Small Bowel Resection and Anastomosis with Intraoperative Small Bowel Endoscopic Snare Polypectomy for Combined Ileal and Jejunal Myoepithelial Hamartoma Presented with Ileo-Ileal Intussusception in Adult Female Patient: Case Report and Literature Review

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Abstract

Background: Intussusception in adult is rare condition account for 5% of all intussusception and 1% to 5% of mechanical intestinal obstruction. Adult intussusception usually developed due to presence of organic lesion that acts as lead point. The clinical presentation of intussusception in adult is non-specific and most patients present with intermittent or chronic presentation.

Case Presentation: In our case report, we present a case of 27-year-old female patient who presented at our emergency department with colicky abdominal pain associated with vomiting and constipation. His medical and family histories were clear. No history of previous abdominal operation. Abdominal Computed Tomography (CT) revealed large ileo-ileal intussusceptions, the patient was resuscitated with IV fluid and shifted to operating room for exploratory laparotomy and she underwent small bowel resection with endoscopic snare polypectomy for jejunal polyp. Histopathology showed myoepithelial hamartoma.

Conclusion: Adult intussusception is rare cause of abdominal pain and should be considered in the differential diagnosis of acute abdomen. Management is surgical and delay diagnosis can lead to serious complications such as intestinal obstruction and ischemia.

Keywords: Adult intussusceptions; Myoepithelial hamartoma; Intestinal obstruction

Introduction

Intussusception is defined as the process in which a segment of the gastrointestinal invaginates into another neighboring segment leading to small bowel obstruction. This case is common among children; nevertheless, it is very rare among adults. The majority of adult cases involving the small bowel and have an evident point as the cause [1]. In the small intestine, the benign causes of adult intussusception e.g. lipoma, adenomatous polyp, and hemangioma are considered the major causes and more common compared to the malignant causes: lymphoma, adenocarcinoma, and melanoma.

Until recent the most common diagnosis ways of adult small bowel intussusception are radiology; however, endoscopy can be used effectively [2]. The best way to consider cases of severe obstruction is surgical intervention [3]. In this report, we describe a case of small bowel polyps complicated with large ileo-ileal intussusceptions and severe high grade decompensated small bowel obstruction.
A 27-year-old lady presented herself at the Emergency Room (ER) in a healthy state with no medical history. She had a complaint of colicky abdominal pain for the last 4 days with no localization associated with melena. The patient also had distension vomiting for 2 days. She denied having experienced any weight loss or alteration of bowel habits. She had no family history of cancer or chronic diseases.

Upon the pre-admission physical examination, she appeared to have an average body build, and she looked in severe pain. She had a blood pressure of 130/90 mmHg, a temperature of 36.9°C, heart pulse rate of 102 beat per minute, and a respiration rate of 20 per minute. During the abdominal examination, there was a clear hyperactive bowel sound, and her abdomen was grossly distended. There was a mild tenderness all over the abdomen. There was no guarding, rigidity, or hernias.

The laboratory examination showed that her White Blood Cells (WBCs) were 20 × 10^9/L, her hemoglobin was 8.5 g/dL, and her platelets were 462 × 10^9/L. The total serum sodium was 133 mmol/L, and potassium was 3.3 mmol/L. The Prothrombin Time (PT) was 13.4 sec, the Partial Thromboplastin Time (PTT) was 26.6 sec, and the international normalized ratio was 0.97. The total serum bilirubin was 6 µmol/L, and the direct bilirubin was 2.8 µmol/L. Alkaline phosphatase was 68 IU/L, Aspartate transaminase was 24 IU/L, Alanine transaminase was 16 IU/L, Amylase was 43 IU/L, and Lipase was 8 IU/L. The Blood urea nitrogen was 6.4 mg/dL, and the Serum creatinine was 54.8 µmol/L. Abdominal Computed Tomography (CT) showed a large ileo-ileal intussusception 15 cm with signs of ischemia (Figure 1). There was a clear polyp inside the intussusception complex with a severe high grade decompensated small bowel obstruction. Moreover, there was another small bowel polyp noted in the jejunum (Figure 2).

A collapsed colon was noted. Based on the findings of CT, the patient was diagnosed to have small bowel obstruction as a result of intussusception, and the surgical intervention decision was made.

Thereafter, the patient was resuscitated with Intravenous (IV) fluid. To prepare her for the surgical operation, nasogastric tube suction was performed to remove all solids, liquids, and gases from the stomach. A Foley catheter tube and prophylactic antibiotics started. Exploratory laparotomy was done through a midline incision.

During the laparotomy, a markedly distended small bowel proximal to the intussuscepted segment was found. This intussuscepted segment was about 15 cm with an area of patchy ischemia (Figure 3). A small bowel resection performed using Gastrointestinal Anastomosis (GIA) stapler. After resection, small enterotomy done for deflation of small bowel and insertion of the intraoperative endoscopy (Figure 4) for snare resection of the jejunal polyp (Figure 5).

Gross pathology of the resected part of ileum showed telescoping of the proximal ileal segment into the distal segment with thickened congested with full thickness mural necrosis and intraluminal polyp (Figure 6). Histological examination of the leading point (intraluminal
Mohamed S Essa, et al., ileal polyp and jejunal polyp showed multiple cystic glands of different sizes associated with bundles of proliferating smooth muscle cells mainly in submucosa (Figures 7-9). These histopathological findings were consistent with Myoepithelial Hamartoma (MEH). The patient’s postoperative course was smooth without any complications and she was discharged on the seventh day in good condition with follow up in outpatient clinic. After one month the patient came back for upper and lower gastrointestinal endoscopy to rule out other gastrointestinal polyps which was negative (Figure 10 and 11).

**Literature Review**

Bowel intussusception is the invagination of the proximal bowel segment (intussusceptum) inside the lumen of nearby segments (intussuscipiens) [4]. Barbette of Amsterdam is the first one who reported intussusceptions in 1674 followed by John Hunter who described the detailed report as “intussusception” in 1789 [5,6]. Bowel intussusception in adults is extremely rare; it accounts for 1% to 5% of mechanical bowel obstruction and 5% of all intussusceptions [4,7]. It also represents 0.1% of adult hospital admissions [8].

Intussusception in an adult is different than in the pediatric age group in many aspects. In adults in about 90% of the cases are developed due to pathological lesions that act as leading points such as carcinomas, Meckel’s diverticulum, benign neoplasm, or polyps, most of these lesions detected intraoperatively. In contrast, intussusceptions in children are usually primary without any pathological lesion [9-11]. The management is also different, in children pneumatic or hydrostatic reduction is the treatment of choice with a success rate approximately 70% to 85% [12,13], while in adult surgery is the definitive management in the form of surgical resection because most are due to pathological lesion and also because of high incidence of malignancy which accounts for approximately 65%, so hydrostatic reduction is absolutely contraindicated in adult. Therefore, 70% to 90% of adult patients need surgical treatment, which is the recommended treatment [14-16]. In our case, the pathological lesion detected on Computed Tomography of the abdomen (CT).

The mechanism of intussusception in adults either primary (idiopathic) which accounted for 8% to 20% and developed most commonly in small bowel or secondary due to an organic lesion in the wall or inside the lumen of the bowel which acts as the lead point that affects normal intestinal peristalsis. Abnormal peristalsis results in an invagination of the proximal bowel segment in the distal segment leading to intussusceptions. This invagination leads to intestinal obstruction and more seriously, affecting the blood supply of the intussuscepted segment result in intestinal ischemia [7,16-18].

Adult intussusceptions are classified according to location and etiologies, regarding gastrointestinal location intussusceptions are classified into four groups:

1. Small bowel (entero-enteric)
2. Large bowel (colo-colic)
3. Ileocolic (invagination of terminal ileum into ascending colon)
4. Ileocecral (in which the ileocecal valve is the leading point) [9,14,19].

From the etiological point of view, the intussusceptions classified into benign, malignant or idiopathic causes. Small bowel intussusception is usually due to intraluminal or extraluminal pathologic lesion such as Meckel’s diverticulum, adenomatous polyps, inflammatory lesions, lipoma, metastases, and lymphoma. Small bowel adenocarcinoma represents 30% of small bowel intussusceptions in adults. Latrogenic intussusception is commonly occurred after insertion of the intestinal tube or after gastrojejunostomy for gastric bypass surgery (retrograde intussusception) [16,20,21]. In contrast large bowel intussusception most commonly due to malignant cause account for 66% [16,18,22]. Marinis A, et al. [4] reported rare cases, 29 years male patient presented with ileocolic intussusception due to terminal ileum diffuse small B-cell non-Hodgkin lymphoma [4].

Study of 745 patients presented with adult intussusceptions
Mohamed S Essa, et al.,

diagnosed intraoperatively, 52% were small bowel intussusceptions (13% ileo-colic, 39% entero-enteric), and 38% were large bowel intussusceptions (4% appendiceal, 17% ileo-cecal, 17% colo-colic) [23].

Small bowel tumors account for 3% to 6% of all tumors that arise in the gastrointestinal tract, and hamartoma represents 1.5% to 4.5% of these tumors [24,25]. Benign tumors of the small bowel rarely symptomatic because of the fluid content and dispensability of the small intestine. Most small bowel benign tumors discovered accidentally during surgery for other causes; therefore, the diagnosis of these tumors preoperatively is challenging. If these tumors become symptomatic, the most common symptom is abdominal pain secondary to intestinal obstruction, followed by hemorrhage [24]. Our patient presented with colicky abdominal pain associated with melena. Hemangiomas and leiomyomas are the most common small intestinal benign tumors causing gastrointestinal bleeding in contrast to other lesions in which the bleeding is rare [26].

The classic triad of pediatric intussusception (abdominal pain, tender palpable lump, and bloody diarrhea) is extremely rare in adults. The presenting symptoms of adult intussusceptions are variable and, including bowel habit changes, constipation, abdominal distension, nausea, vomiting, and gastrointestinal bleeding [7,9,14,27]. Transient intussusceptions are non obstructing intussusceptions occur without any pathological lesion and frequently described in patients with celiac disease [28] or Crohn’s disease [29] and disappear spontaneously without any intervention. In contrast to patients with an organic lesion, the main line of treatment is surgical intervention as in our patient.

The difference in clinical manifestation and radiological findings of adult intussusceptions make the diagnosis difficult preoperatively. Eisen LK, et al. [22] documented that the rate of preoperative diagnosis of adult intussusception 40.7%, while Reijnen HA, et al. [30] reported a higher rate of preoperative diagnosis approximately 50%. An abdominal X-ray is the initial diagnostic test because intestinal obstruction prevails in the clinical picture. Such imaging usually detects evidence of bowel obstruction (air-fluid level) and also gives an idea about the site of obstruction [22,31,32]. Findings on Gastrointestinal contrast studies differ according to anatomical location of intussusceptions, in small bowel intussusception it may show a “stacked coin” or “coiled spring” appearance, while in colo-colic or ileocolic intussusception barium enema may show a “cup-shaped” filling defect or “spiral” or “coiled-spring” appearance [22,33,34].

Abdominal ultrasonography is an effective method for the diagnosis of intussusception in adults and children. Findings on ultrasonography varied depending on the view, in the transverse view it may show “pseudo-kidney” while in a longitudinal view “hay-fork” sign is the main finding. Abdominal ultrasonography is operator dependent. Therefore, it requires skilled radiologist for diagnosis confirmation in addition to obesity and massive air due to abdominal distension decrease diagnostic precision [34-36].

Computed Tomography (CT) of the abdomen is the most sensitive radiological investigation for diagnosis confirmation with diagnostic accuracy ranging from 58% to 100% [7,18,37-41]. The pathognomonic findings on CT are heterogeneous soft tissue mass with layering effect (sausage or target-shaped mass) in addition to the typical results of mesenteric blood vessels inside intestinal lumen [17]. CT can identify the location, nature of the lesion, its relation to...
surrounding structures and staging of the malignant lesion, causing intussusception [22].

Kim YH, et al. [42] reported that abdominal CT could differentiate between intussusceptions with a pathological lesion (with a lead point) from that without pathological lesion (without a lead point). For intussusceptions with lead point the following findings may be observed:

1. Intestinal wall edema,
2. Signs of intestinal obstruction
3. Evidence of bowel ischemia that result in absence of three-layer pattern of bowel wall, and
4. Organic lesion while in intussusception without lead point the previous findings are absent [42].

Management of adult intussusception is mainly surgical because of a high rate of underlying anomalies and malignancy. In contrast to pediatric intussusceptions reduction with barium or air is contraindicated because reduction may lead to the following:

a) Seeding of tumor within the lumen,

b) Vascular dissemination of tumor,
c) Intestinal perforation and spread of microorganisms and neoplastic cells within the peritoneal cavity,
d) High risk of anastomotic complications because of manipulation on fragile and thickened edematous intestinal wall [7,9,16,22,30,43].

The reduction is contraindicated if there is evidence of intestinal ischemia [41]. So patients with ileocecal, ileocolic and colo-colic intussusception particularly those older than 60 years

<table>
<thead>
<tr>
<th>Case</th>
<th>Author</th>
<th>Age</th>
<th>Sex</th>
<th>Symptom</th>
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<th>Intussusception</th>
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<td>26</td>
<td>Our Case</td>
<td>27</td>
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<td>Jejunum and ileum</td>
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<td>MEH</td>
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oncological resection with anastomosis between viable, healthy bowel are mandatory because of a high incidence of malignancy [14, 16,22,30,44,45]. For right-sided colonic intussusceptions, primary resection with ileocolic anastomosis can be done without colonic preparation while intussusceptions on the left side of the colon and rectosigmoid intussusception resection with Hartman’s procedure is better particularly in emergencies as documented by Azar et al. [7].

Reduction can be made in the diagnosed following scenarios:

1. When benign intestinal lesion diagnosed preoperatively.
2. Patients with high risk of short bowel syndrome as in Peutz-Jeghers syndrome, in this case, reduction combined with limited resection with intraoperative endoscopy for polyps excision is preferred, and
3. Postoperative bowel obstruction due to intussusceptions as no recurrence is not present in patients presented with small bowel intussusceptions because of benign lesions treated with reduction and limited resection [7,45-47].

Laparoscopic management of bowel intussusceptions because of benign or malignant causes is feasible based on the general condition of patient and availability of expertise in laparoscopic surgery [48,49].

We have found 26 cases of small bowel hamartoma in both adult (14 patient) and pediatrics (12 patient) (Table 1) [50-71]. Most cases are solitary either in ileum or jejunum except our case in which the hamartoma present in both ileum and jejunum. Fourteen of which caused intussusceptions, eleven of those patients appeared in the ileum and three cases in the jejunum. Twelve of the 26 cases did not cause intussusception. Ten cases presented with gastrointestinal bleeding, either occult bleeding (anemia) or overt bleeding. One case present with difficult defecation. Two cases were asymptomatic.

Most of the cases causing intussusceptions were children (under the age of 18 years old) except two patients were adults (18 and 27 years old) and one patient elderly (82 years old). So we suggest that small bowel hamartoma induced intussusceptions appears mostly in children because hamartoma is congenital abnormality and the lesion rarely increase in size over time. Histopathology was different from 21 cases were myoepithelial hamartoma of intestinal type, three cases had vascular and neuromuscular hamartoma, one patient had adenomyoma and one patient had Peutz-jeghers polyp with malignant transformation [71].

Diagnosis of hamartoma based on the presence of developmental malformation as described by Willis [72]. In our case, the histopathological features are dilated cystic glands with proliferation lined by columnar epithelium with smooth muscle hypertrophy. Gastrointestinal hamartomas are classified by Dawson according to the developmental malformation as described by Willis [72]. In our case, the histological finding was cystic gland proliferation with hypertrophy of smooth muscle fibers that raise the possibility of Peutz-jeghers polyp. The pathognomonic histopathological features of this polyp are muscularis mucosa proliferation with the formation of the ramifying pattern [73,74] which was not present in our patient in addition to no family history of gastrointestinal polyps and no evidence of mucocutaneous pigmentation on examination.

Conclusion
This case report provides useful information about a rare case of adult intussusception and small bowel obstruction as a result of hamartomatous polyposis. The surgical intervention was a demand to help the patient overcome this disease.

Authors’ Contribution

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