

## An Uncommon Burkitt's Lymphoma Form with Ileocecal Localization that Presents as Appendicular Mass

<sup>1</sup>Fuat Ipekci, <sup>1</sup>Yahya Capkis, <sup>2</sup>Gunnur Görgün and <sup>3</sup>Umit Bayol

<sup>1</sup>Department of Surgery,

<sup>2</sup>Department of Hematology,

<sup>3</sup>Department of Pathology, Tepecik Training and Research Hospital, Izmir, Turkey

### ABSTRACT

Burkitt's lymphoma represents an undifferentiated, highly malignant tumor of the B lymphocytes. Burkitt's lymphoma is a highly aggressive lymphoma identified and described in 1958 by British surgeon Denis Burkitt in Africa, in areas known as malaria belt. Currently, Burkitt's lymphoma can be divided into three main clinical variants: the endemic, the sporadic and the immunodeficiency-associated variants. The endemic variant characteristically involves the jaw or other facial bone. In the sporadic form, on the other hand, the jaw is less commonly involved, compared to the endemic variant and the ileo-cecal region is the common site of involvement. The aim of present study is to report a rare Burkitt's Lymphoma case whose clinical presentation mimicked an appendicular plastrone and which was properly diagnosed only after the surgery.

**Key words:** Burkitt's lymphoma, caecal localization, appendicular mass

Insight Surgery 1 (1): 1-3, 2015

### INTRODUCTION

Lymphomas accounts for 12% of childhood malignancies. Forty percent of lymphomas are Hodgkin's lymphoma, while the rest is classified as non-Hodgkin lymphoma. Burkitt's Lymphomas make up 40-50% of non-Hodgkin Lymphomas<sup>1,2,3</sup>. In Turkey, Burkitt's Lymphoma is the second most common disease among the childhood malignancies following leukemias<sup>4,5</sup>. Burkitt's lymphoma can be divided into three main clinical variants: The endemic, the sporadic and the immunodeficiency-associated variants. There are important differences in presentation, epidemiology and clinical features between the endemic and nonendemic form. The endemic variant characteristically involves the jaw or other facial bone. In the sporadic form, on the other hand, the jaw is less commonly involved, compared to the endemic variant and the ileo-cecal region is the common site of involvement. Burkitt's lymphoma has a characteristic distribution of its types in the world<sup>2,4</sup>. Endemic Burkitt's lymphoma is predominantly seen in younger children in Africa with a peak age incidence of 7 years and affects mostly the chin. Sporadic Burkitt's Lymphoma involves the

abdomen and the age, they most commonly seen is 10-11 years<sup>2,3</sup>. It can either cause different kinds of surgical diseases and complaints or simulates them.

### CASE

The patient is a 13 year-old boy with a one week history of malaise and night sweats. He had noticed a mass in his abdomen. He had admitted to a regional health center two months ago with abdominal pain located at the right lower quadrant. After his examination there, he had been given medical treatment for the diagnosis of appendicular plastrone. During treatment, he had been hospitalized because of recurrent pain. He was referred to emergency department with a preliminary diagnosis of acute abdomen and intra-abdominal abscess. On physical examination, there was a large mobile mass in the right lower quadrant. Laboratory examination revealed a white blood cell count of 14.100 mm<sup>3</sup>, a hemoglobin of 11.8 g dL<sup>-1</sup>, uric acid of 7.4 mg dL<sup>-1</sup>. Chest X-ray was normal. On abdomen computed tomography, a cecal mass with air densities in it was observed and this appearance was thought to be consisted with perforated appendicitis and intra abdominal abscess formation. On these grounds he had been operated urgently. Intra-operatively, an

---

**Corresponding Author:** Fuat Ipekci, Department of Surgery, Tepecik Training and Research Hospital, Izmir, Turkey

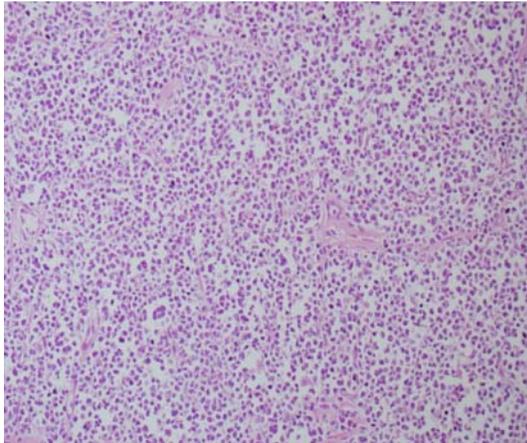


Fig. 1: Pathology results of Burkitt's Lymphoma

obstructive cecal mass of huge dimensions (15×6 cm), with satellite lesions involving almost all of the right colon was observed. There was also reactionary free fluid in the abdomen and all the intestinal mesentery was thickened because of the tumor invasion. Right hemicolectomy and ileo-transversectomy was performed. Samples were taken from mesenteric lymph nodes for pathologic investigation. Final pathology revealed Burkitt lymphoma.

Pathology result was reported as following shown in Fig. 1. Macroscopy: A tumoral lesion of 15×14×6 cm dimensions with rough granules encompassing entirely all the caecum and thickening of the intestinal tissue reaching up to 3.5 cm in some parts of the intestinal wall all along the 32 cm-segment of right hemicolectomy material that involves ileo-caecal valve and appendix. Microscopy: CD 20: Diffuse (+), CD 79a: Diffuse (+) Burkitt's Lymphoma (non-hodgkin lymphoma, small cellular type). Oral medication was started at the third day, of the post-operative period and at the fifth day the patient was discharged after being consulted to hematology department for chemotherapy

## DISCUSSION

Burkitt's lymphoma represents an undifferentiated, highly malignant tumor of the B lymphocytes. Burkitt's lymphoma is a highly aggressive lymphoma identified and described in 1958 by British surgeon Denis Burkitt in Africa, in areas known as malaria belt<sup>6</sup>.

This type of lymphoma has received a variety of names in different classifications of lymphomas and leukemias. However, with the publication of the WHO Classification of Haematopoietic and Lymphoid Tumors, it is once again known as Burkitt's lymphoma.

A high incidence of IgM antibodies to the Epstein-Barr virus (EBV) capsid antigen has been noticed in the serum of patients with Burkitt's lymphoma on immunochemical examination. In recent years, efforts have been focused on improving therapy for this aggressive neoplasm while minimizing treatment-associated toxicity. It is difficult to differentiate Burkitt lymphoma and precise diagnosis based on histologic, immunophenotypic and genetic features remains the critical first step in planning appropriate therapy.

In the middle of the last century, Denis Burkitt noted children with distorted faces and sometimes an accompanying abdominal mass could be observed<sup>6</sup>. These tumors then were believed to be sarcomas. He encountered with Epstein and showed samples from diseased children. Epstein identified a virus which is known as Epstein Bar Virus (EBV) today. It is the first virus which was shown to be related human cancer<sup>6</sup>.

Malign lymphoma is a very rare entity among the gastrointestinal tumours. It can affect the gastrointestinal system as a part of systemic disease or it may originate primarily from gastrointestinal tract. Lymphomas account for 10-15% of the intestinal tumours and nearly 1% of the colon tumours<sup>7</sup>. Because of its hefty lymphoid tissue, lymphomas mainly originate from ileum. Intestinal involvement of lymphoma as a part of systemic disease is very common but sole caecal localization without any other organ involvement is very rare. Symptoms are not different from any other diseases causing intestinal obstruction and therefore, lymphoma is usually diagnosed after laparotomy. In 1/4th of the patients, rectal bleeding can be observed and abdominal mass can be palpated. Double-Contrast Barium Enema, CT and Colonoscopy may help for diagnosis and can determine the level of the obstruction<sup>8</sup>.

Burkitt's Lymphoma is the very aggressive variant of the Non-hodgkin B-Cell Lymphoma. Rapid cell proliferation and the "starry sky" appearance can be observed in the histopathologic samples. These features are especially prominent in children and patients with immunodeficiency for rapid cell proliferation and apoptosis are the typical features of it<sup>9</sup>. Because of these characteristics, Burkitt lymphoma can easily be recognized in the histopathologic samples. It has three forms: Endemic form, characterized by the exponential tumours of the face bones and this form is most commonly seen in Africa. Second form of Burkitt's lymphoma is sporadic form; Sporadic Burkitt's lymphoma occurs worldwide. The abdomen, especially the ileocecal area, is the most common site of involvement. The ovaries, kidneys, omentum,

Waldeyer's ring and other sites may also be involved. This variant is characterized by tumoral proliferation of cells caused by translocation of c-myc oncogen in 8th chromosome. It represents with abdominal masses. The third form is seen in patients with immunodeficiency which goes with widespread lymphadenopathies<sup>7</sup>. Short-duration, high-intensity chemotherapy, sometimes combined with CNS prophylaxis, yields excellent survival in children. The institution of the CODOX-M/IVAC regimen (Magrath protocol)-two cycles of CODOX-M (cyclophosphamide, vincristine, doxorubicin, high-dose methotrexate and intrathecal therapy) alternating with IVAC (ifosfamide, etoposide, high-dose cytarabine and intrathecal therapy) for high-risk disease and for those with low risk disease (e.g., one extranodal site or completely resected abdominal disease with normal LDH), three cycles of CODOX-M, represented a major step forward in the treatment of Burkitt's lymphoma. Children and adults treated with this regimen had similar outcomes; the Event Free Survival (EFS) rate at 2 years was 92% for the group as a whole<sup>2,3</sup>. However, there were significant associated toxicities, because of the rapid, effective tumor cell killing with this aggressive protocol. Careful monitoring is required to avoid the complication of tumor lysis syndrome<sup>5</sup>. Rituximab, a monoclonal anti-CD20 antibody, may improve outcome; but additional investigation is required to determine the role of rituximab in the treatment of Burkitt's lymphoma<sup>1,3</sup>. A good prognosis is associated with resectable abdominal disease<sup>5</sup>.

## CONCLUSION

In Burkitt's Lymphomas of gastrointestinal system, the intestines are directly affected. Very rarely, it may lead to invagination, bleeding, mesenteric ischemia, septic shock and intestinal obstruction. Specifically, sporadic Burkitt's lymphoma most commonly presents in the area of the ileocecal valve. This phenomenon is thought to be related to the confluence of immunoproliferative lymph nodes and Peyer's patches in this area. It can even act like plastron appendicular mass as in this case and can only be diagnosed after operation. Although the primary treatment of Burkitt's lymphoma is chemotherapy. Sporadic variety may be best managed by a combined modality of chemotherapy and surgical extirpation since perforation and bleeding due to the tumor can be observed. Gastrointestinal bleeding might cause morbidity or mortality. Since it is of utter importance to differentiate Burkitt's lymphoma for planning the treatment surgical resection could be a

great help. Moreover, resection of the tumoral mass might help to prevent tumor lysis syndrome related to the chemotherapy.

The manifestations of this acute lysis of cells results in hyperuricemia, hyperkalemia and hyperphosphatemia. Sudden deaths as a result of electrolyte imbalance have been reported. Extirpative resection of the tumor mass in Burkitt's lymphoma has a direct effect on staging and treatment.

Patients with a history of malaise, night sweats and abdominal masses, the possible diagnosis of Burkitt's lymphoma should be kept in mind. In complicated cases like perforation, bleeding and obstructions, surgical resection should be performed. Although the primary treatment is the chemotherapy.

## REFERENCES

1. Shad, A. and I. Magrath, 1997. Non-hodgkin's lymphoma. *Pediatric Clin. North Am.*, 44: 863-890.
2. Cotran, R.S., V. Kumar, T. Collins and S.L. Robbins, 1999. Robbins Pathologic Basis of Disease. 6th Edn., W.B. Saunders Co., California, ISBN: 9780721673356, pp: 662-664.
3. Lanzkowsky, P., 2000. Manual of Pediatric Hematology and Oncology. 3rd Edn., Academic Press, London, pp: 445-469.
4. Cavdar, A.O., S. Gozdasoglu, G. Yavuz, E. Babacan and F. Unal *et al.*, 1993. Burkitt's lymphoma between African and American types in Turkish children: Clinical, Viral (EBV) and molecular studies. *Med. Pediatr. Oncol.*, 21: 36-42.
5. Ertem, U., F. Duru, A. Pamir, N. Tacyildiz and A. Dagdemir *et al.*, 1996. Burkitt's lymphoma in 63 Turkish children diagnosed over a 10 year period. *Pediatr. Hematol. Oncol.*, 13: 123-134.
6. Olaniyi, J.A., 2012. Burkitt lymphoma: A review. *Sci. Rep.*, Vol. 1. 10.4172/scientificreports.337
7. Freedman, A.S. and N.L. Haris, 2006. Clinical and pathologic features of Burkitt's lymphoma. *Up To Date*, 14:3:2006.
8. Yustein, J.T. and C.V. Dang, 2007. Biology and treatment of Burkitt's lymphoma. *Curr. Opin. Hematol.*, 14: 375-381.
9. Ferry, J.A., 2006. Burkitt's lymphoma: Clinicopathologic features and differential diagnosis. *Oncologist*, 11: 375-383.