Unusual cause of adult intussusception: diffuse large B-cell non-Hodgkin’s lymphoma: a case report and review

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Abstract. – BACKGROUND: Intussusception is defined as the telescoping of a segment of the gastrointestinal tract into an adjacent one. A demonstrable etiology is found in 70% to 90% of cases in adult patients, and about 40% of them are caused by a primary or secondary malignant tumor. The aims of this study were to give an overview of the literature on intussusception due to gastrointestinal lymphoma.

MATERIALS AND METHODS: We present a case of ileocecal intussusception secondary to non-Hodgkin’s lymphoma (NHL), as well as a literature review of studies published in the English language on intussusception secondary to lymphoma, accessed through PubMed and Google Scholar databases.

RESULTS: Thirty-six published cases of intussusception caused by lymphoma were evaluated, and a case of ileocecal lymphoma in a 62 year-old woman is herein presented. In the reviewed literature, 33 reports meeting the aforementioned criteria were included in this review. The patients were aged from 16 to 86 years (mean, 48.2 +/- 19.0 y). Twenty-nine were male and seven were female. According to the localization of lymphoma, 24 patients had ileo-colic intussusception, 10 had enteric, and 2 had colic intussusception. In terms of the diagnosis, 34 patients were diagnosed with various types of NHL, and two patients were diagnosed with HL.

CONCLUSIONS: Despite the rarity of intussusception cases secondary to malignant causes, particularly lymphoma, it is rather difficult to diagnose preoperatively by surgeons. Because there exists a risk of malignancy in a substantial portion of adult intussusception cases, resection should be performed in a manner consistent with the oncological principles.

Key Words: CD20, Intussusception, Lymphoma.

Introduction

Intussusception occurs when a more proximal portion of bowel (intussusceptum) invaginates into more distal bowel (intussuscipiens)3-31. The pathomechanism is thought to involve altered bowel peristalsis at the intraluminal lesion, which is then a lead point for the intussusception. Although intussusception is a common condition in children, it is a rare entity in the adult population. Adult intussusception represents 5% of all intussusceptions and 1% to 5% of all cases of intestinal obstruction in adults31-35. Intussusception in adults is distinct from pediatric intussusception in many aspects. In contrast to intussusceptions in children, a demonstrable etiology is found in 70% to 95% of cases in the adult population, and approximately 40% of them are caused by primary or secondary malignant neoplasms1,2,4,32,34,35. In general, the majority of lead points in the small intestine consist of benign lesions. Malignant lesions account for up to 30% of cases of intussusception in the small intestine. Intussusception occurring in the large bowel is more likely to have a malignant etiology and represents 63% to 68% of cases32,33,35-37. When we take a look at malignant diseases leading to intussusception, while adenocarcinoma, particularly metastatic carcinoma, is found to be the most frequent cause in the colon; primary adenocarcinoma, gastrointestinal stromal tumors (GISTs), lymphoma, and carcinoid tumors are seen in the small intestine35.

The gastrointestinal tract is the most common extranodal site affected by lymphoma, accounting for 5% to 20% of all cases. Primary gastrointestinal lymphoma, however, is very rare, constituting only about 1% to 4% of all gastrointestinal malignancies. Gastrointestinal lymphoma is usually secondary to widespread nodal diseases. The most frequently affected sites are the stomach, followed by the small bowel (20%-30%) and colon4,38. Histopathologically, almost 90% of primary gastrointestinal lymphomas are B-cell non-Hodgkin’s lymphoma (NHL), followed by T-cell NHL and Hodgkin’s lymphoma (HL).

Primary malignant tumors of the small intestine are very rare, accounting for less than 2% of all...
gastrointestinal malignancies. Intestinal lymphoma constitutes 10% to 20% of all small intestine neoplasms and 20% to 30% of all primary gastrointestinal lymphomas. The ileum is the most common site affected by small intestine lymphoma, followed by the jejunum and duodenum. While intussusception is a very rare presentation of NHL, the most common lymphoma causing intussusception is diffuse large B-cell NHL.

We herein present a rare case of an ileocecal intussusception caused by primary intestinal NHL in a 62 year-old female patient.

**Materials and Methods**

In this paper, we present and discuss a new case of ileocecal intussusception caused by lymphoma. In addition, for this review, a search of the English medical language literature in PubMed and Google Scholar was conducted for every case report, series, letter to the editor, original article, and literature review related to gastrointestinal intussusception caused by lymphoma. Furthermore, reference lists of the articles obtained and previous reviews were examined. Key words used were intussusception, intussusception and lymphoma, intussusception due to lymphoma, intussusception due to Hodgkin’s lymphoma, lymphoma presenting as intussusception, intussusception due to Burkitt lymphoma, and intussusception due to Non-Hodgkin’s lymphoma. The search included all articles from 2000 to May 2011. The articles containing adequate information such as patient age, sex, localization of the tumor, diagnostic tools, surgical approach, type of lymphomas, follow-up, and oncologic management were included in this study, while studies and comment articles with insufficient clinical and demographic data were excluded. Patients younger than 16 years of age were excluded from the study because they are under the practice of pediatric surgery.

**Results**

**Case Report**

A 62 years-old female patient presented to our Clinic with complaints of abdominal pain, constipation, and fatigue. She said that she had these symptoms for 6 months, that they had worsened over time, and that she had lost 5 to 6 kg. No disease was detected in her medical background other than hypertension, and no abnormal findings were revealed in her physical and rectal examinations. Her liver and kidney function test results, tumor marker levels (CEA and CA 19-9), and complete urine test results were within normal limits. Her hemoglobin level was 10 g/dL (12.5-16), WBC level was 5.8 K/µL, and platelet count was 378,000 K/µL. No abnormal finding was detected on plain abdominothoracic X-rays. The patient underwent oral + intravenous contrast-enhanced abdominal computed tomography (CT). Two different masses were detected in the cecum and mesocele (Figure 1). Therefore, a colonoscopy was scheduled. A mass that completely filled the ileocecal valve and did not allow the colonoscope to enter was detected during colonoscopy. Nonspecific inflammation was noted in a pathology specimen taken via endoscopy. Laparotomy was performed because of the low hemoglobin level, weight loss, and masses on CT. During laparotomy, a solitary mass of 4 to 5 cm was detected in the mesocolon along with a mass leading to ileocolic invagination (Figure 2A, B). The patient underwent right hemicolecctomy and tip-to-side ileotransversotomy. She was discharged on the fifth postoperative day. A tumoral tissue of 6 × 3 cm invaginating toward the cecum and originating from the ileum 3 cm proximal to the ileocecal valve was cut and stained for histopathology. Tumor cells were stained by CD45 and CD20 diffusely, but CD3, CD30, PanCK, EMA, and AK were stained in the immunohistochemical survey. The Ki67 proliferation index was highly positive in the same cells. The diagnosis of anaplastic diffuse large B-cell NHL was established with regard to the afore-

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**Figure 1.** Contrast-enhanced computed tomography showing two homogenous soft tissue masses located both ileocecal region and mesenteric area (Lymphadenopathy).
Figure 2. Intraoperative images. The mass leading to ileocecal invagination (A) and appearance of the mass leading to invagination after the specimen was cut (B).

Discussion

Description and History

Intussusception is defined as a proximal segment of bowel (intussusceptum) that telescopes into the lumen of the adjacent distal segment (intussuscipiens)\(^2,7,33\). The first report of intussusception was made in 1674 by Barbette of Amsterdam. Intussusception or introsusception, as it was called then, was also detailed in 1789 by John Hunter. In 1871, Sir Jonathan Hutchinson was the first to successfully operate on a child with intussusception\(^1,39\).

Pathophysiologic Mechanism

The exact mechanism of intestinal intussusception is still unclear. However, it is believed that any lesion in the bowel wall or irritant within the lumen that alters normal peristaltic activity and that forms leading edges for an intussusceptum is able to initiate an invagination. Ingested food and subsequent peristaltic activity of the bowel produces an area of constriction above the stimulus and relaxation below, thus, telescoping the lead point through the distal bowel lumen. The most common locations are at the junctions between freely moving segments and retroperitoneally or adhesively fixed segments\(^2,3,32,35,36,39,40\).

Literature Review

The English medical literature published between 2000 and 2011 in the PubMed and Google Scholar databases was reviewed, and 33 reports concerning 36 cases of intussusception due to lymphoma meeting the aforementioned criteria were included in this review. The patients were aged from 16 to 86 years (mean, 48.2 ± 19.0 y). Twenty-nine were male (mean, 45.8 ± 18.1 y) and seven were female (mean, 58 ± 18.9 y). According to the localization of lymphoma, 24 patients had ileo-colic intussusception, 10 had enteric, and 2 had colic intussusception. In terms of the diagnosis, 34 patients were diagnosed with various types of NHL, and two patients were diagnosed with HL. The clinicopathologic characteristics of the 36 patients are summarized in Table I.
Table I. A summary of 36 cases of intussusception due to lymphoma reported in the literature from 2000 to 2011.

<table>
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<th>Reference</th>
<th>Year</th>
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Etiology and Location

Adult intussusception can be classified according to etiologic factors: primary or idiopathic and secondary. Primary or idiopathic adult intussusception accounts for about 8% to 20% of cases and is more likely to occur in the small intestine. Secondary intussusception, which is more commonly present in the adult population, is associated with a pathological condition involving a lead point. The causative lead point can be a benign polyp, lipoma, appendix, Meckel’s diverticulum, or a malignant tumor such as a primary or metastatic adenocarcinoma, GISTs, lymphoma, or carcinoid tumor34,40,41.

Adult intussusceptions have been classified into three major categories according to their gastrointestinal locations: enteroenteric, ileocolic or ileoileal, and colocolic41. Enteroenteric intussusception is confined to the small bowel. In ileocolic intussusceptions, the ileum invaginates through the ileocecal valve. Colocolic intussusception is confined to the colon90. In the results of our screening of over 14 papers in which 396 intussusception cases were presented, enteroenteric intussusception was detected in 203 cases (51.3%), ileocolic or ileoileal in 83 (20.9%), and colocolic in 110 (27.8%)2,7,23,31,37,41-44.

Adult intussusception occurs more frequently in the small bowel (50%-88%) than in the large bowel (12%-50%)16. In adults, it is important to differentiate between small bowel and colonic intussusception: in 30% of cases of small bowel intussusception, a malignant underlying lesion can be found, whereas in 30% to 68% of cases of large bowel intussusception, a malignant etiology is expected. Colon adenocarcinoma is the most important cause of malignant large bowel intussusception. Lymphoma, lymphosarcoma, and leiomyosarcoma have also been reported as tumoral masses inducing intestinal invagination. Benign lesions provoking large bowel intussusception include lipoma, leiomyoma, adenomatous polyp, and endometriosis, and up to 13% of colonic intussusception cases remain unexplained. Malignant causes of small bowel intussusception are predominantly metastases. Only rarely does a primary small bowel malignancy include adenocarcinoma, carcinoid, GIST, or lymphoma tumors. Nonmalignant lesions include benign tumors, Meckel’s diverticulum, inflammatory fibroid polyps, lymphoid hyperplasia, adhesions, intestinal tubes, jejunostomy feeding tubes, and trauma45. It was also concluded from the same literature research that of the 396 cases, intussusception developed secondary to benign and malignant causes in 252 (63.6%) and 144 (36.4%) cases, respectively. When we take a look at cases in which malignancy was detected, primary or secondary malignancy was detected in 50 (24.5%) of enterenteric cases, 34 (41%) of ileocolic cases, and 60 (54.5%) of colocolic cases. Furthermore, details of malignant tumors included 67 primary gastrointestinal adenocarcinomas, 15 GISTs, 23 lymphomas, 4 carcinoid tumors, 10 metastatic melanomas, 6 metastatic adenocarcinomas, 5 metastatic lung cancers, 5 leiomyosarcomas, 4 intestinal mesotheliomas, 2 NETs, 1 metastatic esophagus cancer, 1 stomach cancer, and 1 ileal Kaposi’s sarcoma2,7,23,31,37,41-46. These results are consistent with the fact that the risk of intussusception secondary to malignancy rises gradually from proximal to distal. An important finding is that lymphomas, which are rated third in terms of primary gastrointestinal malignancies, are rated second in terms of those leading to intussusception. However, because it is not clear in the literature whether a predisposing factor exists in cases with intussusception secondary to lymphoma, our interpretations are limited.

Because the main topic of this paper is intussusception and lymphoma, a discussion of the general features of gastrointestinal lymphomas follows.

Lymphoma is a type of cancer that affects lymph cells and tissues, including white blood cells, lymph nodes, and the spleen. It may arise anywhere outside of the lymph nodes and may develop in the gastrointestinal tract, including the stomach, ileum, and colon13. Lymphoma is divided into two major categories: HL and NHL. HL most commonly occurs in the cervical lymph nodes. The extranodal forms are rare and account for less than 1%. The most common site of involvement is the gastrointestinal tract, followed by the pulmonary system, thyroid, skin, genitourinary system, and central nervous system. To our knowledge, 14 patients with small bowel HL have been reported in the English-language literature since 19671,22,46. NHL is much more common than HL. The gastrointestinal tract is the most commonly involved site for extranodal NHL, accounting for 20% to 50% of all extranodal disease. The most common primary site is the stomach, followed by the small intestine, colon, and rarely other gastrointestinal organs. Gastrointestinal NHL represents between 1% and 4% of all gastrointestinal malignancies and 10% to 20% of small bowel ma-
Both of these malignancies may cause similar symptoms, but the conditions themselves differ. The distinction between HL and NHL is made upon histopathological examination of the cancerous material.

While HL develops from a specific abnormal B lymphocyte lineage, NHL may derive from either abnormal B or T cells. There are five subtypes of HL and about 30 subtypes of NHL. Because there are so many different subtypes of lymphoma, the classification of lymphoma is complicated. There are many classifications of gastrointestinal tract lymphoma. For simplicity, clinical staging is based on the Ann Arbor classification, and histopathologic staging is based on the World Health Organization (WHO) classification. According to this classification, lymphomas of the gastrointestinal tract generally fall into one of six categories: extranodal marginal zone mucosa-associated lymphoma tissue (MALT lymphoma), follicular lymphoma, mantle cell lymphoma, diffuse large B-cell lymphoma, and Burkitt’s lymphoma. Many of the NHL subtypes look similar, but they are functionally quite different and respond to different therapies with different probabilities of cure. HL subtypes are microscopically distinct, and typing is based upon the microscopic differences as well as extent of disease.

There are some well-known risk factors for gastrointestinal lymphomas. They include Helicobacter pylori infection, inflammatory bowel diseases, autoimmune disorders, immunodeficiency syndromes, immunosuppression, transplantation, celiac disease, and nodular lymphoid hyperplasia. In our literature search, five cases of intussusception that developed secondary to lymphoma had been infected with HIV and one case had undergone pancreas and kidney transplantation 11 years ago.

The most appropriate treatment modalities for primary gastrointestinal lymphomas are still controversial. Some Authors advocate that only surgical procedures performed with regard to oncological principles are sufficient, while others support that addition of chemotherapy to surgery increases survival. Generally, chemotherapy is recommended along with surgery in cases with poor prognostic factors, such as high LDH level, T-cell phenotype, extranodal involvement of ≥ 2, Ann Arbor stage of III to IV, age of > 60 years, and ECOG performance status of ≥ 2. High positivity rates of the Ki67 proliferation index immunochemically correlate with the aggressive course of a tumor. Thus, one must take this into consideration when scheduling a treatment approach. In summary, while surgical procedures alone performed consistently with oncological principles are enough in localized primary and in low-grade intestinal lymphomas, the most appropriate approach is to perform different chemotherapy protocols along with surgery in cases with the aforementioned poor prognostic factors. In secondary gastrointestinal lymphomas, chemotherapy should be performed first with regard to features of primary disease, followed by large or limited surgery with regard to the status of disease in the intestinal system. However, the size of the surgical procedure should be determined according to intraoperative findings in cases requiring urgent surgery because of signs and symptoms of intestinal obstruction.

Clinical Presentation

The clinical presentation of adult intussusception varies considerably. The presenting symptoms are nonspecific, and the majority of cases in adults have been reported as chronic, consistent with partial obstruction. Colicky abdominal pain (85%-100%) is the most common presenting symptom in patients with intussusception, followed by nausea (41%-75%), vomiting (35%-70%), bleeding (16.4%-27.3%), and diarrhea and constipation (22.5%-69%). In contrast to intussusceptions in children, palpation of an abdominal mass during clinical examination is reported in 9.1% to 62.5% of adult patients with intussusception. The most common age of presentation is around the fifth and sixth decades of life with a slight male preponderance. We analyzed 14 reports that we used as references in this study. A total of 396 patients ranging between 15 and 93 years of age (mean, 50.6 ± 18.1 y), 205 of whom were male and 191 of whom were female, are presented in this study. When we compare our results with our own literature analysis, intussusception cases secondary to lymphoma have a slightly younger mean age with a prominent male preponderance.

Diagnostic Methods

The preoperative diagnosis of adult intussusception is difficult because the clinical presentation is often nonspecific and the condition is rare. An accurate diagnosis is based on a good medical history, thorough physical examination, and...
specific imaging modalities, such as X-rays, ultrasound (US), CT, magnetic resonance imaging (MRI), enteroclysis, endoscopic procedures, diagnostic laparoscopy, scintigraphy, angiography, capsule endoscopy, and FDG-PET/CT. Typically, abdominal X-rays are the first diagnostic tool because obstructive symptoms dominate the clinical picture in most cases. A barium enema was the gold standard for diagnosing intussusception until the mid-1980s, when studies established that US could accurately diagnose the condition. Around the same time, the discovery was made that air could be used to both diagnose and treat intussusceptions. Enteroclysis is rarely used in the diagnosis of intussusception. This invasive double-contrast imaging method requires intubation using a special catheter with a double lumen and balloon to the proximal jejunum and is performed under fluoroscopy, MRI, or CT imaging. Enteroclysis shows not only the inside of the lumen, but also has a high sensitivity and specificity for revealing small and mucosal lesions. Colonoscopy is useful only in cases in which colonic involvement is strongly suspected, and it allows the lesion to be diagnosed and biopsied. Colonoscopically, intussusception is seen as an intraluminal mass directed centrally and distally. However, a diagnosis made by colonoscopy is rare, and most of the time the diagnosis is performed during surgery. Capsule endoscopy and digital balloon endoscopy are newer means of diagnosing various gastrointestinal disorders. Capsule endoscopy is a noninvasive diagnostic test used to locate the source of gastrointestinal bleeding and identify the cause of other intestinal disorders, including intussusception and various tumors. Intussusception in capsule endoscopy has been reported to appear as a mass lesion of the small bowel. Although obstructive symptoms are contradictory to capsule endoscopy, this new modality for the evaluation of the small bowel could be very helpful in cases with long-standing abdominal pain and negative radiologic examinations, either CT or barium studies, to exclude the possibility of malignancy. Double-balloon enteroscopy, also known as push-and-pull enteroscopy, can examine approximately 70 to 150 cm of the small bowel, and double balloon enteroscopy can examine the full length of the small bowel, both antegrade and retrograde. Diagnostic laparoscopy (DL) may assist in the diagnosis of intussusception in cases in which the diagnosis is suspected but not confirmed by preoperative workup. Moreover, DL can help to establish the cause and is less traumatic than exploratory laparotomy. US is considered to be a useful tool for the diagnosis of intussusception in both children and adults. Its classical imaging features include the target or doughnut sign in the transverse view and the pseudokidney, sandwich, or hayfork sign in the longitudinal view. However, obesity and the presence of massive air in the distended bowel loops limit the image quality and subsequent diagnostic accuracy. Overall, US has a sensitivity of 98% to 100% and a specificity of 88% for diagnosing intussusceptions. Abdominal CT is currently considered the most sensitive radiological method for confirming intussusception, with a reported diagnostic accuracy of 58% to 100%. On CT, a bowel-within-bowel configuration suggested by mesenteric fat and vessels compressed between the walls of the small bowel is pathognomonic. MRI is not applied routinely to diagnose intussusception in children or adults. This modality is reserved for evaluating select patients in whom an atypical sonographic appearance suggests a pathological lead point, such as lymphoma. Angiography and scintigraphic studies have shown diagnostic efficacy, but they are not routinely used. Despite the presence of several reports indicating an increase in FDG activity at the intussusception area detected via FDG-PET/CT, no study is available regarding its routine use. We conducted an analysis of over 396 patients showing that the percentage of establishing an intussusception diagnosis in the preoperative period varied between 30.7% and 83.3%. When we examined instruments used for verifying the diagnosis, the most successful results were achieved with laparoscopy (100%), followed by CT (57%-93.9%), colonoscopy (42.1%-45.4%), and barium enema (45.4%-73%).

**Approach to Adult Intussusception**

There is no universal approach to the management of adult intussusception. Most of the debate focuses on the issue of primary en bloc resection versus initial reduction followed by resection. Reduction by surgery before resection may theoretically permit a more limited resection. However, the risk of potential intraluminal seeding or venous tumor dissemination during the manipulation of a malignant lesion should be considered. The prevalence of malignancy as the cause of enterocentric intussusception is as high as 30%, and the vast majority are metastatic. For these reasons, most Authors recommend...
initial reduction of externally viable small bowel before resection\textsuperscript{36,39}. Conversely in the large bowel and ileocolic region, it is more likely that the intussusception will have a malignant etiology (up to 68\%). The majority of these lesions arise as a primary lesion, in which resection without reduction is recommended\textsuperscript{36,37,39}.

References


