

ILEAL DIFFUSE LARGE B CELL LYMPHOMA AS A LEAD POINT FOR AN ADULT ILEO-ILEO-COECCAL INTUSSUSCEPTION

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ABSTRACT

Intussusception is defined as an invagination of one segment of the gastrointestinal tract and its mesentery (intussusceptum) into the lumen of an adjacent distal segment of the gastrointestinal tract (intussusciens). Adult intussusception is a rare condition which can occur in any site of gastrointestinal tract from the stomach to the rectum. More than 80% of the cases of adult intussusception are reported to be caused by neoplasms. Primary malignant lesions (carcinoma and lymphoma) are the most common underlying malignant lesions. In adults, intussusception typically manifests as an acute or chronic obstruction and its presentation is similar to that of bowel obstruction. Diagnosis may be delayed because of its longstanding, intermittent, and non-specific symptoms.

A case of 40-year old male patient with abdominal discomfort, distension, pain, diarrhea and occasional constipation in the few weeks' period prior to admission on our department is presented. Gastrografen contrast-enhanced imaging bowel investigation was performed and it showed a narrowed segment of the terminal ileum with an enlarged segment proximally, 20-30cm in length. This finding was an indication for an abdominal CT scan and an ileo-colonic invagination was detected. The terminal ileal segment was invaginated to the cecum and ascending colon. The circulation of the invaginated segment was not compromised. As soon as the diagnosis was made, a laparotomy was performed. The intraoperative finding was an ileo-ileo-coecal invagination with an ileal tumor lesion and local fibrinopurulent peritonitis in the site of the invagination, accompanied with uncomplicated Meckel's diverticulum. The ileo-coecal invagination was spontaneously reduced and a resection of the terminal ileum including the Meckel's diverticulum with ileo-coecal latero-lateral anastomosis was performed. The resected segment was subject to a pathohistological analysis. The pathohistological finding was a diffuse large B cell lymphoma of the terminal ileum.

Keywords: ileo-ileo-coecal intussusception, B cell lymphoma, Meckel's diverticulum

INTRODUCTION

Intussusception is defined as an invagination of one segment of the gastrointestinal tract and its mesentery (intussusceptum) into the lumen of an adjacent distal segment of the gastrointestinal tract (intussusciens).¹ Adult intussusception is a rare condition which can occur in any site of gastrointestinal tract from stomach to rectum.¹

Approximately 5% of all intussusceptions occur in

adults, accounting for 1% of all bowel obstructions. Intussusceptions are classified according to their location (enteroenteric, ileocolic, ileocecal, or colocolic) and cause (benign, malignant, or idiopathic).² Unlike children, where most cases are idiopathic, intussusception in adults has an identifiable etiology in 80- 90% of cases.¹ More than 80% cases of adult intussusception are reported to be neoplasms.^{3,4} Primary malignant lesions

(carcinoma and lymphoma) are the most common underlying malignant lesions.^{3,5,6}

The spectrum of clinical presentations depends on the site of the intussusception, the timing of the clinical presentation, and the predilection for spontaneous reduction.¹ In adults, intussusception typically manifests as an acute or chronic obstruction and its presentation is similar to that of small and large bowel obstruction.^{1,7,8} Unlike intussusception in children, an acute abdomen is very often present in adults. The most common symptom in the acute presentation is abdominal pain (71-100%), intermittent abdominal pain and vomiting (40-60% of the cases) and bleeding per rectum (8-27% of the cases).^{1,5}

Diagnosis can be delayed because of its longstanding, intermittent, and non-specific symptoms and most cases are diagnosed during an emergency laparotomy.⁹ Several procedures are used in diagnosing an intussusception.

Plain abdominal film report signs of acute intestinal obstruction (air-fluid levels) with average sensitivity about 25%.¹

Barium enema signs of intussusception include a spiral, “coil spring” or “stacked coin” appearance with narrowed central canal. These signs result from the retrograde filling of the contrast between the walls of the invaginated bowel loop. The narrowed central canal is the edematous, obstructing intussusceptum. Barium enema remains limited to the ileocolic or colonic lesions.¹

Ultrasonography is characterized with a “target and doughnut” sign (an even thickened hypoechoic outer and a central hyperechoic core on transverse view), a “crescent-in-doughnut” sign (an even outer hypoechoic rim with a central hyperechoic crescent) or a “multiple concentric rings” sign (a mass with multiple alternating hypoechoic and hyperechoic concentric rings).¹

Color Doppler may be helpful in determining the degree of vascular compromise of the involved segments. ^{1,10}

Abdominal CT is the most useful diagnostic tool, not only for detecting an intussusception, but it also helps in identifying the underlying cause. While the appearance of intussusception is characteristic on a CT scan, its etiology usually cannot be established. ^{5,8,12}

After the bowel intussusception is diagnosed, it should be treated adequately. There is no universal agreement upon

the correct treatment of adult intussusception, although surgical intervention is considered to be necessary.^{3,11} The type of intervention depends on the patient's medical history and intraoperative findings.^{5,11} All patients with enteric lesions who have not had a previous laparotomy should undergo a resection without reduction, because of the high incidence of associated malignancy and should be treated by oncological principles.^{1,13} Primary resection without reduction should also be performed due to theoretical risks of perforation and seeding of the bowel's microorganisms or tumor to the peritoneal cavity and/or venous embolization.¹⁰

CASE REPORT

Our patient was a 40-year old male complaining of abdominal discomfort, distension, pain, diarrhea and occasional constipation during the few weeks' period prior to admission to our department. The onset of medical condition started with epigastric pain. A gastroscopy was performed, which showed an inflamed gastric mucosa, and the H. pylori test was positive. Oral diet and treatment for H. pylori infection was recommended. The suggested therapy did not resolve the condition, on the contrary, a week before the admission, the patient had symptoms of bowel obstruction presented with vomiting, absence of gases and stool. The condition was partly resolved with an intravenous rehydration and bowel peristaltic stimulation. Two days prior the admission, a gastrografin contrast-enhanced imaging bowel investigation was performed, which showed a narrowed segment of the terminal ileum with an enlarged segment proximally, 20-30cm in length. Despite the narrowing, the contrast passed through the segment into the colon. (Fig.1)

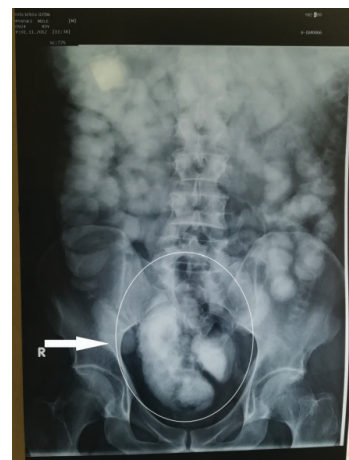


Fig.1: Gastrografin contrast-enhanced imaging bowel investigation

This finding was an indication for an abdominal CT scan. A day before the admission, a contrast-enhanced CT scan was performed and an ileo-colic invagination was detected. The terminal ileal segment was invaginated to the cecum and ascending colon. The vascularization of the invaginated segment was not compromised. (Fig. 2-4)

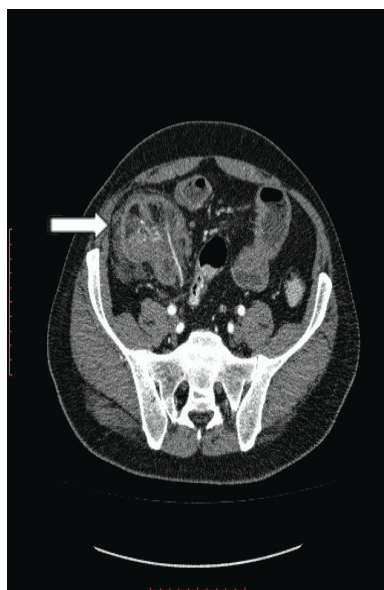


Fig.2: Transverse plane



Fig.3: Coronal plane

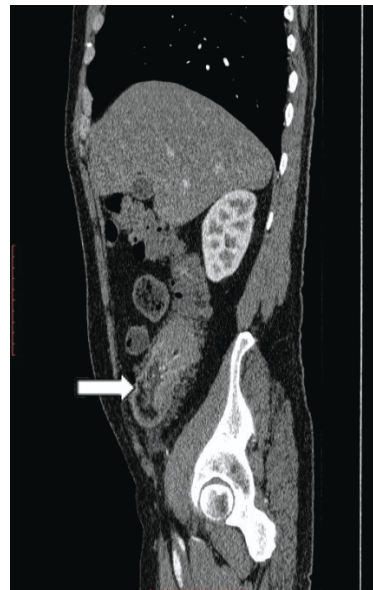


Fig.4: Sagittal plane

A laparotomy was performed and the intraoperative finding was an ileo-ileo-coecal invagination with ileal tumor lesion and a local fibrinopurulent peritonitis of the site of the invagination. (Fig. 5, 6) A secondary finding was an uncomplicated Meckel's diverticulum. (Fig. 7)

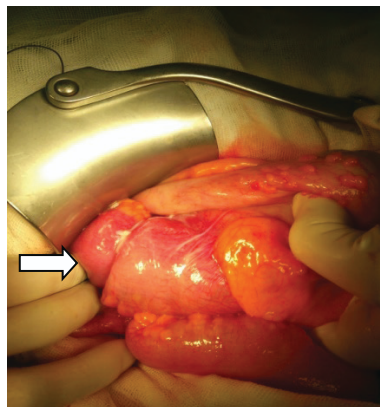


Fig.5: Ileo-ileo-coecal invagination

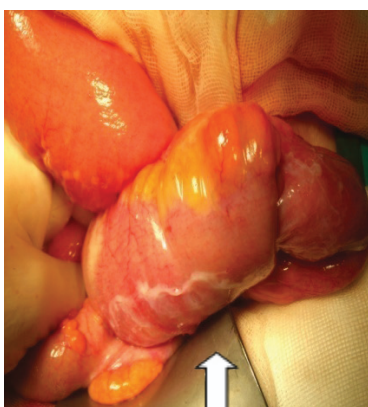


Fig.6: Local fibrinopurulent peritonitis

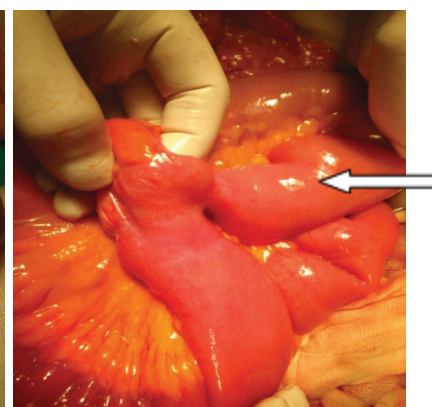


Fig.7: Meckel's diverticulum

The ileo-colic invagination reduced spontaneously and resection of the terminal ileum including the Meckel's diverticulum with ileo-coecal latero-lateral anastomosis was performed. The resected segment was a subject to pathohistological analysis. (Fig. 8, 9) The pathohistological finding was a diffuse large B cell lymphoma of the terminal ileum.

After the surgery, the patient was suggested to consult a hematologist for appropriate chemotherapy. Prior to the chemotherapy treatment, a whole-body PET scan was performed. There were no signs for metastases. (Fig. 10) The 5 years follow up period was uneventful



Fig.8: Resected bowel segment

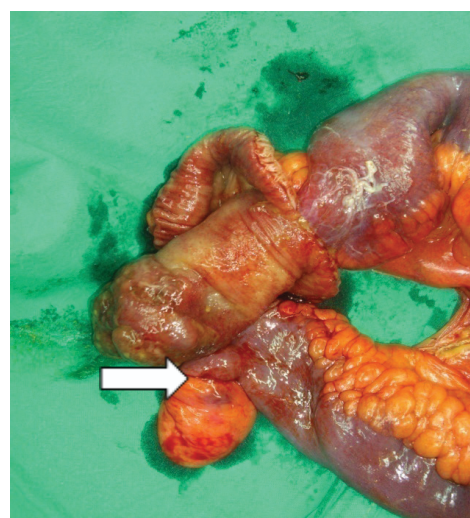


Fig.9: Ileal tumor lesiona

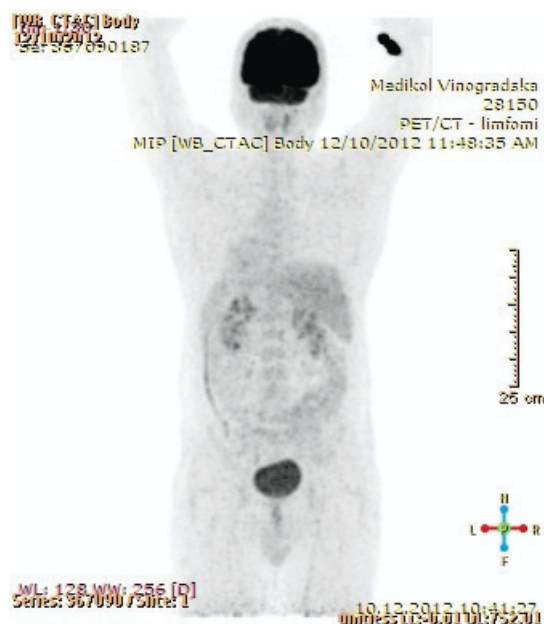


Fig. 10: PET scan

DISCUSSION

Adult intussusception is a rare condition which can occur in any site of gastrointestinal tract from stomach to rectum.¹ Intussusception commonly occurs at the junctions between freely-moving segments and retroperitoneally or adhesionaly fixed segments.¹⁰ More than 80% of the cases of adult intussusception are reported to be neoplasms. ³ Malignant causes of small bowel intussusception include primary leiomyosarcomas, malignant gastrointestinal stromal tumors, carcinoid tumors, neuroendocrine tumors and lymphomas.¹ Non-Hodgkin lymphomas of the ileum can be the leading point

for ileo-colic intussusceptions, such as in our presented case.^{7,14,15} Carcinoma of the appendix is a rare etiological factor for small bowel intussusception.

Diagnosis can be delayed because of its longstanding, intermittent, and non-specific symptoms and in most cases are diagnosed during an emergency laparotomy. Several procedures are used to set the diagnosis of intussusception. Abdominal CT is the most useful diagnostic tool, not only for detecting an intussusception, but it also helps in identifying the underlying cause.^{5,8}

Intussusceptions seen on CT that had a neoplastic lead point are significantly longer and have a significantly larger diameter than non-neoplastic ones.^{5,8,12} Proximal dilatation of the small bowel is significantly more common in intussusceptions with a neoplastic lead point, which correlates with the finding during our gastrografen contrast investigation.

After setting the diagnosis of adult intussusceptions, primary resection without reduction should be the method of choice for treatment of these patients. Bowel resection with oncological principles should be followed in every case where a malignancy is suspected. The theoretical risks of preliminary manipulation and reduction of an intussuscepted bowel include: (1) intraluminal seeding and venous tumor dissemination, (2) perforation and seeding of microorganisms and tumor cells to the peritoneal cavity and (3) increased risk of anastomotic complications of the manipulated friable and edematous bowel tissue.^{7,16} Reduction of the intussuscepted bowel is considered safe for benign lesions in order to limit the extent of resection or to avoid the short bowel syndrome

in certain circumstances.^{7,17,18}

In the presented case, the intraoperative finding was an ileo-ileo-coeccal invagination with ileal tumor lesion and local fibrinopurulent peritonitis in the site of the invagination. According to the basic principles, ileo-colonic invagination was spontaneously reduced and resection of the terminal ileum including the uncomplicated Meckel's diverticulum with ileo-coeccal latero-lateral anastomosis was performed. The patohistological finding was diffuse large B cell lymphoma of the terminal ileum, which belongs to the group of Non-Hodgkin's lymphomas of the ileum. They are reported as a most common malignant leading points for ileo-colic intussusceptions.^{3,5}

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