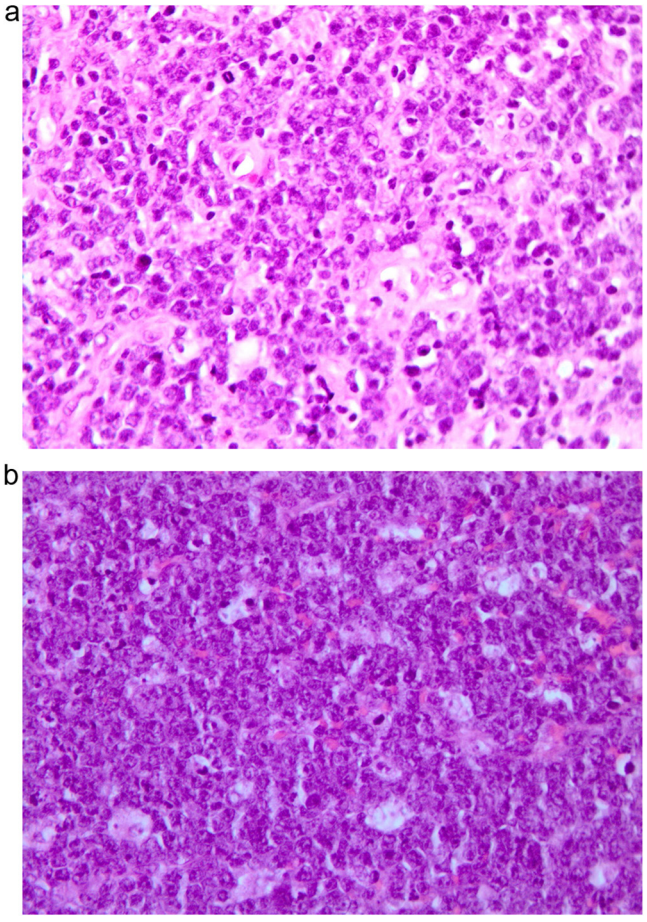

Fig. 3. Primary extranodal B cell non-Hodgkin's lymphoma of the small intestine; the histopathological pattern of the "starry-sky" (H&E, x40).

Immunohistochemical findings revealed the tumor cell phenotype as CD20+, bcl6+ (moderate), bcl2-, CD10+, CD43-, TDT-, CD3-, CK AE1/3-, chromogranin-. CD10 indicated strong positivity (Fig. 4). Proliferative activity, measured by Ki67 expression, was found in almost all cells (Fig. 5). FISH (fluorescence *in situ* hybridization) analysis identified the t(8;14) (C-MYC/IGH) chromosomal translocation. These histopathological and phenotypic analyses pointed to the diagnosis of BL.

Extensive disease staging, including evaluation of the bone marrow aspirate smear and bone biopsy, as well as multi-slice helical