

Adult Ileocolic Intussusception Due To Diffuse Large B-Cell Lymphoma: A Case Report.

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Abstract

Background: Adult intussusception constitutes 5% of all intussusceptions. Gastrointestinal non-Hodgkin lymphomas represent 1%-4% of all gastrointestinal malignancies. Diffuse large B-cell lymphoma is the most common lymphoma, while 40% of these tumors arise in extranodal sites.

Case presentation: Herein, we report a 77-year-old female who appeared in the emergency department with a two-day history of abdominal pain localized in the right lower quadrant, associated with nausea and lack of appetite. Computed tomography of the abdomen revealed an ileocolic intussusception; however, it did not detect a pathological lead point. A right hemicolectomy was performed. Histopathological report evinced the diagnosis of a diffuse large B-cell lymphoma.

Conclusion: Diffuse large B-cell lymphoma is the most common lymphoma causing intussusception. Neither surgery nor radiotherapy alone is preferred for patients with localized disease. The combination of surgery with chemotherapy is superior to any other treatment combination in localized disease.

Keywords: Intussusception; extranodal disease; diffuse large B-cell lymphoma; computed tomography; management.

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I. Introduction

Intussusception is defined as the invagination of one bowel segment (intussusceptum) into an adjacent part (intussusciptens). Adult intussusception constitutes 5% of all intussusceptions. The reported incidence is 2-3 cases/million/year. Bowel intussusception is divided based on the involvement segments into enteric, ileocolic, ileocecal, and colonic type (1,2). Enteric intussusception is the most frequent type (49.5%), followed by ileocolic (29.1%) and colonic intussusception (19.9%) (3).

Contrary to pediatric intussusception, adult intussusception is brought about by a pathological lead point in 90%, while the remaining 10% is idiopathic. Approximately two-thirds of the cases with a lead point are caused by benign or malignant neoplasms, while 50% of them are malignant (4, 5). Benign tumors are the commonest etiology of enteric intussusception, while malignant tumors are the most frequent cause of ileocolic and colonic intussusception (3).

Clinical diagnosis of intussusception is very challenging as the manifestation is variable and non-specific. Computed tomography is the gold standard for diagnosing intussusception with a reported sensitivity of 58%-100% and specificity of 57%-71% (6). A pathological lead point mainly causes adult intussusception; thus, surgical resection is indicated. Surgical exploration is recommended when symptoms and signs of bowel obstruction are present, a pathological lead point is identified on computed tomography, and colonic or ileocolic intussusception is diagnosed (2, 7).

Non-Hodgkin Lymphoma (NHL) is much more common than Hodgkin lymphoma (HL). Diffuse large B-cell lymphoma (DLBCL) is the most common lymphoma and composes 30%-58% of NHL. Approximately 40% of these tumors arise in extranodal sites. The stomach is the most typical site of gastrointestinal NHL, followed by the small intestine, colon, and rectum. As for the small intestine, the ileum is usually affected (60%-

65%), followed by the jejunum (20%-25%) and duodenum (8%) (6). The combination of surgery with chemotherapy is superior to any other treatment combination in localized disease (8). Here we describe the case of a 77-year-old female who was diagnosed with ileocolic intussusception and underwent right hemicolectomy. Histopathological report evinced the diagnosis of diffuse large B-cell lymphoma.

II. Case presentation

A 77-year-old female visited the emergency department with a two-day history of abdominal pain localized in the right lower quadrant. The pain was described as cramping, and it was associated with nausea and lack of appetite. Her medical history was unremarkable. Physical examination revealed a soft, non-distended abdomen, with tenderness in the right lower quadrant and no evidence of peritonism. Bowel sounds were normal, while digital examination disclosed the presence of soft stool.

Laboratory studies were within normal limits, and the Covid-19 test was negative. Abdominal x-ray disclosed three atypical air-fluid levels without evident bowel dilatation. Contrast-enhanced computed tomography of the abdomen was performed and showed an ileocolic intussusception and enlarged right paracolic nodes. However, it did not detect a pathological lead point.

The patient underwent an exploratory laparotomy as ileocolic intussusception is usually linked with malignancy. A right hemicolectomy with stapled side to side antiperistaltic anastomosis was carried out. The patient recovered uneventfully, and she was discharged on the seventh postoperative day.

The pathologic report showed an infiltrating tumor of all bowel layers located in the terminal ileum. The mass extended through the serosa in two adjacent lymph nodes. It consisted of medium to large cells, with eccentric nuclei and vesicular chromatin with few nucleoli, in diffuse sheets with confluent foci of large cells, but no prominent starry sky pattern, with a background of few admixed small lymphocytes. The immunohistochemistry findings revealed a B lineage origin (CD19, CD20, CD22, CD79a+) with a germinal center phenotype (Bcl6, CD10, CD38+), a high Ki-67 index (>40%), and few CD3 positive cells. CD5, CD30, Bcl2, and Cyclin D1 were negative. The diagnosis was compatible with a diffuse large B-cell lymphoma with a germinal center phenotype. Therefore, the patient was referred to the oncology unit for adjuvant chemotherapy.

III. Discussion

Intussusception is defined as the prolapse of one segment (intussusceptum) of the bowel into an adjacent part (intussusciptum). It is more frequent in children than adults, with an approximate ratio of 20:1 (1). In adults, intussusception is extremely rare, with a reported incidence of 2-3 cases/million/year (2). It constitutes 5% of all cases of intussusception and accounts for 1% of bowel obstruction. It is found in less than 1 in 1300 abdominal operations (1). Usually, it involves people after the fifth decade of life, with no gender predominance (1, 7).

The bowel intussusception is divided into four categories based on the involvement bowel segments:

- Enteric: The intussusception is confined to the small bowel.
- Ileocolic: An ileal segment invaginates into the colon via the ileocecal valve.
- Ileocecal: The ileocecal valve acts as the lead point.
- Colonic: The intussusception is confined to the large bowel (1, 2).

Hong et al. conducted a systematic review and meta-analysis of 40 retrospective case series that include 1229 adults with intussusception. Enteric intussusception is the most frequent type (49.5%), followed by ileocolic (29.1%) and colonic intussusception (19.9%) (3).

Contrary to pediatric intussusception, adult intussusception is brought about by a pathological lead point in 90%, while the remaining 10% is idiopathic. The pathological lead point can be in the lumen of the bowel, inside the wall, or extramural. Approximately two-thirds of the cases with a lead point are caused by benign or malignant neoplasms, while 50% of them are malignant. The remaining cases are brought about by postoperative adhesions, Crohn's disease, Meckel diverticulum, infections, and intestinal ulcers (4, 5, 9).

When dividing etiology by location, benign tumors are the commonest cause of enteric intussusception (39.4% vs 22.5%). The main type of malignant tumors is metastatic carcinoma (48.7%), followed by lymphoma (26.2%) and GIST (21.3%). On the contrary, malignant tumors are the most frequent etiology of ileocolic and colonic intussusception (36.9% vs 34.4%, and 46.5% vs 36.8%, respectively). The main type of malignant tumors in ileocolic and colonic intussusception is primary adenocarcinoma (61.7% and 78.8% respectively), followed by lymphoma (28.1% and 16.8% respectively) and metastatic carcinoma (13.4% and 14.4% respectively) (3).

Clinical diagnosis of intussusception is very challenging as the manifestation is variable and non-specific. Typically, patients develop symptoms and signs of bowel obstruction. The classical triad of abdominal pain, palpable mass, and bloody stool are rarely present. Pain is the most typical symptom. Associated symptoms encompass nausea, vomiting, bloating, failure to the gas passage, change in bowel habits, obstipation, constipation, and gastrointestinal bleeding (3, 7, 10). Wang et al. reported that cramping abdominal pain was

present in 80% of cases, whereas a palpable mass was found in <9% of patients (9). Symptoms are usually acute, lasting days to weeks. Rarely, they can be chronic, lasting years (7). Physical examination may reveal distention with tenderness ranging from mild to severe (consistent with peritoneal irritation). Other findings include decreased or absent bowel sounds, guaiac-positive stool, and a palpable mass (11).

Imaging modalities are the critical element for the diagnosis of intussusception. Although plain abdominal x-rays are helpful in detecting bowel obstruction and perforation, they are insensitive to the diagnosis of intussusception (12). Abdominal ultrasound displays a sensitivity of 98%-100% and a specificity of 88% (6). Nonetheless, its ability to detect lead points is limited. The classic feature is the target or doughnut sign in the transverse view and the pseudo-kidney sign in the longitudinal view. The target sign is brought about by the edematous intussusciens forming an external ring around the centrally based intussusceptum, while the layers of intussusception form a pseudo-kidney sign (12). Computed tomography is the gold standard for diagnosing intussusception with a reported sensitivity of 58%-100% and specificity of 57%-71% (6). It can detect intussusception, identify the pathologic lead point, highlight the location of the lead point and its relationship with the surrounding tissues, and diagnose bowel perforation or ischemia. The appearance of intussusception on computed tomography depends on the imaging plane. The characteristic finding is the bowel-within-bowel appearance, in which two concentric enhancing/hyperdense rings are present. This appearance is called target sign when imaged at right angles to the lumen and sausage-shaped soft tissue mass when imaged longitudinally. The two rings are formed by the outer intussusciens and the central intussusceptum. The mesentery (fat and vessels) will create a crescent of tissue around the compressed innermost lumen, surrounded by the two layers of the outer enveloping bowel. The lead point will be visualized more distally (1, 2, 7, 13).

A pathological lead point mainly causes adult intussusception; thus, surgical resection is indicated. Surgical exploration is recommended in the following situations:

- Intussusception with associated symptoms and signs of bowel obstruction.
- Intussusception with an identifiable pathological lead point on computed tomography.
- Colonic or ileocolic intussusception: They are usually associated with malignancy.

Some authors propose a wait and see policy with serial clinic and imaging assessment to ensure spontaneous resolution in entero-enteric intussusception without a lead point and short affected segment (<3.5 cm) (2, 7). Hong et al. reported that only 4.6% of patients were treated conservatively (3). When indicated, surgery might be performed open or laparoscopically, depending on the patient's condition and the skill and experience of the surgeon. Regardless of the surgical approach, a dilemma exists whether intraoperatively reduction or direct resection of the affected segments should be performed. When imaging studies, colonoscopy, or intraoperative appearance suggest the presence of malignancy, or the surgeon suspects ischemia that milking the intussusceptum out of the intussusciens might lead to perforation, the reduction is obviated. The entire segment should be resected en bloc (2, 7). Azar et al. reported that resection and creating an end colostomy (Hartmann procedure) for left-sided or rectosigmoid cases is a safe and efficient option, whereas resection with primary anastomosis for entero-enteric or right-sided cases should be performed (4).

Lymphoma is a type of cancer that affects lymph cells and tissues, including white blood cells, lymph nodes, and the spleen. It is divided into two categories: Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL). NHL is much more common than HL. Diffuse large B-cell lymphoma (DLBCL) is the most common lymphoma and composes 30%-58% of NHL. Although most of these tumors originate in lymph nodes, almost 40% develop in extranodal sites. The gastrointestinal tract is more frequently affected, followed by the pulmonary system, thyroid, skin, genitourinary system, and central nervous system. Gastrointestinal NHL constitutes 1%-4% of all gastrointestinal malignancies and 10%-20% of small bowel malignancies. The stomach is the most typical site of gastrointestinal NHL, followed by the small intestine, colon, and rectum. As for the small intestine, the ileum is usually affected (60%-65%), followed by the jejunum (20%-25%) and duodenum (8%) (6, 8, 14).

NHL has been correlated with the following conditions, drugs, and chemical agents:

- Infections: helicobacter pylori, human immunodeficiency virus (HIV), Epstein-Barr virus (EBV), hepatitis-C virus (HBV), human T-cell lymphotropic virus-1 (HTLV-1), campylobacter jejuni. inflammatory bowel disease, celiac disease, and immunosuppression
- Inflammatory bowel disease
- Coeliac disease
- Autoimmune disorders
- Hereditary immunodeficiency disorders
- Immunosuppression (6, 14, 15)

Neither surgery nor radiotherapy alone is preferred for patients with localized disease. Several studies demonstrated improved overall survival in patients who underwent surgery combined with chemotherapy, particularly those with early disease stages. The combination of surgery with chemotherapy is superior to any

other treatment combination in localized disease. Cyclophosphamide, doxorubicin, vincristine, and prednisone with or without rituximab are currently used. (8).

IV. Conclusion

Adult intussusception constitutes 5% of all intussusceptions. It is divided based on the involvement segments into enteric, ileocolic, ileocecal, and colonic types. Enteric intussusception is the most frequent type (49.5%), followed by ileocolic (29.1%) and colonic intussusception (19.9%). Contrary to pediatric intussusception, adult intussusception is brought about by a pathological lead point in 90%, while the remaining 10% is idiopathic. Computed tomography is considered the gold standard for diagnosing intussusception with a reported sensitivity of 58%-100% and specificity of 57%-71%. Surgical exploration is recommended when symptoms and signs of bowel obstruction are present, a pathological lead point is identified on computed tomography, and colonic or ileocolic intussusception is diagnosed.

Diffuse large B-cell lymphoma (DLBCL) is the most common lymphoma and composes 30%-58% of NHL. Approximately 40% of these tumors arise in extranodal sites. The stomach is the most typical site of gastrointestinal NHL, followed by the small intestine, colon, and rectum. As for the small intestine, the ileum is usually affected (60%-65%), followed by the jejunum (20%-25%) and duodenum (8%). Surgery combined with chemotherapy is superior to any other treatment combination in localized disease.

Abbreviations:

HL: Hodgkin Lymphoma

NHL: Non-Hodgkin Lymphoma

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5. Kefala MA: Analysis and interpretation of data.
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