Adult Intussusception Secondary to Lymphangioma of the Cecum: A Case Report

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We report the case of a patient with ileocolic intussusception caused by cecal lymphangioma. A 45-year-old man visited our hospital with a 2-month history of frequent episodes of watery diarrhea (≥5 times/day) and intermittent abdominal pain. A cecal submucosal tumor with mucosal ulceration and partial obstruction of the colonic lumen was identified by colonoscopy. He was admitted to our hospital 2 days later due to aggravation of his abdominal pain. Physical examination revealed tenderness over the right abdomen with no peritoneal signs. A double-contrast lower gastrointestinal series showed a right-side colonic lesion with indentation and a peripheral, beak-like sign. Abdominal computed tomography scanning revealed an intra-abdominal mass with the characteristic sausage sign, highly suggestive of intussusception. The patient subsequently underwent right hemicolectomy. The final diagnosis was ileocolic intussusception with cecal lymphangioma, which was confirmed by histopathology. He had an uneventful recovery with follow-up in our hospital. This case highlights the possibility of colonic lymphangioma as the leading point of adult intussusception, and this should be taken into consideration as a possible diagnosis in this uncommon clinical condition.

Key Words: cecum, intussusception, lymphangioma
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Case Presentation

A 45-year-old man visited our hospital owing to a 2-month history of frequent episodes of watery diarrhea and intermittent abdominal pain. He had previously been healthy, with no specific underlying disease. Colonoscopic examination revealed a cecal submucosal tumor with mucosal ulceration and partial obstruction of the colonic lumen (Figure 1). He was admitted to our hospital 2 days later owing to aggravation of his abdominal pain after meals.

At the time of his visit, his blood pressure was 116/78 mm Hg, pulse rate 98 beats per minute, and body temperature 36.7°C. Physical examination revealed tenderness in the right abdomen with no peritoneal signs. Laboratory tests showed no evidence of leukocytosis, with a normal C-reactive protein level (<5 mg/L) and a white blood cell count of 8,440/μL. A subsequent abdominal computed tomography (CT) scan showed an intra-abdominal mass over the right upper abdomen, with the characteristic sausage sign (Figure 2A). A double-contrast lower gastrointestinal series showed an obstruction of the right-side colon by a gas-containing lesion with a peripheral beak-like sign (Figures 2B and 2C).

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Laparotomy was performed for suspected intussusception. One huge, right-sided colonic mass was identified during the operation (Figure 3A) and right hemicolectomy with end-to-side ileocolostomy was performed. The final diagnosis of ileocolic intussusception with cecal lymphangioma was confirmed by histopathology (Figures 3B and 3C).

The patient made an uneventful recovery 21 months after the operation, and has continued to attend follow-up sessions in our clinic without the detection of any complications.

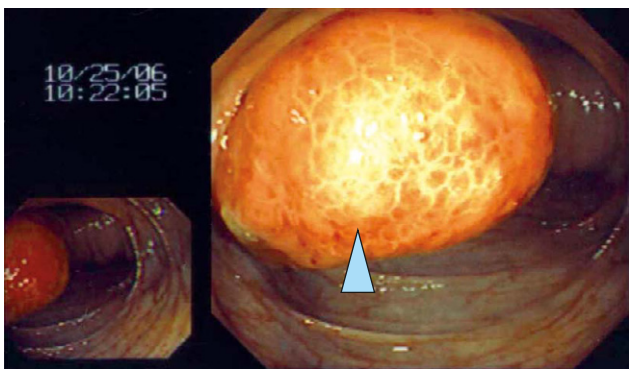

Figure 1. *Colonoscopy shows colonic submucosal tumor (arrowhead) with mucosal ulceration and partial obstruction of the lumen.*

Discussion

In this case report, we present an adult with ileocolic intussusception with a cecal lymphangioma as the leading point. In contrast to intussusception in pediatric patients, adult intussusception is relatively rare and only accounts for 5–16% of all cases of intussusception [1]. Clinical manifestations of intussusception may include acute, subacute, or chronic, nonspecific symptoms. The most common presenting symptoms in adult patients with intussusception are abdominal pain, vomiting, bleeding, melena, and anemia [2]. Altered bowel habits or unexplained diarrhea are uncommon. A number of different techniques have been reported to be useful in the diagnosis of intussusception. Abdominal sonography may show target signs on transverse views and pseudo-kidney signs on longitudinal views [3]. Cup-shaped filling defects and spiral or coil-spring appearances are characteristic of

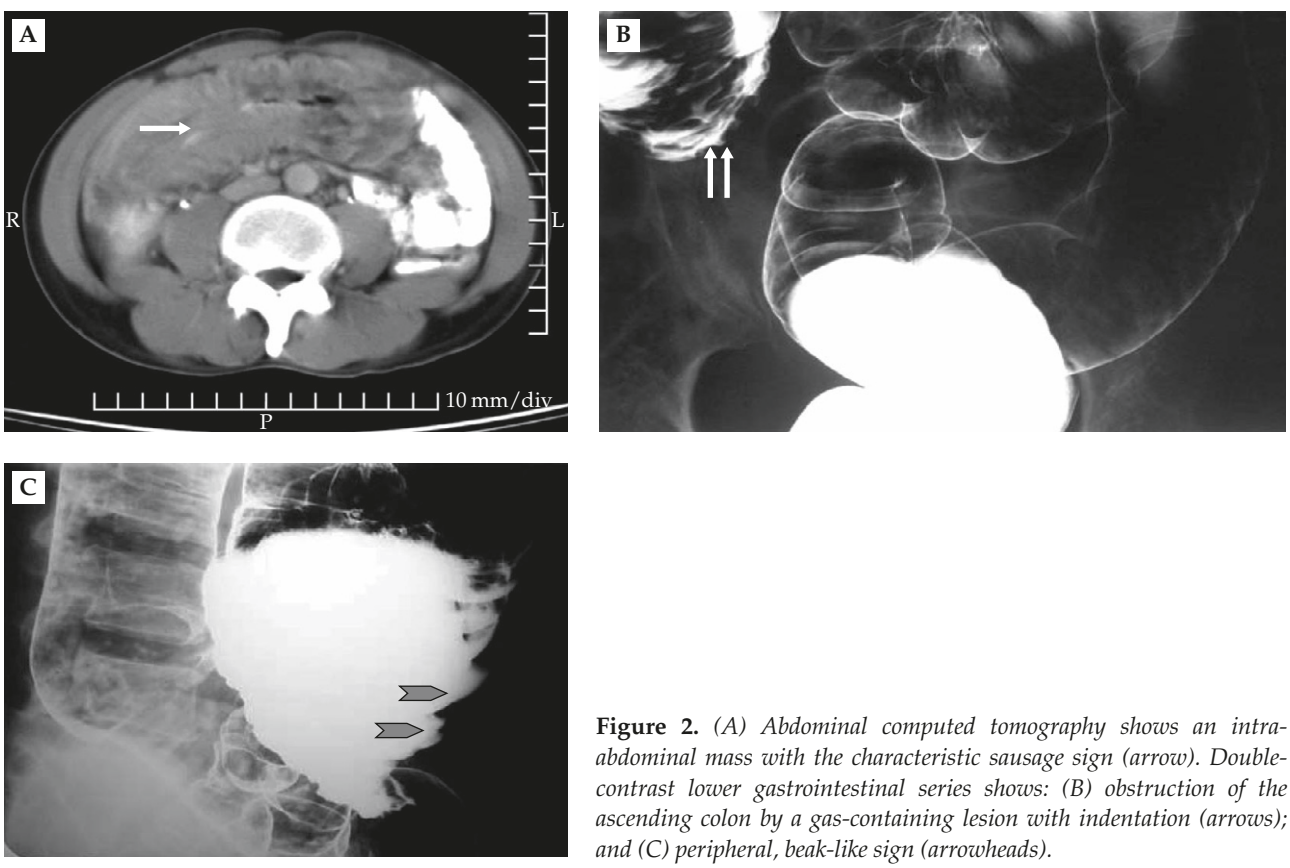

Figure 2. *(A) Abdominal computed tomography shows an intra-abdominal mass with the characteristic sausage sign (arrow). Double-contrast lower gastrointestinal series shows: (B) obstruction of the ascending colon by a gas-containing lesion with indentation (arrows); and (C) peripheral, beak-like sign (arrowheads).*

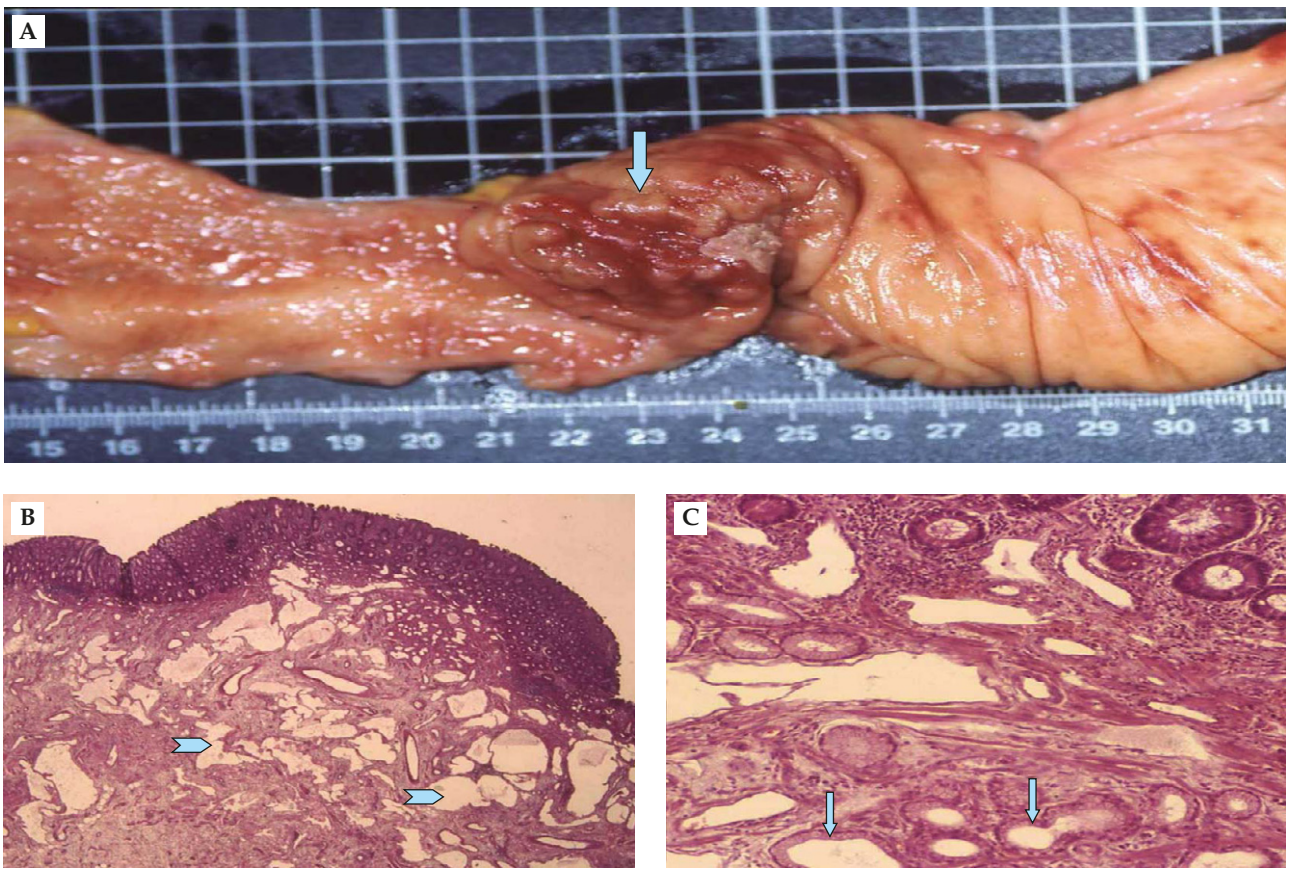

Figure 3. *(A) Cecal tumor (arrow) as the leading point of ileocolic intussusception. The resected tumor measured 3.0 × 3.0 × 1.2 cm in diameter. Microscopically, the lymphangioma showed: (B) loose myxoid stroma (arrowheads); and (C) variably sized channels lined by flattened lymphatic endothelial cells (arrows) in the submucosal layer.*

intussusception in barium enema studies [4]. Several reports have suggested that abdominal CT is the most useful radiologic method for diagnosing intestinal intussusception [2,4–6]; the usual finding on CT scans is an inhomogeneous, target- or sausage-shaped soft tissue mass with a layered effect.

The organic lesion noted in the patient in this report was a cecal lymphangioma. The exact histogenesis of lymphangiomas remains unclear. Chung et al described the histologic findings, which consisted of aggregates of lymphocytes in stroma, multiple dilated lymphatic channels with flat endothelial linings, and surrounding one layer of smooth muscle tissue [7]. Most lymphangiomas are identified in patients older than 40 years of age, with a slight male predominance. They are most likely to be found in the head, neck or axilla regions. Intra-abdominal lymphangiomas are rare and comprise < 5% of all lymphangiomas [8].

There have been more than 330 cases of colorectal lymphangiomas reported in the medical literature since Chisholm and Hillkowitz reported the first case in 1932 [9], and most cases have been documented in Eastern countries. The most common site for colorectal lymphangiomas appears to be the transverse or ascending colon [8]. However, after reviewing the literature, we found only four reported cases of adult intussusception associated with colonic lymphangiomas [10–13].

The most common clinical signs and symptoms of lymphangioma are intermittent, cramping abdominal pain. Less frequent findings include bleeding, diarrhea, sensation of fullness, abdominal mass, or protein-losing enteropathy [8]. Colonoscopy is the main diagnostic tool. Most colorectal lymphangiomas are usually identified preoperatively as submucosal tumors, as in this patient. The differential diagnosis should include other colonic submucosal tumors, such as lipomas, leiomyomas, cavernous hemangiomas, carcinoid tumors, carcinomas or mucoceles [13]. The colonoscopic features that suggest a lymphangioma

include a translucent and lustrous smooth surface, polypoid contours, yellowish or pinkish color, and easy compressibility. Two reports have suggested that endoscopic ultrasound could be a useful tool for the diagnosis of lymphangiomas of the gastrointestinal tract [13,14].

A key point in the treatment of colonic lymphangiomas is removal of the entire lesion, rather than just puncture and drainage. Surgical intervention, including segmental or wedge resection, is the most commonly reported therapy, though endoscopic therapy using snare polypectomy has been increasingly applied for the successful management of lymphangiomas. Huguet et al suggested that therapeutic colonoscopy could be used for most lesions <2.5 cm in diameter [8]. Larger tumors are usually identified in patients presenting with complications (e.g. intussusception, massive bleeding, or protein-losing enteropathy) [15], and surgical treatment should be considered in these patients, since perforation of colonic wall during polypectomy has been reported [8,10,13,16]. We reviewed the literature using the MEDLINE database and found four cases of adults with intussusception of the colon caused by colonic lymphangiomas. All cases occurred on the right side of the colon (3 cases in the cecum, 1 case in the ascending colon), all were female, and all underwent laparotomy (2 cases underwent right hemicolectomy, another 2 cases underwent ileocecalectomy) [10–13].

In this patient, we performed right hemicolectomy instead of segmental resection, without reduction of the intussusception, because of an inability to preoperatively exclude the possibility of colonic malignancy by colonoscopy (i.e. a colonic mass with mucosal ulceration). Two reports have suggested a higher incidence of malignancy in colonic intussusceptions, with 50–100% of colonic lesions in this condition being malignant [2,4]. For ileocolic intussusceptions, Barussaud et al [17] and Wang et al [4] also reported 41.6% and 50% of cases, respectively, with malignant lesions. Because of the increased incidence of malignancy in colonic intussusception, several reports have suggested that this type of intussusception should be left unreduced and should be resected as a single mass [1–2,17].

The benign nature of the lesion in some patients, such as in this study, means that further aggressive surgery could be avoided if reliable methods were available for distinguishing between benign and malignant lesions before resection of the intestine. Wang et al [4] reported their experience with the successful use of intraoperative colonoscopy to identify benign leading lesions of lipoma in two patients with ileocolic intussusception before resection, so allowing limited resection after reduction, and preservation of the ileocecal valve. As previously mentioned, an inability to exclude the possibility of colonic malignancy by preoperative examination was the reason for our determined management of the patient in the current study. We suggest that more experience in intraoperative identification of the possible etiologic agents of adult intussusception and in the differentiation of the properties of benign and malignant lesions is still needed to allow surgeons to confidently choose the appropriate treatment.

In conclusion, we reported our clinical experience with the successful management of an adult patient with intussusception, with a cecal lymphangioma as the leading point. We also reviewed the literature. This case highlights the possibility of colonic lymphangioma as the leading point of adult intussusception. Due to difficulties in preoperative differential diagnosis of the etiologies of colonic intussusception in adults, we suggest that patients should undergo laparotomy. This approach is supported by the relatively higher incidence of malignancy, compared with the rarity of colonic lymphangioma, as an etiologic factor, and the successful experiences of the five reported cases (including this one). Greater experience in perioperative differentiation of the possible etiologic agents of adult intussusception is still needed to allow clinicians to confidently choose the least invasive treatments.

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盲腸淋巴管瘤作為誘發成人腸套疊發生的導引點 — 病例報告

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我們報告了一個盲腸淋巴管瘤誘發成人腸套疊發生的案例。一位 45 歲的男性病患因為每天多次的水瀉合併間歇性腹痛，且症狀已持續二個月，而來到本院求診。大腸鏡檢發現一個盲腸黏膜下腫瘤合併黏膜潰瘍及盲腸的部分阻塞，二天後此病人因為腹痛加劇而入院。理學檢查發現右腹壓痛但無腹膜炎徵象，雙重對比劑下消化道攝影檢查顯示右側結腸阻塞處有鋸齒狀顯影及病灶邊緣鳥嘴狀顯影特徵，腹部電腦斷層檢查發現有右腹腹腔內腫塊合併香腸狀特徵，高度懷疑腸套疊。病人隨後接受了右半結腸切除術，術後病理診斷證實是腸套疊合併盲腸淋巴管瘤，病人術後恢復良好並在本院定期追蹤。這個病例彰顯了結腸淋巴管瘤也是誘導成人腸套疊發生的可能導引點病灶之一，而臨床醫師在面對及處理成人腸套疊的狀況時也應將此病灶列入可能的診斷。

關鍵詞：盲腸，腸套疊，淋巴管瘤

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