

# Burkitt Lymphoma of Appendix not Presenting with Acute Abdomen: Case Report

## Akut Batın ile Başvurmayan Apendiksin Burkitt Lenfoması

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**ABSTRACT** Burkitt's lymphoma is a high grade B cell neoplasm typically presenting with abdominal mass in children in non-endemic regions. Primary appendicular lymphoma (PAL) is very rarely observed and comprises about 0.015% of all gastrointestinal lymphomas. Cases with PAL generally present with acute abdomen and are usually diagnosed in the postoperative period. In this study, a case of Burkitt Lymphoma in our clinic, which has originated from the appendix, without symptoms of acute abdomen with right lower quadrant swelling for 2 months, has been evaluated. Right hemicolectomy+end-lateral ileotransversostomy were performed on the patient. Postoperative treatment was continued with the classic combination of cyclophosphamide, doxorubicine, vincristine and prednisone (CHOP). On the control computed tomography performed 12 months later, there was no tumor residue detected in the abdomen and the pelvis.

**Key Words:** Appendiceal neoplasms; burkitt lymphoma; abdomen, acute

**ÖZET** Burkitt lenfoma endemik olmayan bölgelerde çocuklarda abdominal kitle ile klinik bulgu veren, yüksek grade B hücreli neoplazmdır. Apendiksin primer lenfoması (APL) tüm gastrointestinal lenfomaların %0,015'ini oluşturur. APL hastaları genellikle akut batın ile başvururlar ve daha çok postoperatif dönemde tanı alırlar. Bu çalışmada, yaklaşık 2 aydan beri sağ alt kadranda şişliği bulunan ve akut apandisit bulguları bulunmayan apendiks kaynaklı Burkitt lenfoma olgusu değerlendirildi. Olguya sağ hemikolektomi+uç-yan ileotransversostomi gerçekleştirildi. Postoperatif dönemde klasik siklofosfamid, doksorubisin, vinkristin ve prednizolon (CHOP) tedavisi uygulandı. Postoperatif 12. ayda çekilen kontrol bilgisayarlı tomografide abdomen ve pelviste tümöral doku tespit edilmedi.

**Anahtar Kelimeler:** Apendiks tümörleri; burkitt lenfoması; karın, akut

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**B**urkitt's lymphoma is a high grade B cell neoplasm presenting typically with abdominal mass in children in non-endemic regions. Primary Burkitt lymphoma of the appendix is a very rare tumor. Acute appendicitis is the most common clinical manifestation of tumors originating from the appendix.<sup>1,2</sup> In this study, a case of Burkitt Lymphoma in our clinic originating from the appendix with right lower quadrant swelling has been presented.

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## CASE REPORT

A 16-years-old female patient presented to our clinic with the complaints of such as rebound and defense (Figure 1). There were no complaints of nausea, vomiting, weight loss or changes of intestinal habits. On the abdominal examination, there was minimal sensitivity with palpation. On the performed laboratory evaluation, WBC was  $11.77 \times 10^3/\mu\text{L}$  and the other laboratory values were within normal limits. On abdominal ultrasonography (AUSG), a heterogenic, lobulated contoured hypoechoic lesion of about  $7 \times 10 \times 15$  cm in size in right lower quadrant in abdomen was reported. On abdominal computerized tomography (CT), a mass lesion at the right abdominal lower quadrant of  $14 \times 11 \times 10$  cm size, including fluid density areas at its center, causing compression onto the surrounding intestinal loops and not significantly infiltrating the surrounding tissues was visualized. Several lymph nodes sized  $<1$  cm at the superior medial part of the mass were reported. The ureter, bladder, uterus and ovaries were of normal appearance. Colonoscopy was recommended to the patient, but it was not accepted by the patient. After obtaining consent from the patient for the operation, preparations were made for exploration. On exploration, a mass sized  $10 \times 15$  cm at the right abdominal lower quadrant, which had not invaded the surrounding tissues was determined.

The mass had invaded the cecum. Right hemicolectomy+end lateral ileotransversostomy were performed (Figure 2). In the pathologic evaluation, tumor formation of the appendix compatible with Burkitt lymphoma invading the muscularis propria and subserosa away from the  $10\mu$  serosal surface was reported. The proximal and distal resection margins were free of malignancy. Twenty nine lymph nodes were detected in the specimen and they were free of malignancy. There was a uniform and homogeneous proliferation of transformed lymphoid cells with medium-sized, round nuclei, non-cleaved, dispersed chromatin, several small nuclei and abundant basophilic cytoplasm that may give the cells a cohesive appearance. A high mitotic rate and "starry sky" appearance resulting from

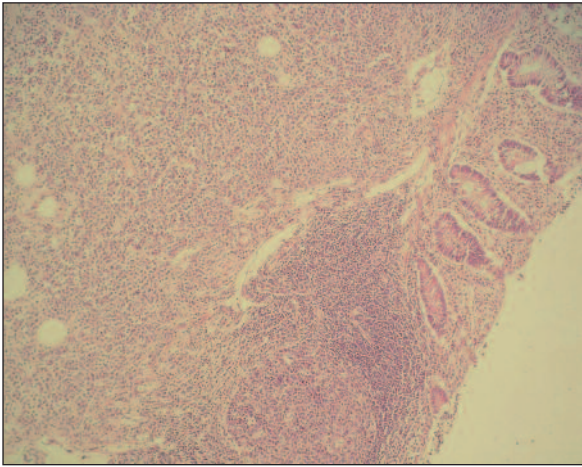


FIGURE 1: Clinical presentation of Burkitt lymphoma.

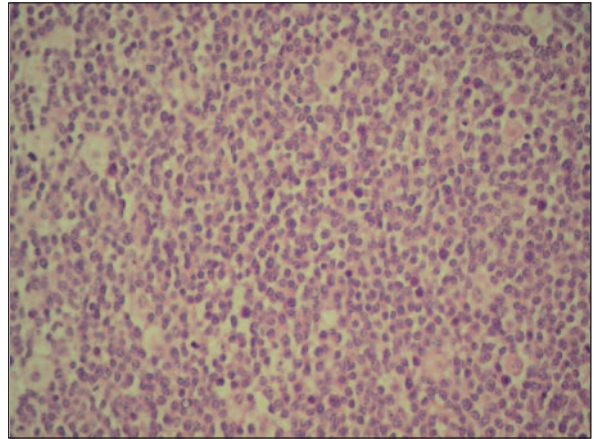


FIGURE 2: Intraoperative view of Burkitt lymphoma.

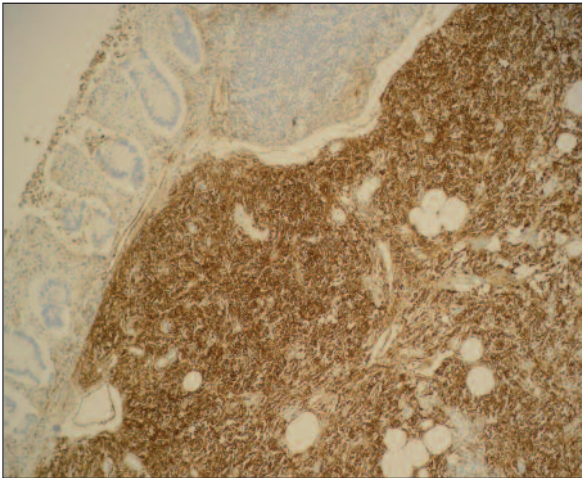
the presence of numerous tingible-body macrophages were typically noted. The tumor cells were CD-20, Bcl-2 and CD-10 positive and CD-5, CD-23, Bcl-6 negative. The important feature of Burkitt lymphoma is Ki-67 staining of more than 95% of cells (Figure 3-8). The carcino-embryonic antigen (CEA) level was  $1.5$  ng/mL and within normal limits. Postoperative treatment was continued with the classic combination of cyclophosphamide, doxorubicine, vincristine and prednisone (CHOP). On the control CT performed 12 months later, there was no tumor residue detected in the abdomen or the pelvis.



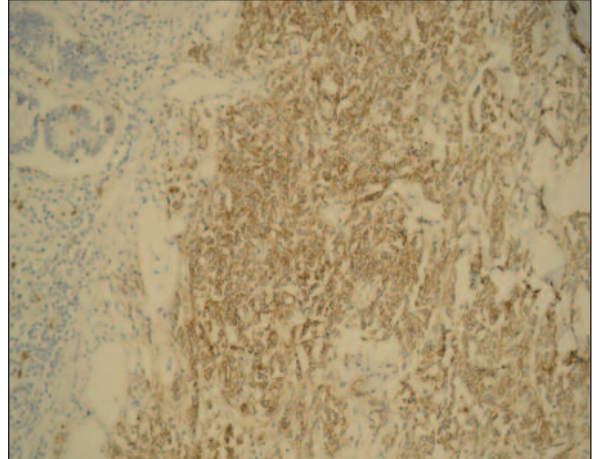
**FIGURE 3:** Low power view showing flat mucosa on the right and lymphoma on the left. (HEX40).



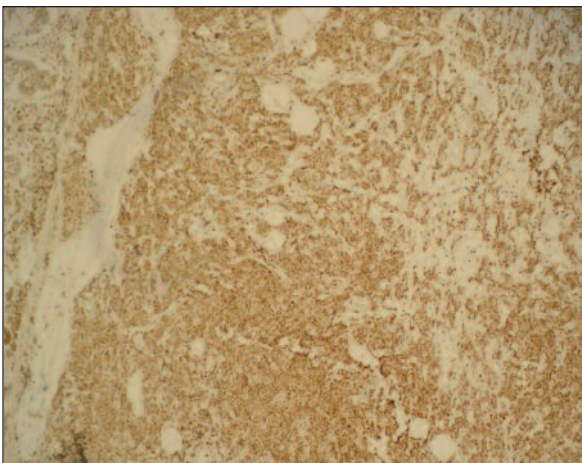
**FIGURE 4:** There was a uniform and homogeneous infiltrate of small non-cleaved follicular center cells that have several nucleoli and amphophilic cytoplasm. Numerous mitotic figures were present. The macrophages create a starry sky appearance at low power (HEX200).



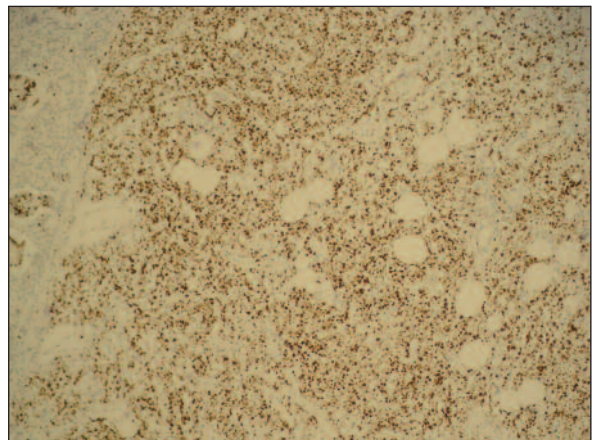
**FIGURE 5:** The tumor cells were CD 20 positive (x100).



**FIGURE 6:** Tumor cells were CD 10 positive (x200).



**FIGURE 7:** The tumor cells were bcl 2 positive (x100).



**FIGURE 8:** Ki 67 staining of more than %95 of the cells (x100).

## DISCUSSION

Burkitt lymphoma is a B cell non-Hodgkin lymphoma (NHL) type generally present with extranodal invasion. Most commonly, children and immune suppressant patients are affected by this disease.<sup>3</sup> Malignant lymphomas consist 1-4% of the all gastrointestinal malignant neoplasms.<sup>4</sup> Primary appendicular lymphoma is very rare and generally diagnosed in the postoperative period. In the study of Collins and colleagues conducted on 71 000 appendectomy specimen, they found primary appendicular lymphoma incidence approximately 0.015%.<sup>5</sup> In the study of Ruoff et al. conducted on 7970 appendectomy specimen, they detected appendix neoplasm in 51 cases. Only in 1 case, a lymphoma originating from the appendix has been diagnosed.<sup>6</sup> Acute appendicitis due to luminal obstruction is the most common clinical manifestation. Other clinical manifestations are: an

asymptomatic palpable mass, intussusception, gastrointestinal bleeding, ureteral obstruction, hematuria and incidental detection in imaging methods.<sup>7</sup> Diffuse wall thickening and hypoechoic appearance on ultrasound typically mimics cystic dilatation. The diameter of the lymphomatous appendix on CT is generally over 3 cm.<sup>8</sup> It is a sufficient surgical procedure in NHL cases originating from the appendix that do not exceed the submucosa.<sup>9</sup> Chemotherapy is the main basis of treatment in Burkitt lymphoma. The classic combination of CHOP has been in use for along time.<sup>10</sup>

Malignant neoplasms of the appendix are very rarely observed. They generally present as acute appendicitis. Preoperative diagnosis is very difficult. In this case report, we evaluated the Burkitt lymphoma of the appendix, which is very rare and which does not present with the picture of acute abdomen, and the short term results of this case.

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