



Case report

Burkitt lymphoma involving jejunum in children

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ABSTRACT

A 5-year-old boy presented with 3-month bloody stool from unknown origin and progressive anemia. In this case report, we review the incidence, diagnosis, pathology, treatment and prognosis of Burkitt Lymphoma.

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1. Case presentation

A 5-YEAR-OLD BOY PRESENTED WITH 3-MONTH BLOODY stool from unknown origin and progressive anemia. The colonoscopy and emission computed tomography (ECT) showed negative results. Physical examination revealed palpable abdominal mass and computed tomography (CT) of abdomen showed a gassy tumor with a gas-fluid level (Panel A). Suspected preoperative diagnosis was intestinal tumor. An exploratory laparotomy was performed. An isolated jejunum tumor with 50 cm length was resected (Panel B and C) and Burkitt lymphoma involving jejunum was confirmed by the postoperative pathological examination (Panel D). The immunohistochemistry staining demonstrated that CD10, CD20 and CD99 were positive and Ki-67 lymphocytes accounted for more than 90 percents. The boy underwent postoperative chemotherapy- Methotrexate (MTX) in combination with Cytarabine and Rituximab. At follow-up two years after resection, the boy was completely cured and thrived.

Laboratory data at initial hospitalization

Weight	18 kg
Height	110 cm
RBC	2.90*10 ⁹ /L
WBC	4.75*10 ⁹ /L
Differential count	
Neutrophils	59.4%
Lymphocytes	23.5%
Monocytes	6.6%
Hemoglobin	77.9 g/L
Hematocrit	21.8%
Platelet	198*10 ⁹ /L
AFP	2.34 ng/ml
C-Reactive Protein	86.6 mg/L
Albumin	30.7 g/L
ALT	17U/L
AST	30U/L
Sodium	139.7 mmol/L
Potassium	3.91 mmol/L
Chloride	105.1 mmol/L
Bicarbonate	19.5 mmol/L

2. Discussion

This case is extremely rare and reminds us three important points: 1. The pediatric digestive tract hemorrhage should not be restricted to some regular diseases, for example, Meckle's diverticulum, intussusception and so on. 2. Attention must be paid to the characteristic radiological imaging. The gas-fluid level of the tumor is very meaningful for the differential diagnosis. 3. The increasing

Abbreviations: ECT, emission computed tomography; CT, computed tomography.

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morbidity of pediatric intestinal tumor should be much more emphasized to avoid misdiagnosis and missed diagnosis.

2.1. The incidence of Burkitt lymphoma

Burkitt's lymphoma accounts for 40–50% of pediatric non-Hodgkin's lymphomas [1]. In the United States, primary pediatric Burkitt lymphoma has accounted for less than 5% of pediatric cancers over the past 40 years [2]. Worldwide, the incidence of lymphomas in children can be quite variable. In equatorial Africa, Burkitt lymphoma is the most common childhood malignancy, with frequent involvement of the intestines [3]. Nonetheless, although intestinal cancers are rare in China, the most prevalent intestinal malignancy in pediatric patients is lymphoma, and among lymphomas, non-Hodgkin lymphomas (NHLs), and specifically, Burkitt lymphoma, predominate [4].

2.2. Primary manifestation of burkitts lymphoma

A small percentage of patients with Burkitt's lymphoma present as if they had acute leukemia, with extensive bone marrow and peripheral-blood involvement. This form of leukemia accounts for only 1–2% of cases of childhood acute lymphoblastic leukemia. More commonly, patients with Burkitt's lymphoma present either as this child did, with a large abdominal mass and abdominal pain and distention, or with intussusception of the small bowel related to lymphoma involving the terminal ileum [5].

2.3. Pathology of Burkitt lymphoma

Burkitt lymphoma comprises sheets of medium-size lymphoid cells with scattered tingible-body macrophages which have abundant pale cytoplasm [6]. This creates a “starry sky” appearance (Panal D). The tumor cells are monomorphic, with high nuclear-to-cytoplasmic ratio, cytoplasmic molding, coarse chromatin, inconspicuous nucleoli, and many mitotic figures (Panal D). Flow cytometry reveals a predominant population of CD19⁺ and CD20⁺ B cells that express CD10 but not CD23 or CD5 and have monotypic expression of lambda immunoglobulin light chain. Immunohistochemical analysis showed that the tumor cells were negative for terminal deoxynucleotidyl transferase, a marker of lymphoid precursor cells. The tumor cells had the immunophenotype of germinal-center B cells in that they were positive for CD10 and the transcription factor Bcl-6 but negative for the antiapoptotic protein Bcl-2. The Ki-67 proliferation accounts for more than 90% (Panal D).

2.4. Therapeutic strategy and prognosis of Burkitt lymphoma

Burkitt's lymphoma is highly curable [5] and the majority of children with the disease are cured with relatively short courses of systemic multiagent chemotherapy. A risk-adapted treatment strategy is important in planning the intensity and duration of treatment for patients with Burkitt's lymphoma. Several groups that conduct clinical trials have developed risk-stratification systems comprising two to four risk groups that incorporate tumor stage, extent of disease and surgical resection, and serum lactate dehydroge dehydrogenase level. Patients with the most limited disease (completely resected abdominal disease or stage extra-abdominal disease) have a prognosis that approaches 100% event-free survival after very brief, low-intensity treatment, such as two or three cycles of chemotherapy. This patient, who has more extensive disease, would benefit from more dose-intensive chemotherapy. Effective treatment is the most challenging in patients with overt bone marrow and central nervous system involvement, which this patient did not have. Therapy directed at

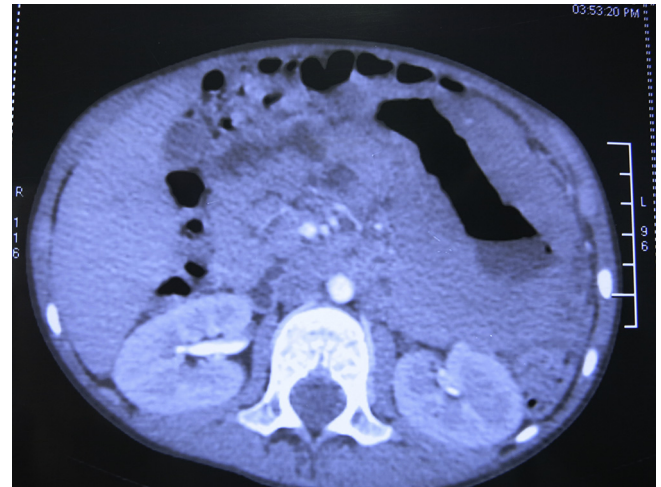


Fig. 1. CTof abdomen showed a gassy tumor with a gas-fluid level.

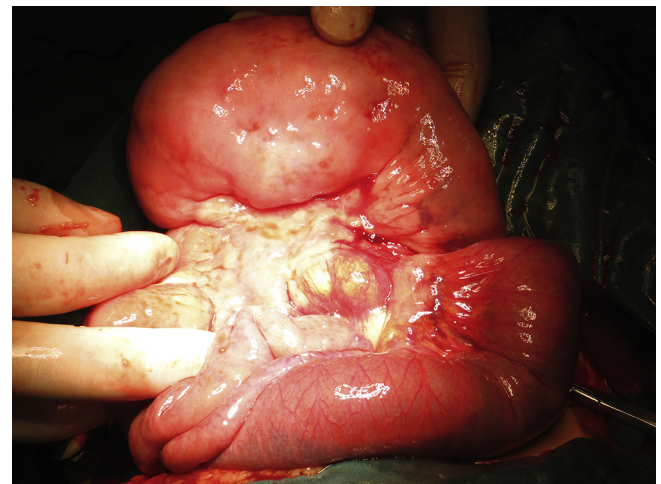


Fig. 2. An isolated jejunum tumor with 50 cm length was resected.

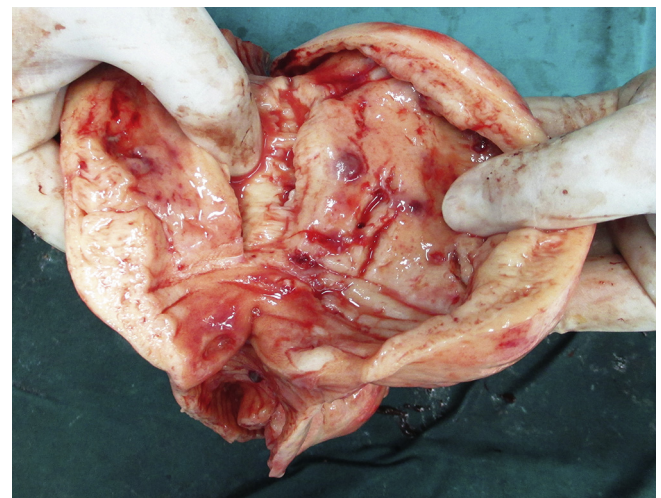


Fig. 3. Section view of the tumor.

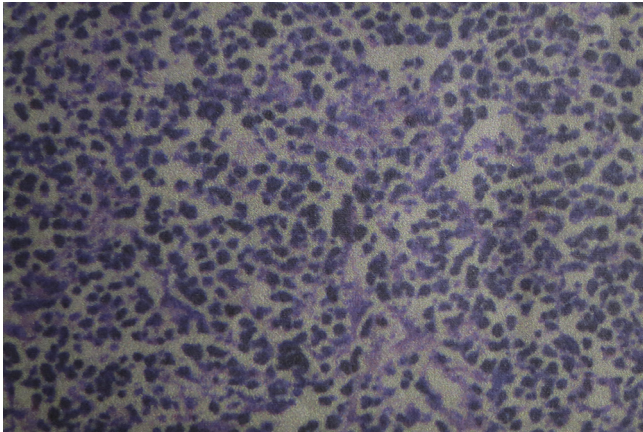


Fig. 4. The pathology of Burkitt lymphoma: “starry sky” appearance.

the CNS (in the form of intrathecal chemotherapy) is an essential component of treatment for all patients. Supportive care is also crucial, because of the risk of the tumor lysis syndrome in patients with a large tumor burden; rasburicase has improved our ability to effectively manage this complication [7].

Non-Hodgkin's Lymphoma (NHL) is the most common intestinal malignancy in children, and Burkitt's lymphoma is the most frequently encountered histologic subtype. In pediatric patients, intestinal involvement of the lymphoma is a common finding. As over half of these intestinal tumors are unresectable at the time of presentation, chemotherapy is the mainstay of treatment. However, as the tumor responds to chemotherapy, regression of the tumor in the bowel wall can result in intestinal perforation [8].

In conclusion, Burkitt's lymphoma involving jejunum is extremely rare in children. Burkitt's lymphoma is highly curable and the majority of children with the disease are cured with relatively short courses of systemic multiagent chemotherapy. Because Burkitt's lymphoma is not common in mainland China, it is necessary for us to ameliorate the management and treatment of the tumor to improve the prognosis in the near future (Figs. 1–4)..

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Conflict of interest

All authors have no conflicts of interest to disclose.

Contributor's statements

Zhi Li: Dr. Li conceptualized and designed the study, drafted the initial manuscript, and approved the final manuscript as submitted.

Jiexiong Feng: Dr. Feng carried out the initial analyses, reviewed and revised the manuscript, and approved the final manuscript as submitted.

Xiaoyi Sun: Dr. Sun, correspondence author, critically reviewed the manuscript, and approved the final manuscript as submitted.

Key learning points

This case is extremely rare and reminds us three important points:

- The pediatric digestive tract hemorrhage should not be restricted to some regular diseases, for example, Meckle's diverticulum, intussusception and so on.
- Attention must be paid to the characteristic radiological imaging. The gas-fluid level of the tumor is very meaningful for the differential diagnosis.
- The increasing morbidity of pediatric intestinal tumor should be much more emphasized to avoid misdiagnosis and missed diagnosis.

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