Acute Intestinal Intussusception: Beware of Burkitt’s Lymphoma

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Abstract
Burkitt’s lymphoma can show in a variety of ways, and it’s frequently identified in children owing to the development of an abdominal tumor. This aggressive, fast growing tumor can induce either indirect symptoms due to pressure phenomena or direct involvement of the intestine lumen, resulting in intestinal obstruction or intussusception. We are reporting a case of intestinal intussusception in a patient with Burkitt’s lymphoma.

Keywords
Burkitt’s lymphoma, intestinal intussuception, imaging

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Introduction
Acute intestinal intussusception is a common cause of acute abdominal pain in pediatric population. In 25% of cases, there is an underlying cause. Burkitt’s lymphoma is among the rare causes of intussusception and it is an aggressive, malignant non-Hodgkin’s lymphoma with rapid growth of B cells. We report a case of an 8-year-old boy with intussusception due to Burkitt’s lymphoma. The patient underwent abdominal ultrasound which was complemented by a CT scan showing intestinal intussusception and colonic parietal thickening suspicious for malignancy, without evidence of upstream occlusion. The histopathology confirmed Burkitt’s lymphoma as the etiology responsible for this intussuscepted mass.

Case Report
This is a 7-year-old child with no known medical issues. The symptomatology began 4 months ago, with epigastric discomfort, vomiting, and constipation. The whole event happens in the context of a 7 kg weight loss and a change in overall condition. A general practitioner examined the patient and prescribed symptomatic therapy. Due to the exacerbation of the symptoms, the patient went to the emergency department.

A somewhat swollen, sensitive abdomen with a solid epigastric mass measuring about 7 cm was discovered during physical examination. An ileo-colonic invagination on a parietal digestive thickening was revealed during an abdominal ultrasound examination (Figure 1). A CT scan of the abdomen revealed a sub-hepatic mass similar to the ileo-colic invagination, as well as mesenteric adenopathies (Figure 2).

A cytopunction was conducted, and the results indicated Burkitt’s lymphoma. As an emergency, treatment with injectable Cyclophosphamide was started. One week after starting therapy, an ultrasound examination revealed a dramatic reduction in digestive thickness and the resultant intestinal invagination.

Discussion
Non-Hodgkin’s malignant lymphoma (NHL) represents 1% to 4% of malignant digestive tumors in children. The ileum and the colon, and more specifically the ileocecal region, are the segments most often affected because of their high concentration of lymphoid tissue. It is the third most common cancer in children, with 3 histological forms: B-cell NHL representing 65% of all

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NHL, including both Burkitt’s lymphoma and diffuse large B-cell lymphoma, lymphoblastic lymphoma and anaplastic large cell lymphoma. Burkitt’s lymphoma is the most common type representing 40-50% of NHL. The World Health Organization recognizes 3 forms Burkitt’s lymphoma: an endemic form and 2 other variants. The predisposing factors for the development of Burkitt’s lymphoma are mainly frequent childhood infections with Epstein Barr virus and disadvantaged social environments.

The clinical features normally seen with BL in the pediatric population include massive involvement of the gastrointestinal tract which is the usual site of involvement especially in sporadic forms, involvement of the jaw or face (25%), regional adenopathy, Central Nervous System involvement in 15% to 30% of cases, and other digestive signs due to intestinal obstruction or gastrointestinal bleeding.

Intussusception is a condition in which a segment of the digestive tract (proximal part) is invaginated into the distal part. It was first described in 1674 by Barbette and successfully treated surgically in 1831 by Wilson. In the pediatric population, it is frequently due to an unknown cause in 75% of cases and in 25% of cases it is secondary to an underlying cause (Meckel’s diverticulum, polyp, rheumatoid purpura, lymphoma, etc). It should be noted that 18% of Burkitt’s lymphomas are revealed by intestinal invagination and that this percentage increases with age, reaching 50% around the age of 5 to 6 years.

Burkitt’s lymphoma affects mainly the terminal part of the ileum, the cecum and the appendix. This is due to the high concentration of lymphoid tissue in the ileocecal junction. Burkitt’s tumor, like all lymphomas, extends into the submucosal and mucosal layer circumferentially, and manifests as a parietal digestive thickening or as a mural mass. The mural mass may sometimes invaginate into the downstream digestive segment.

Abdominal ultrasound is the most appropriate examination in case of suspicion of intestinal intussusception in children, it allows to make a positive diagnosis with an etiological orientation. It reveals hypo-echogenic digestive thickening with loss of parietal stratification. Sometimes it allows the visualization of a large hypo-echogenic tumoral mass. In case of intussusception, it allows to identify the intussusception coil constituted by the intussusceptum and intussuscipiens parts, producing target sign in transverse section due to the superposition of hypoechoic and echogenic layers with a central part corresponding to the mesenteric fat, as well as the “Sandwich” or “Hot Dog” sign in longitudinal section. Ultrasound can also be used to look for signs of severity such as vascular insufficiency with a thin layer of fluid within the bladder and distension of the upstream digestive tract, indicating intestinal obstruction.

The CT scan shows a voluminous necrotic tumor with little enhancement after injection. It also allows to distinguish the hypodense, circumferential, asymmetric digestive parietal thickening, poorly enhanced after injection. The intestinal lumen may be narrowed or, on the contrary, be the site of an aneurysmal dilatation. The narrowing is a consequence of the mass effect and the aneurysmal appearance is secondary to invasion of the autonomic plexus or muscularis propria. The intussusceptum is seen telescoping into the intussucipiens with fat and the mesenteric lymph nodes.

The 18FDG PET scan shows hyperfixation of the tumor and affected lymph nodes. The PET scan is
essential for patient follow-up and to distinguish fibrosis from residual tumor.  

If the diagnosis of acute intestinal intussusception on Burkitt’s lymphoma is made, patients are usually treated with cytotoxic chemotherapy which allows the tumor to melt away spectacularly just after the beginning of the treatment.

Therapeutic management is multidisciplinary, including surgery, chemotherapy, radiotherapy and immunotherapy, and different combinations may be chosen on a case-by-case basis.

Conclusion

Children’s intussusception is a prevalent cause of abdominal pain. Despite the fact that the majority of cases are idiopathic, roughly 10% of them have a pathologic lead point. Burkitt’s lymphoma does not have a common cause. Burkitt’s lymphoma is most commonly related with intussusception in children.

Author Contributions

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Figure 2. Injected abdominal CT scan (A, B, C, D) showing an asymmetric, hypodense colonic parietal thickening, narrowing the digestive lumen (white arrows). This thickening involved the ileocolic junction, the cecum and part of the ascending colon with reversal of the affected part of the colon into the downstream colon, giving a target appearance (red arrow) on the axial section and a sandwich appearance (blue arrow) on the coronal section.
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