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Case Report

Ileocecal Burkitt's Lymphoma Presenting as Ileocolic Intussusception With Appendiceal Invagination and Acute Appendicitis

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Intussusception is a common cause of abdominal pain in children. Although most cases are idiopathic, about 10% of cases have a pathologic lead point. Burkitt's lymphoma is not a common etiology. Burkitt's lymphoma might present primarily as intussusception in children but has rarely been associated with appendicitis. We report a case in which a 10-year-old obese boy who initially presented with acute appendicitis due to ileocolic intussusception with appendiceal invagination. He underwent one-trocar laparoscopy and antibiotic treatment. The symptoms recurred 10 days after discharge. Colonoscopy disclosed ileocecal Burkitt's lymphoma as the pathological lead point. This case emphasizes the importance of the age of the patient and the anatomic location of the intussusception related to possible etiology, and hence the most appropriate surgical procedure.

Key Words: appendicitis, childhood cancer, intussusception, lymphoma

Intussusception occurs when a segment of bowel, the intussusception, prolapses and invaginates into another segment, the intussuscipiens.[1] The peak age of intussusception in children is between 3–18 months, with an incidence of 0.1–0.4% of live births.

Intussusception is the second most common cause of abdominal emergency in children.[1] Most pediatric ileocolic intussusceptions are idiopathic. In adults and occasionally in children over 2 years of age, a pathologic lead point for intussusception can be found.[2] In children, the incidence of identifiable lead point in pediatric intussusception has been reported as 1.5–12.0%.[3]

We report an unusual case of ileocecal Burkitt's lymphoma presented as acute appendicitis due to ileocolic intussusception with appendiceal invagination.

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Case Report

A 10-year-old obese boy with unremarkable medical history was admitted to our hospital owing to a 10-day history of intermittent periumbilical pain accompanied with nausea. He had not experienced associated fever or vomiting. His body weight was 70 kg and there was no history of upper respiratory or urinary tract infection.

Physical examination revealed tenderness over the periumbilical area, but there was neither abdominal distension nor palpable masses. His pulse rate was 90 beats/min and blood pressure was 118/80 mmHg. Abnormal laboratory data included C-reactive protein of 115.4 mg/L and a white blood cell count of 15.8×10^9/L, with 76.9% neutrophils, 14.7% lymphocytes, 5.4% monocytes and 2.8% eosinophils.

Abdominal radiograph revealed a soft tissue mass in the right upper quadrant and transabdominal ultrasonographic evaluation showed a target lesion approximately 6.4×4.4 cm in size in the right upper abdomen. In addition, an eccentrically invaginated hypoechoic lesion within the target lesion was also found. Abdominal computed tomograms (CT) disclosed intestinal obstruction, suspicious ileocecal intussusception with bowel wall thickening and a tubular hypodense lesion. An eccentrically located intraluminal lesion, swollen appendix as well as adjacent inflammatory strandings were also observed (Figures 1 and 2). The patient received one-trocar laparoscopy as intussusception caused by appendicitis was suspected. Laparoscopy revealed partial invagination of the appendix into the cecum and appendicitis with mass formation. Reduction of intussusception and appendectomy was performed, but no intraluminal mass was disclosed. Histopathological examinations showed appendicitis with an organized abscess. Postoperative antibiotics with cefazolin, gentamicin and metronidazole were prescribed. He was discharged without any symptoms of the gastrointestinal tract 7 days post operation.

Ten days later, the patient suffered from poor oral intake, postprandial abdominal pain and a weight loss of 6 kg in 10 days. White blood count was 11.3×10^9/L, with 69.8% neutrophils, 21.2% lymphocyte, 4.2% monocytes and 4.5% eosinophils. Other laboratory findings were normal. Transabdominal ultrasonography and abdominal CT scan disclosed persistent ileocolic intussusception with marked wall edema of A-colon and pericolic strandings as well as somewhat tubular soft tissue density at the central part of the intussusceptum. Colonofibroscopy revealed a mass in the ascending colon, and thus a biopsy was performed. Histopathological examination demonstrated tissue fragments with proliferative capillaries and dense infiltrations of small lymphoid cells with starry-sky appearance. The lymphoid cells

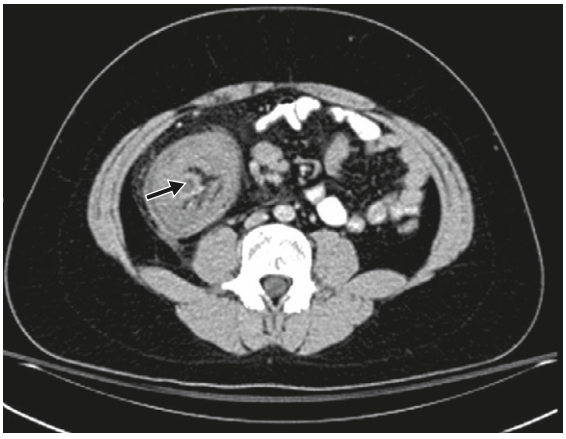

Figure 1. Abdominal computed tomogram at the level of the lower abdomen shows ileocecal intussusception with bowel wall thickening and a tubular hypodense eccentrically located intraluminal lesion (arrow) which might be an invaginated appendix.

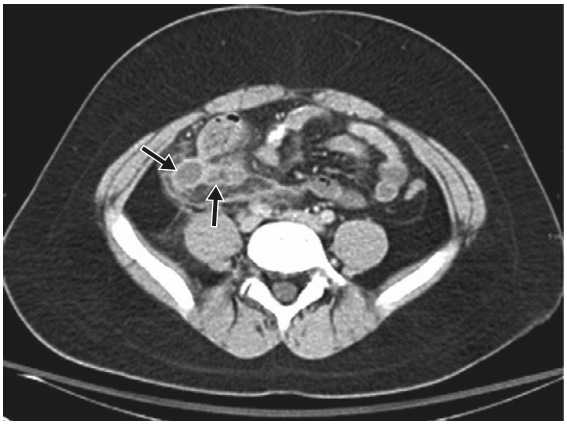

Figure 2. Computed tomogram at the level of the upper pelvis shows a swollen appendix (arrows) with prominent adjacent inflammatory strandings and dilated terminal ileal loops.

bore slightly irregular nuclei and small nucleoli. Mitotic figures were frequently noted. Subsequent immunohistochemical studies were positive for CD20, CD10 and Ki-67. Non-Hodgkin's lymphoma stage II was diagnosed according to Murphy classification. The B-non-Hodgkin's lymphoma chemotherapy protocols designed by the Taiwan Pediatric Oncology Group were administered.

Follow-up abdominal CT at 6 months delineated a progressive mass in the ileocecal region and surgical resection of the tumor was performed. Recurrent lymphoma was noted again at 7 months, even when undergoing chemotherapy with ifosfamide, carboplatin and VP-16. Unfortunately, 8 months after starting chemotherapy, the patient expired due to refractory Burkitt's lymphoma and septic shock.

Discussion

Underlying pathological causes of intussusception can be identified in 1.5–12.0% of cases.[3] These include Meckel's diverticulum, polyps, duplications, mesentery cysts, intestinal hematoma and lymphoma.[4] Lymphoma, although uncommon, arouses the most concern due to its malignant nature, and represents 6.5% of pathologic lead points of intussusception in children.[3] Navarro et al reported that long duration of symptoms and weight loss were two important clinical clues to the presence of gastrointestinal lymphoma.[5] These symptoms were present in our case.

Burkitt's lymphoma commonly presents as an abdominal mass and is often associated with abdominal pain, nausea and intestinal obstruction caused by direct compression of the bowel lumen or intussusception.[6] The peak age for gastrointestinal Burkitt's lymphoma in children is 5–15 years.[7] However, to our knowledge, Burkitt's lymphoma associated with ileocecal intussusception with appendiceal invagination, and masquerading as appendicitis has not previously been described.

Both transabdominal ultrasonogram and abdominal CT revealed findings suggestive of intussusception and appendicitis.[8–10] Although an intraluminal tubular-like lesion was suspected as a lead point, laparoscopy only detected appendicitis and intussusception.

The clinical presentation and operative findings were highly suggestive of appendicitis with intussusception. Unfortunately, the ascending colon lesion did not resolve after laparoscopic appendectomy. Indeed, subsequent rapid weight loss was an important indicator for the presence of an underlying colonic lesion.

Only 10% of non-Hodgkin's lymphoma is confined to the gastrointestinal tract.[11] Phillips et al emphasized that underlying factors such as ulcerative colitis, chronic antigenic stimulation in an immunosuppressed host, or the interaction of Epstein-Barr virus with immunosuppression, might predispose patients to develop primary colonic lymphoma.[12] However, no such underlying factors could be found in this particular patient.

Definitive treatment and management of intussusception should be individualized according to the age of the patient and the anatomic location of the intussusception. Since neoplasm is a possible etiology in older children, explorative laparoscopy and complete surgical resection, with or without intraoperative reduction depending on the anatomic location of the intussusceptions, should be a consideration. However, as shown in this case, transumbilical one-trocar appendectomy may not be sufficient for complete evaluation of submucosal bowel pathologies.

The role of surgery in gastrointestinal Burkitt's lymphoma remains controversial. Burkitt's lymphoma, arising from B-lymphocytes, responds dramatically to combination chemotherapy inducing initial rapid tumor resolution and often long-term remission.[13,14] By contrast, Magrath et al suggested aggressive operative debulking, defined as > 90% tumor removal, prior to chemotherapy following analysis of a large single institutional series of patients with Burkitt's lymphoma in Uganda.[15] In our case, Burkitt's lymphoma was localized but did not respond well to chemotherapy. In such circumstances, early surgical intervention and total tumor removal may be mandatory.

In conclusion, complete examination and acquisition of adequate biopsies via exploratory laparoscopy should be considered in children greater than 5 years of age, with intussusception as a neoplasm being a possible underlying cause of the presenting problem. When an older child with intussusception has appendicitis, intussusception secondary to intestinal lymphoma should be considered as a differential diagnosis. With ileocecal intussusceptions and appendicitis, procedures vary from reduction of the intussusception with appendectomy in benign conditions to a right hemicolectomy in the face of cancer.

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