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Burkitt's Lymphoma of Intestine Presenting as Ileocolic Intussusception

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ABSTRACT

Burkitt lymphoma, as the most common non-Hodgkin Bcell lymphoma of childhood, is rarely detected in the gastrointestinal tract. Intussusception secondary to Burkitt's lymphoma is an uncommon presentation. We describe an unusual case of intestinal Burkitt's lymphoma in a four and the half-year-old girl who presented with intermittent colicky pain three times. Imaging studies were suggestive of intussusception. The patient was subjected to the surgery of bowel resection, which revealed a creamy-gray oval-shaped mass. Histopathology through immunohistochemistry study confirmed the Burkitt lymphoma. Owing to rather nonspecific clinical and radiological features, the preoperative diagnosis of Burkitt lymphoma remains a challenging task for pediatric surgeons and radiologists. Therefore, in case of any clinical suspicion, further examinations, such as CT scan in children are recommended.

Introduction

In 1958, Dr. Dennis Parsons Burkitt initially described Burkitt Lymphoma (BL) in a 5-year-old Ugandan boy with swelling of both jaws [1]. He described three variants of BL, including endemic (mainly in Africa), sporadic, and non-endemic or American BL. In this regard, the sporadic type tends to exist in the gut lymphoid tissues, regularly as masses in the terminal ileum, the Waldeyer ring, and even with the inclusion of abdominal organs with the main inclusion of distal ileum, mesentery, or caecum. Among these, Burkitt's lymphoma is an unusual and aggressive form of Non-Hodgkin's Lymphoma (NHL) accounting for 40% of all childhood NHLs [2]. In children, the most common site for intestinal lymphoma is the ileocecal region [3].

Moreover, Burkitt lymphoma-caused Intussusception as a result of an acute abdomen is rare and has deceptive symptoms difficult to diagnose. This lymphoma is

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diagnosed after laparotomies for bowel perforation, appendicitis, obstruction, and hemorrhage [4]. In the present case, we explain Burkitt's lymphoma of the intestine as ileocolic intussusception.

Case Presentation

A four and half-year-old girl child presented to the emergency department with sudden onset of periumbilical colicky pain with low-grade fever and constipation for one month. An examination was also unremarkable: 37.4 centigrade, pulse: 80 /minute, and blood pressure 120/75 mmHg. No mucosal bleeding, generalized lymphadenopathy, or abdominal mass was observed. Blood investigations revealed a hemoglobin level of 10.1 g%, a total leukocyte count of 15.6×10^9/L with 61% neutrophils. The platelet counts 561000 (cell/mm3). Erythrocyte sedimentation rate (ESR) by Westergren's method was 10 mm. All other routine laboratory investigations showed a normal situation.

Ultrasonography examination of the abdomen showed intestinal loops with target signs 33 mm in greatest diameter, which was favorable for intussusception (Figure 1). Then the hydrostatic reduction was done, which was successful and confirmed by a follow-up ultrasound. After two days, the patient was discharged in good general condition. After ten days, she returned with hypogastric and periumbilical colicky pain extended to the right lower quadrant (RLQ), associated constipation, anorexia, and low-grade fever. The examinations showed a mild tenderness in RLQ, and the ultrasound showed ileocecal invagination measuring 43×41 mm in RLQ, which failed despite twice hydrostatic reduction. Finally, she underwent surgery with a possible diagnosis of appendicitis. Appendectomy was done, and after two days, she was discharged with the good general condition.

At third admission for acute abdomen after two weeks, she underwent an emergency laparotomy, and no CT scan was performed at each hospital admission. Intraoperative findings showed ileocolic invagination, and a right hemicolectomy was done.

The gross examination of the resected bowel consisted of a 10 cm segment of the ileum and cecum. It showed a creamy-gray ovoid mass with surface ulceration measuring 4×3×1.5 cm and encompassing 80% of the ileocecal valve edge. Microscopic findings showed total wall thickness of bowel infiltrated of medium lymphoid cells with scant clear to eosinophilic cytoplasm, high nucleocytoplasmic ratio, brisk mitotic activity area necrosis, and hemorrhage (Figure 1).

The primary differential diagnosis was high grad NHL, possibly lymphoblastic lymphoma or Burkitt's lymphoma. The BL was confirmed by immunohistochemical studies such as positivity for CD20, PAX5, CD10, Bcl6 markers, and negative expression of CD3, CD5, CD23, Bcl2, and TDT markers,ki-67 labeling index was 98%. (Figure 1). After confirmation of diagnosis, a bone scan was done, which showed bone metastasis. Three months after the first hospitalization, she is on an LMB96 chemotherapy regimen and has received the first dose of medication.

Discussion

NHL is the third most common pediatric cancer. This health problem is divided into three histological subtypes, including Bcell NHL (65%) with both Burkitt's lymphoma and diffusing anaplastic large cell lymphoma (15%), large Bcell lymphoma, and lymphoblastic lymphoma (20%) [5]. BL has a classical predilection for the head and neck regions in children with the gastrointestinal tract and kidneys being potential sites of involvement [6].

Burkitt's lymphoma is submucosal and grows underneath an intact mucosa. With the advancement of the disease, ulceration of the mucosa occurs as seen in our case. Due to the aggressive behavior and volumetric doubling of this neoplasm, the patient may have an acute abdomen as well as bowel obstruction, bleeding, or rarely intussusception. Children younger than 2 years of age are more affected (75% of cases) [7]. Infrequently intussusception is the first clinical sign of Burkitt's lymphoma. It is a potentially life-threatening pediatric emergency that leads to early detection of the disease [7]. An approximate incidence of 1-4 per 2000 children is reported for intussusception, which is regarded as a pediatric surgical emergency [8]. Although the classical triad for diagnosing the intussusception includes "red-currant jelly stools", palpable abdominal mass, and abdominal colic, a diagnostic problem exists owing to the intussusception subtler presentation in post-infancy children aged 2-3 years probably without classic triad of symptoms [4, 9]. Eighty percent of the cases with intussusception with Burkitt's lymphoma had abdominal pain, nausea, constipation, vomiting, diarrhea, malaise, or fatigue [7, 10]. These symptoms along with constant abdominal pain also were reported by our patient. Nevertheless, in 95% of cases, recurrent symptoms lasting over a week, highlight the significance of diagnosing these atypical signs for intussusception [11]. Imaging modalities, especially ultrasonography and CT scan, combined with the clinical examination are nearly 100%

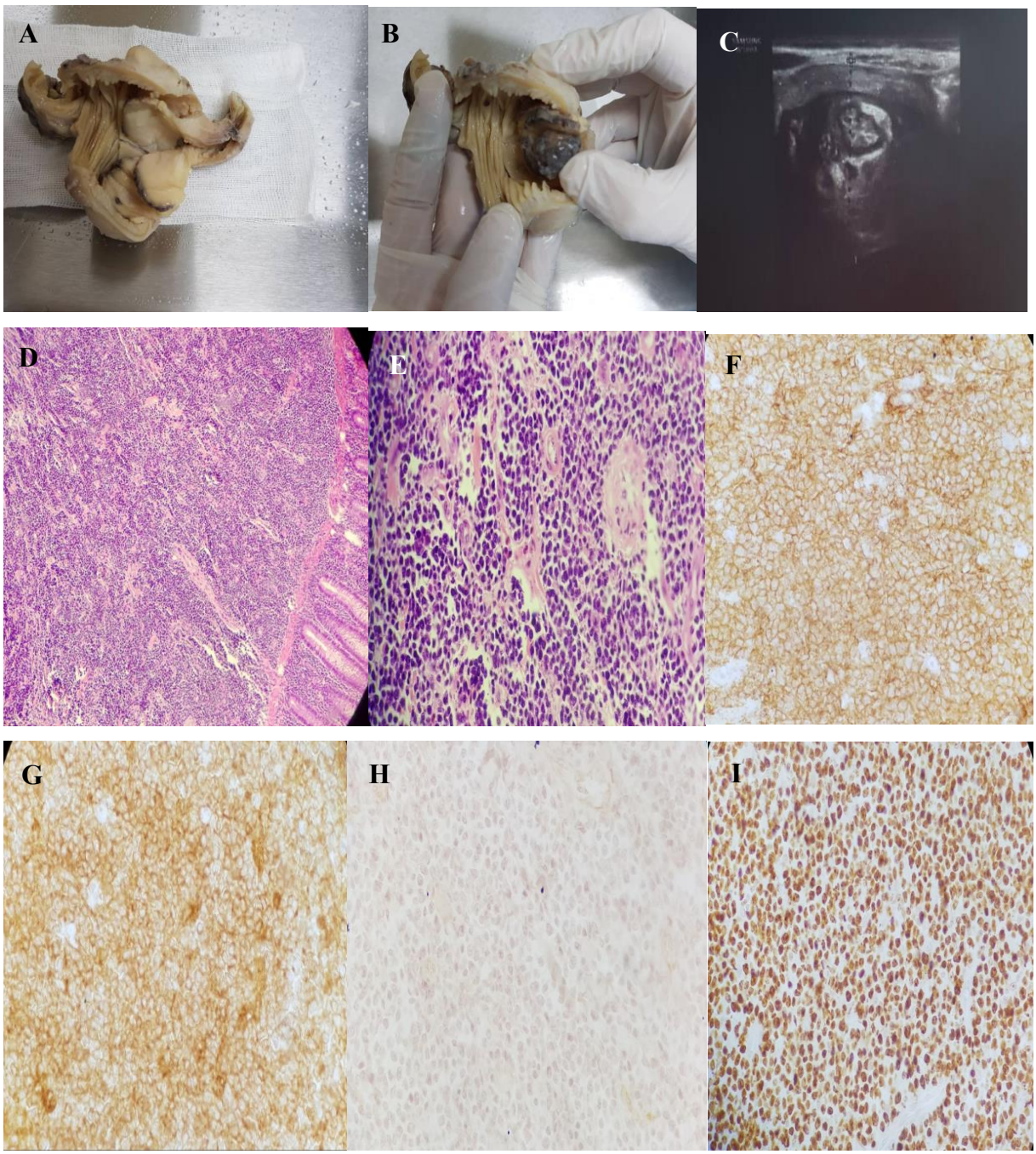

Figure 1. Gross, Microscopic (H and E) and immunohistochemical study of Burkitt's lymphoma

A and B) Gross specimen of the resected bowel showing a creamy-gray fleshy tumor with ulcerated mucosa and involving the full thickness of the bowel;

C) Ileocecal intussusception in abdominal ultrasound;

D) Photomicrograph shows diffuse infiltration of Muscularis propria by monomorphic lymphoid cells (H and E, ×100);

E) The lymphoid cells are medium-sized with a scant amount of Eosinophilia cytoplasm and central nucleus showing increased nucleus cytoplasmic ratio (H and E, ×400);

F) CD20 (Immunostain×400).

G) CD10 (Immunostain×400);

H) TDT (Immunostain×400);

I) Ki67 (Immunostain×400).

diagnostic for intussusception [6]. Ultrasonography with a false-negative rate of 0% is a trustworthy screening instrument for children at low risk for intussusceptions. Consequently, like our case, some received a contrast enema that is therapeutic and diagnostic (the gold standard in diagnosing intussusception) [12]. Laparotomy is still the gold standard in the treatment and diagnosis to ensure the entire localized tumor excision with proper margins [10]. The effectiveness of early tumor resection is not confirmed in children with extensive intra-abdominal Burkitt's lymphoma, even in the bowel obstruction setting. The reason is that surgical debulking of extensive disease may provoke tumor lysis syndrome, resulting in hyperuricemia and acute renal failure. Gahukamble et al. reported that the post-emergency laparotomy mortality in the untreated Burkitt's lymphoma setting was very high and did not enhance by aggressive tumor resection [13]. The complete surgical resection may be beneficial for patients with small localized tumors that can be carefully resected [14].

Conclusion

Ultrasonography is the most effective examination for diagnosing intestinal intussusceptions with a reported accurateness of over 100%. Also, in combination with clinical observation, it is an efficient technique for diagnosing the intussusceptions. CT scan is presently frequently recommended as the initial imaging modality for patients with recurrent abdominal symptoms and detecting intra-abdominal masses, such as neuroblastoma, Wilms tumor, and Burkitt's lymphoma. Finally, a multidisciplinary approach assures efficient treatment.

Ethical Considerations

Compliance with ethical guidelines

This study was approved by the Ethics Committee of the Hormozgan University of Medical Science (Code: IR.HUMS.REC.1399.189).

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Conflict of interest

The authors declared no conflict of interest.

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