

Multi-Detector Computed Tomography Findings of Inflammatory Myofibroblastic Tumor of the Ileum with Ileoileal Intussusception: A Case Report

염증성근섬유모세포종양에 의해 유발된 회장장중첩증의 다중검출 전산화단층촬영 소견: 증례 보고

Jeong Ah Hwang, MD, Il Young Kim, MD, Young Tong Kim, MD, Hyeong Cheol Shin, MD

Department of Radiology, Soonchunhyang University College of Medicine, Cheonan Hospital, Cheonan, Korea

Inflammatory myofibroblastic tumor (IMT) of the ileum rarely develops and moreover, intussusception of the tumor is even rarer. We report a case of IMT of the ileum with ileoileal intussusception in which we obtained multi-detector computed tomography images that revealed dilatation of the proximal ileum as well as luminal mass lesion in the distal ileum with intussusceptum.

Index terms

Abdomen
Neoplasm
Inflammatory Myofibroblastic Tumor
Small Bowel Intussusception
CT

Received October 29, 2012; Accepted January 2, 2013
Corresponding author: Il Young Kim, MD
Department of Radiology, Soonchunhyang University
College of Medicine, Cheonan Hospital,
31 Suncheonhyang 6-gil, Dongnam-gu,
Cheonan 330-721, Korea.
Tel. 82-41-570-3515 Fax. 82-41-579-9026
E-mail: ilykim@schmc.ac.kr

Copyrights © 2013 The Korean Society of Radiology

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal neoplasm that is typified by a myofibroblastic and mixed inflammatory cell infiltrate (1). The tumor presents itself most commonly in children and young adults, and it occurs most frequently in the lungs (1). Although IMT has been reported in various extrapulmonary sites, its occurrence in the ileum is uncommon. Furthermore, its presentation as intussusceptions is rare and thus, only a few cases have been reported (2). To the best of our knowledge, there is no report in the radiology literature regarding IMT-induced intussusception.

We report multidetector computed tomography (CT) imaging findings of an IMT of the ileum in an adult causing an ileoileal intussusception, and further discuss its radiological and pathological features.

CASE REPORT

A 55-year-old man was admitted to our hospital with a one-week history of abdominal pain. He had no history of any surgical operation and systemic disease. A physical examination revealed a distended abdomen with tenderness in the entire abdomen as well as hyperactive bowel sounds. Plain abdominal radiographs revealed dilated small bowel loops with different heights of air-fluid level and a thickening of valvulae conniventes with mechanical ileus. Contrast-enhanced abdominal CT demonstrated dilated small bowel loops and invagination of a segment of the distal ileum with mesentery into the adjacent thick-walled intussusciens (Fig. 1A, B). A 2.5 cm sized lead mass with enhancing thin wall in the distal portion of the intussusceptum was identified. The CT attenuation value of the mass was measured at about 34 Hounsfield units (HU) on the unen-

hanced image and approximately 41 HU on the enhanced image (Fig. 1C, D). There was a small amount of pelvic ascites and no abnormal lymphadenopathy. The differential diagnosis included Meckel diverticulum, duplication cyst, gastrointestinal stromal tumor, or leiomyoma with intussusception.

An exploratory laparotomy revealed ileoileal intussusceptions at the distal ileum along with edematous small bowel proximal to the lesion. The patient underwent a segmental resection of the intussuscepted ileum as well as an ileoileostomy.

Grossly, the resected specimen of the ileum measured 8 cm in length and 3.5 cm in circumference. There was a solid, well-defined, and intraluminal pedunculated mass measuring 2.5 × 2.0 cm in the ileum (Fig. 1E). The cut surface of the mass revealed a whitish tissue and there was no hemorrhage or necrosis in the mass (Fig. 1F). Microscopically, the well-demarcated tumor involved a submucosa and a muscle layer. The tumor was composed of an admixture of proliferating the spindle cells and inflammatory cells embedded in a highly vascularized stroma. There were mild cellular atypia and mitotic figures (4/50 high power field) (Fig. 1G). Immunohistochemistry of the tumor

cells was positive for the smooth muscle actin (Fig. 1H). The resected tumor of the ileum was diagnosed as polypoid IMT.

DISCUSSION

IMT is a rare mesenchymal neoplasm that is typified by a myofibroblastic and mixed inflammatory cell infiltrate (1). Since its first description in 1937, the understanding of IMT has evolved from a reactive inflammatory process to a neoplasm of an uncertain malignant potential (2). Disagreement and uncertainty about the histogenesis of IMTs has resulted in a number of synonyms, which include plasma cell granuloma, plasma cell pseudotumor, inflammatory myofibrohistiocytic proliferation, omental-mesenteric myxoid hamartoma, and, most commonly, inflammatory pseudotumor (3).

The etiology of IMT is poorly understood. Since IMT may be seen following abdominal surgery and trauma, it may represent an infectious or inflammatory process (4). However, based on the role of oncogenic viruses and cytogenetic abnormalities, including the anaplastic lymphoma kinase gene rearrangements

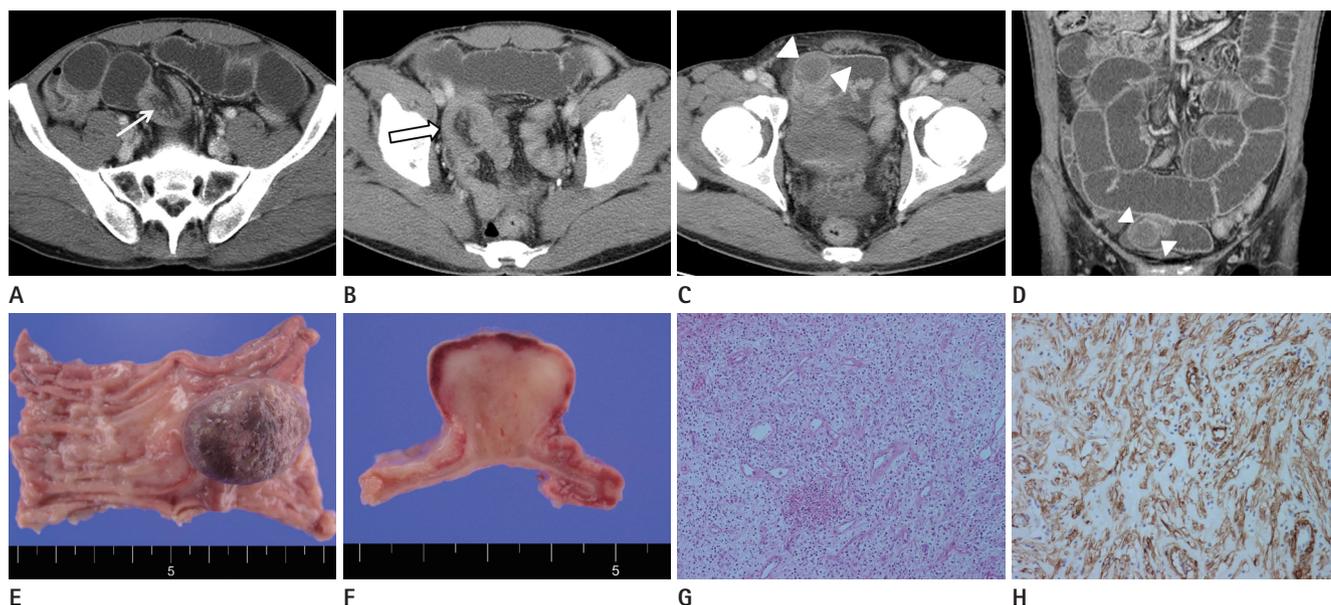


Fig. 1. A 55-year-old man with abdominal pain for one week.

A, B. Contrast enhanced axial CT scan demonstrates invaginated mesenteric fat and vessels (arrow) as well as bowel wall thickening of the intussusceptum (open arrow).

C, D. Axial (**C**) and coronal (**D**) reformatted images show a 2.5 cm sized lead mass (arrowheads) with enhancing thin wall in the distal portion of the intussusceptum. The CT attenuation value of the mass is measured about 34 Hounsfield unit (HU) on unenhanced image (not shown) and about 41 HU on enhanced image.

E, F. Gross specimen showing intra-luminal pedunculated solid mass measuring 2.5 × 2.0 cm. Cut surface shows myxoid component and there is no evidence of hemorrhage or necrosis.

G, H. Microscopic view of the lesion shows that the tumor is composed proliferation of spindle-shaped myofibroblastic cells and infiltration of inflammatory cells embedded in a highly vascularized stroma (H&E staining, × 40). Immunohistochemistry was positive for smooth muscle actin.

on chromosome 2p23, clonal chromosome abnormalities, and DNA aneuploidy, a neoplastic origin for IMT has been supported more recently (2, 3).

IMT occurs most commonly in the lungs. Extrapulmonary sites have also been reported, which include the mesentery and omentum, genitourinary tract, gastrointestinal tract, retroperitoneum, pelvis, head and neck, trunk, and extremities (1). Among extrapulmonary IMTs, 43% of the sites arise from mesentery and omentum whereas the small bowel accounts for only 1.2% of the affected sites (1). Moreover, IMTs arising from the small bowel causing intussusception are rare (2, 3). Intussusception is primarily a disease among infants and children, and about 5% of the cases occur in adults. Whereas 90% of childhood-type intussusceptions are idiopathic, an underlying pathologic process is usually identifiable in over 90% of adult cases (5). Small bowel intussusception without a lead point is more common than intussusception with a lead point. A lead point intussusception involving the small bowel is generally due to a benign condition and less often due to a neoplasm, which is usually a metastatic lesion (6).

IMTs arising from small bowel causing intussusception are extremely rare. Although it is hard to discern the exact underlying disease on radiologic examination, as long as IMT can cause an intussusception, the tumor should be considered as one of the causes of intestinal obstruction.

The clinical presentation of the IMT depends to some extent on its site of origin. Patients with intra-abdominal tumors may also be present with abdominal mass, abdominal pain, vomiting, constipation, and bowel obstruction. IMT may be a cause of chronic obstruction and of acute obstruction owing to intussusception (3).

Although the radiological examinations confirm obstructive features and the presence of intra-abdominal mass lesions, IMT is not a radiologic distinct entity (7). There are many factors that affect the appearance of an intussusception. These include the presence of a lead point, the configuration of the lead mass, the degree of bowel wall edema, and the amount of invaginated mesenteric fat (6). Identification of a lead mass that is separate and distinct from bowel loops is not easy. However, a mass that is seen on CT can serve as a reliable radiologic indicator of intussusceptions with a lead point, even though it is hard to discern the exact underlying disease in most cases (6). The CT ap-

pearance of IMT is variable. The mass may be hypoattenuated or isoattenuated on unenhanced scans; moreover, calcification has been observed in the cases of the pancreas, stomach, and liver. Enhancement usually occurs but is not pronounced, and a variety of patterns have been noted including early peripheral, with delayed central filling, and heterogeneous, homogeneous, and no enhancement (8). In our case, the CT attenuation value of the mass measured at about 34 HU on the unenhanced image and approximately 41 HU on the enhanced image.

The combination of conventional hematoxylin and eosin staining and appropriate immunohistochemical staining pathological studies can reliably distinguish IMTs with other mesenchymal tumors (9). The confirmation of the anaplastic lymphoma kinase protein/gene rearrangement is also useful in distinguishing IMT from other spindle cell neoplastic mimickers (3).

The mainstay of treatment for this tumor is surgical resection with wide margins. Incompletely resected tumors may have local recurrence within 1 year; however, recurrence has been seen up to 9 years after resection of the primary tumor (10).

In conclusion, ileoileal intussusceptions rarely occur in adults, particularly that due to IMT. It is hard to detect the exact underlying disease of intussusceptions on radiologic examination. Although intra-luminal IMT of small bowel loop may be a rare cause of intussusception, IMT may be considered as one of the causes of intestinal obstruction.

REFERENCES

1. Coffin CM, Watterson J, Priest JR, Dehner LP. Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. *Am J Surg Pathol* 1995;19:859-872
2. Milne AN, Sweeney KJ, O'Riordain DS, Pauwels P, Debiec-Rychter M, Offerhaus GJ, et al. Inflammatory myofibroblastic tumor with ALK/TPM3 fusion presenting as ileocolic intussusception: an unusual presentation of an unusual neoplasm. *Hum Pathol* 2006;37:112-116
3. Ntloko S, Gounden A, Naidoo M, Madiba TE, Singh Y, Ramdial PK, et al. Intestinal inflammatory myofibroblastic tumour. *S Afr J Surg* 2011;49:190-193
4. Demirkan NC, Akalin T, Yilmaz F, Ozgenc F, Ozcan C, Alkanat MB, et al. Inflammatory myofibroblastic tumor of

- small bowel wall in childhood: Report of a case and a review of the literature. *Pathol Int* 2001;51:47-49
5. Agha FP. Intussusception in adults. *AJR Am J Roentgenol* 1986;146:527-531
 6. Kim YH, Blake MA, Harisinghani MG, Archer-Arroyo K, Hahn PF, Pitman MB, et al. Adult intestinal intussusception: CT appearances and identification of a causative lead point. *Radiographics* 2006;26:733-744
 7. Ciftci AO, Akçören Z, Tanyel FC, Senocak ME, Çağlar M, Hiçsönmez A. Inflammatory pseudotumor causing intestinal obstruction: diagnostic and therapeutic aspects. *J Pediatr Surg* 1998;33:1843-1845
 8. Narla LD, Newman B, Spottswood SS, Narla S, Kolli R. Inflammatory pseudotumor. *Radiographics* 2003;23:719-729
 9. Greenson JK. Gastrointestinal stromal tumors and other mesenchymal lesions of the gut. *Mod Pathol* 2003;16:366-375
 10. Kim KA, Park CM, Lee JH, Cha SH, Park SW, Hong SJ, et al. Inflammatory myofibroblastic tumor of the stomach with peritoneal dissemination in a young adult: imaging findings. *Abdom Imaging* 2004;29:9-11

염증성근섬유모세포종양에 의해 유발된 회장장중첩증의 다중검출 전산화단층촬영 소견: 증례 보고

황정아 · 김일영 · 김영통 · 신형철

염증성근섬유모세포종양이 회장에 대단히 드물게 발생할 수 있다. 더욱이 이 종양에 의한 장중첩증은 더욱 드물다. 저자들은 염증성근섬유모세포종양에 의해 유발된 장중첩증을 경험하였기에 이에 대한 multi-detector computed tomography (MDCT) 영상소견을 보고하고자 한다. MDCT에서 회장말단부의 폐쇄에 의한 확장과 종양에 의한 장중첩증의 영상소견을 보였다.

순천향대학교 의과대학 천안병원 영상의학과