

Jejunal lymphangioma: an unusual cause of intussusception in an adult patient

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Key words

Intussusception • Adult • Submucosal lymphangioma • Small intestine

Summary

Adult intussusception is a relatively rare clinical entity. Almost 90% of cases of intussusception in adults are secondary to a pathologic condition that serves as a lead point. Lymphangioma of the small bowel is an unusual tumour that has been rarely reported to cause intussusception. In this paper, we present a rare case of adult intussusception due to jejunal lymphangioma. A 22-year-old female patient with a medical history significant for anaemia presented with intermittent colicky abdominal pain, diarrhoea and oedema of the inferior limbs for the past three months. Ultrasonography and CT scan revealed a typical target sign with dilated intestinal loops. At laparotomy, a jejuno-jejunal intussusception

was found. Partial resection of the jejunum was performed. Macroscopic examination of the surgical specimen revealed a pedunculated polyp measuring 2 cm in diameter. Histological sections of the polyp revealed in the lamina propria and submucosal layer of the jejunum several markedly dilated thin-walled lymphatic spaces lined with single layers of flat endothelial cells. The final pathologic diagnosis was submucosal lymphangioma. This case report indicates that intussusception, although rare in adults, should be considered in the differential diagnosis of abdominal pain. Moreover, it should be taken into consideration that lymphangioma is one of the possible lesions that can cause intussusception.

Introduction

Intussusception of the bowel is defined as the telescoping of a proximal segment of the gastrointestinal tract within the lumen of the adjacent segment. Intussusception is rare in adults. Almost 90% of cases of intussusception in adults are secondary to a pathologic condition that serves as a lead point, such as carcinomas, polyps, Meckel's diverticulum, strictures or benign neoplasms, which are usually discovered intraoperatively¹⁻³. Lymphangioma of the small bowel is an unusual neoplasm that has been rarely reported to cause intussusception. Herein, we report a new case of submucosal lymphangioma of the jejunum that acted as the lead point to cause intussusception in an adult female patient.

Clinical history

A 22-year-old female patient with a medical history significant for anaemia, presented with a 3-month history

of intermittent colicky abdominal pain, diarrhoea and oedema of the inferior limbs. On admission, physical examination revealed left lower quadrant pain and tenderness. Laboratory tests showed a microcytic, hypochromic anaemia. Abdominal X-ray revealed markedly dilated bowel loops with multiple air-fluid levels. Ultrasonography revealed a typical target sign with dilated intestinal loops (Fig. 1). CT scan demonstrated a "target lesion", a sign pathognomonic for intussusception and compression of iliac vein by the overlying iliac artery (Cockett syndrome) (Fig. 2). Emergency laparotomy was performed. At laparotomy, a jejuno-jejunal intussusception was found. Partial resection of the jejunum was performed. Macroscopic examination of the surgical specimen revealed a pedunculated polyp measuring 2 cm in diameter. The polyp was soft and compressible and was macroscopically multicystic containing a clear fluid. Histological sections of the polyp showed numerous and variable-sized dilated lymphatic channels in the mucosa and submucosa of the jejunum (Fig. 3). The lymphatic channels contained eosinophilic mate-

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Fig. 1. Ultrasonography showing a typical target sign with dilated intestinal loops.



Fig. 2. Computed tomography scan demonstrated a "target lesion", a pathognomonic sign for intussusception.

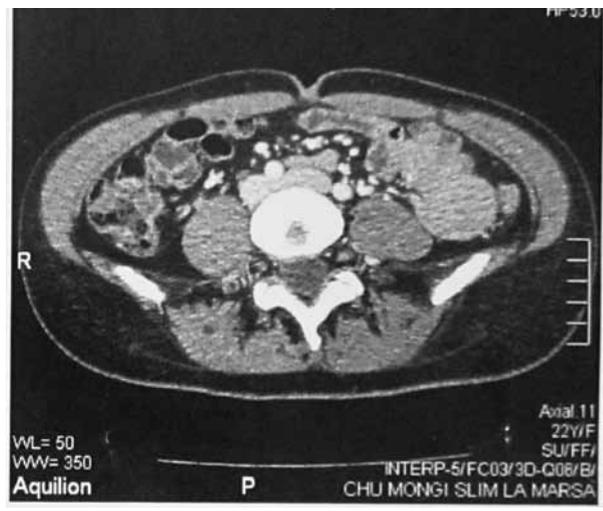


Fig. 3. Histological section of the polyp showing dilated lymphatic vessels in the mucosa and submucosa of the small intestine filled with eosinophilic material (Haematoxylin and eosin, magnification $\times 40$).

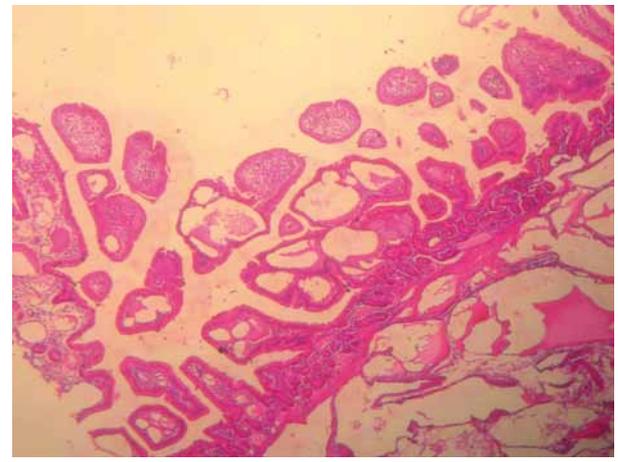
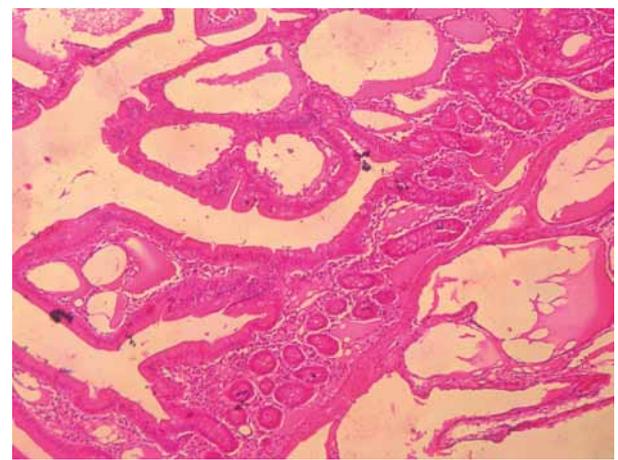


Fig. 4. The lymphatic channels contained eosinophilic material and were lined by flattened endothelium (Haematoxylin and eosin, magnification $\times 200$).



rial and resulted in broadening of the villi in some areas. They were lined by flattened endothelium (Fig. 4). The non-involved small intestine demonstrated villi of normal height and appearance. The final pathologic diagnosis was submucosal lymphangioma. The postoperative course was uneventful and there were no signs of recurrence at three weeks after surgery.

Discussion

Adult intussusception is a relatively rare clinical entity, and accounts for 1% of patients with bowel obstruction and 5-10% of all intussusceptions^{2,4}. The site of intussusception is more commonly the small bowel (83.3%) than the colon (16.7%)⁴. Intussusceptions have been

classified according to their location into four categories: (1) entero-enteric, confined to the small bowel, (2) colo-colic, involving the large bowel only, (3) ileo-colic and (4) ileo-cecal⁵. In our patient, intussusception was confined to the jejunum and was thus classified as entero-enteric. Intussusceptions have also been classified according to aetiology (benign, malignant or idiopathic). The most common benign cause of enteric intussusception is postoperative adhesions. Malignancy is more commonly associated with colonic intussusception than with enteric intussusception^{6,7}. In our case, submucosal lymphangioma acted as the lead point to cause intussusception in an adult patient. Lymphangiomas are uncommon benign tumours and occur mainly in children. The most common sites are the head, neck and axillary lesions. Intra-abdominal lymphangiomas

Tab. I. Classification and histological features of different subtypes of lymphangioma.

Histological subtype	Pathological features
Capillary lymphangioma	Small thin-walled lymphatics usually situated superficially in the skin.
Cavernous lymphangioma	Larger lymphatics having a connection with normal adjacent lymphatics.
Cystic lymphangioma	Lymphatic spaces of various sizes having no connection with normal adjacent lymphatics.
Haemolymphangioma	Lymphangioma with a vascular component.

are less common, accounting for only about 9% of all lymphangiomas¹². The aetiology of lymphangiomas remains unclear. A well-established theory suggests that lymphangiomas arise from sequestrations of lymphatic tissue during embryologic development^{3,7}. However, it is suggested that abdominal trauma, lymphatic obstruction, inflammatory process, surgery or radiation therapy may lead to the secondary formation of such a tumour. Unlike the typical paediatric presentation of acute onset, episodic abdominal pain, currant jelly stools and vomiting, adults often present with a vague history of symptoms that might include diarrhoea, constipation and weight loss. Nausea, vomiting and abdominal pain are the most common manifestations among adults. Radiologic studies are helpful, but not always diagnostic^{6,7}. In our case, lymphangioma was not detected using imaging investigations, but was incidentally discovered on macroscopic examination of the surgical specimen. Ultrasonography has been used to evaluate suspected intussusception. The classic features include the “target and doughnut sign” on transverse view and the “pseudo-kidney sign” in longitudinal view [3]. Abdominal computed tomography is the most accurate diagnostic procedure, revealing intussusception in 78% of cases⁷. The CT appearance is complex, including the outer intussusciptens, the inner intussusceptum and an eccentric fat density mass representing the intussuscepted mesenteric fat. According to the cut axis, the intussusception appears as a “sausage” or a “target” mass⁸. Although few reports have described magnetic resonance imaging (MRI) of adult intussusception, the general imaging characteristics of intussusception on MRI are similar to those on CT³. Intussusception in adults needs surgical exploration because of the risk of ischaemia and possible malignancy of the intussusception’s lead point. The advised surgical approach is usually total resection of the affected segment⁹. Laparoscopic exploration and definitive treatment may be performed if there are no contraindications⁶. Intestinal resection is recommended for all types of invagination in adults, without previous disinvagination¹⁰. Histologically, lymphangiomas are classified into three subtypes (Tab. I): *capillary*, *cavernous* and *cystic*. This classification is based on their microscopic characteristics. A fourth subtype, *haemangiolympangioma*, is also recognized. A simple capillary lymphangioma is usually situated superficially in the skin and composed of small thin-walled lymphatics. Cavernous lymphangioma consists of larger lymphat-

ics having a connection with normal adjacent lymphatics. Cystic lymphangioma consists of lymphatic spaces of various sizes that contain serous, chylous, bloody or purulent fluid, but has no connection with normal adjacent lymphatics¹¹. As suggested by their name, haemangiolympangiomas are lymphangiomas with a vascular component. Lymphangiomas may also be classified into *microcystic*, *macrocytic* and *mixed* subtypes according to the size of their cysts.<http://en.wikipedia.org/wiki/Lymphangioma> - cite_note-Gigu.C3.A8re-1

In summary, our patient represents an interesting case of intussusception that highlights the elusive, indolent course that adult intussusception can take. This case report indicates that intussusception, although rare in adults, should be considered in the differential diagnosis of abdominal pain. Furthermore, it should be taken into consideration that lymphangiectasia is one of the possible lesions that can cause intussusception.

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