Journal of Turgut Ozal Medical Center 2016;23(4):431-4
OLGU SUNUMU/CASE REPORT DOI: 10.5455/jtomc.2016.03.036

Ascites and hepatic involvement: a rare and atypical presentation of Burkitt Lymphoma

Burkitt lenfoma'nın asit ve karaciğer tutulumunun olduğu, nadir görülen atipik prezentasyonu

Yuksel Seckin[1], Yasir Furkan Cagin[1], Melda Comert Ozkan[2], Yilmaz Bilgic[1], Oguzhan Yildirim[1], Mehmet Ali Erdogan[1], Yahya Atayan[1], Hale Kirimlioglu[3], Ismet Aydogdu[4]

[1]İnönü University, Faculty of Medicine, Department of Gastroenterology, Malatya, Turkey
[2]İnönü University Medical Faculty, Department of Heamatology, Malatya, Turkey
[3]Acıbadem University Medical Faculty, Department of Pathology, Malatya, Turkey,
[4]Celal Bayar University Faculty of Medicine, Department of Heamatology, Malatya, Turkey

Abstract
Burkitt lymphoma (BL) is an uncommon form of lymphoma in adults. In this disease, extensive involvement of the omentum and the peritoneum is rare. We present this case with ascites because it is such a rare and overlooked clinical presentation of BL.
A previously healthy 20-year-old man was admitted to our hospital with abdominal pain, distension, and night sweats. Physical examination revealed massive ascites. Computerized tomography showed liver metastasis and ascites, but no pathologic thoracal or abdominal lympadenopathy was reported. Atypical lymphocytic cells with cytoplasmic vacuoles were determined on cytologic examination of ascites. We detected starry sky view on the bone marrow examination. On immunophenotyping, cells were CD20 positive, but negative for CD30, CD79a/Tdt. Patient was diagnosed as BL.
Burkitt lymphoma should be considered in young patients presenting with ascites and hepatic mass even without lymphadenopathy or focal gastrointestinal masses.
Keywords: Ascite; Hepatic İnvolvement; Burkitt Lymphoma.

Öz
Burkitt lenfoma (BL), erişkinlerdeki lenfomanın sık olmayan formlarından biridir. Bu hastalıkta omentum ve peritonun yaygın tutulumu nadirdir. Burkit lenfomanın nadir ve gözden kaçmış klinik sunumu ile gelen bu assitli olguyu sunuyoruz.
Daha önce herhangi bir hastalığı olmayan sağlıklı, 20 yaşındaki erkek hasta, karın ağrısı, şişkinlik ve gece terlemesi şikayetleri ile hastanemize başvurdu. Fizik muayenesinde assiti mevcut idi. Bilgisayarlı tomografide karaciğerde metastaz ile uyumlu lezyonlar ve asit izlenirken patolojik herhangi bir torakal ve abdominal lenfadenopati rapor edilmedi. Assit mayi sitolojisinde stoplazmik vakouller içeren atipik lenfositler izlendi. Kemik iliği aspirasyonunda yıldızlı gökyüzü manzarası gözlendi. İmmunofenotipinde CD20 pozitif iken, CD30, CD79a/Tdt negatif görüldü.
Assitli, lenfadenopatisiz karaciğer veya fokal gastrointestinal kitlesi olan genç hastalarda Burkitt lenfoması düşünülmesi gerekir. Burkitt lenfoma; lenfadenopati veya fokal gastrointestinal kitleler olmadan bile assit ve karaciğerde kitle ile başvuran genç hastalarda düşünülmelidir.
Anahtar Kelimeler: Assit; Hepatik Tutulum; Burkitt Lenfoma.

Received/Başvuru: 07.03.2016
Accepted/Kabul: 24.03.2016

Correspondence/İletişim
Yasir Furkan Cagin
İnönü University, Faculty of Medicine, Department of Gastroenterology Malatya, Turkey
E-mail: yafur@hotmail.com

How to cite this article/Atıf için
Seckin Y, Cagin YF, Ozkan MC, Bilgic Y, Yildirim O, Erdogan MA, Atayan Y, Kirimlioglu H, Aydogdu I. Ascites and hepatic involvement: a rare and atypical presentation of Burkitt Lymphoma. J Turgut Ozal Med Cent 2016;23(4):431-4.

2016;23(4):431-4
DOI:10.5455/jtomc.2016.03.036

INTRODUCTION

Burkitt lymphoma (BL) is an uncommon form of lymphoma in adults. Burkitt lymphoma is a highly aggressive B-cell malignancy with a high propensity for central nervous system involvement and abdominal spread, as well as a high incidence of tumor lysis syndrome due to rapid cell growth and turnover (1). BL is the most common form of childhood cancer in Central Africa and the sporadic North American form is very rare. Its most common presentation includes abdominal masses involving bowels, predominantly the cecum and the distal ileum along with extensive mesenteric and retroperitoneal lymphadenopathy (2,3).

BL can also involve the liver, spleen, kidneys and pancreas. Extensive involvement of the omentum and the peritoneum is rare (3). Peritoneum involvement is significantly associated with lymphadenopathy and ascites (4). We report a case who initially admitted with ascites, and then diagnosed as BL.

CASE REPORT

A previously healthy 20-year-old male was admitted to our hospital with abdominal pain, distension, and night sweats. Physical examination revealed hepatomegaly, reduced respiratory sound in the left lung and massive ascites. The abdomen was distended, tens, and tender. No cervical, axillary and inguinal lymphadenopaty was present. Complete blood count showed; white blood cells (WBC):13.300/mm^3, hemoglobin:14 g/dl, platelet:227.000 / mm3 and erythrocyte sedimentation rate: 13 mm\h. Biochemical parameters were as follows: albumin:3.4 g/dl, total bilirubin:0.37 mg/dl, direct bilirubin:0.12 mg/dl, aspartate aminotransferase (AST):85 U/L, alanine aminotransferase (ALT):23 U/L, alkaline phosphatase (ALP):76 U/L, gamma glutamyl transferase (GGT):24 U/L, lactate dehidrogenase (LDH): 2574 U/L, and uric asid acid:7.4 mg/dl. Viral markers were negative for HBsAg, anti-HCV, anti-HIV, Epstein–Barr virus (EBV). Carsino Carcino-embryonic antigen (CEA), alpha-fetoprotein (AFP), CA19-9 were within a normal range.

A contrasted thorax computerized tomography (CT) scanning revealed pleural effusion on the left lung and atelectasis on the left inferior lobe. Cranial magnetic resonance scanning was normal. Abdominal ultrasonography revealed multiple hypoecoic lesions (the largest size of lesions was 2.5 cm), massive ascites and omental and bowel-wall thickening. In addition, abdominal CT showed the thickening of the omentum, intestine and peritoneum. No cervical, abdominal and thoracal lymphadenopaty was revealed on in the CT. Esophagogastroduodenoscopy was normal. Paracentesis was performed. Macroscopic appearance of paracenteses fluid had intensive content and hemorragical. Ascite tests indicated WBC:130.300/mm^3, total protein:3.7 g/dl, albumin: 2.4 g/dl, LDH:26712 U/L, glucose:4 mg/dl. Serum-ascite albumin gradient was calculated as 1. Ascite resistance bacteria were not found. On cytologic examination of ascites, atypical lymphocytic cells were determined (Figure 1). Peripheral blood smear examination revealed cells with blastic appearance with vacuolated cytoplasm. The starry sky view was detected on the bone marrow examination (Figure 2). Immunophenotyping revealed that cells were CD10, CD20 positive, but negative for CD30, and CD79a/Tdt (Figure 3). The growth fraction (Ki-67) was 100%. The patient was diagnosed as BL. Spinal fluid examination was cytologically normal. Chemotherapy protocol including cyclophosphamide 1200 mg/m^2/day (1. day), daunorubycine 45 mg/m^2/day (1-3. days), vincristine 1.4 mg/m^2/day (1, 8, 15, 22. days), and prednisolone 60 mg/m^2/day (1-14. days) was initiated. CNS prophylaxis was done with intrathecal installations of cytosine arabinoside (30-45 mg/m^2 day 7) and methotrexate (12.5 mg day 10). After 14 days, abdominal ultrasonography revealed minimal ascites, and no lesion on the liver.

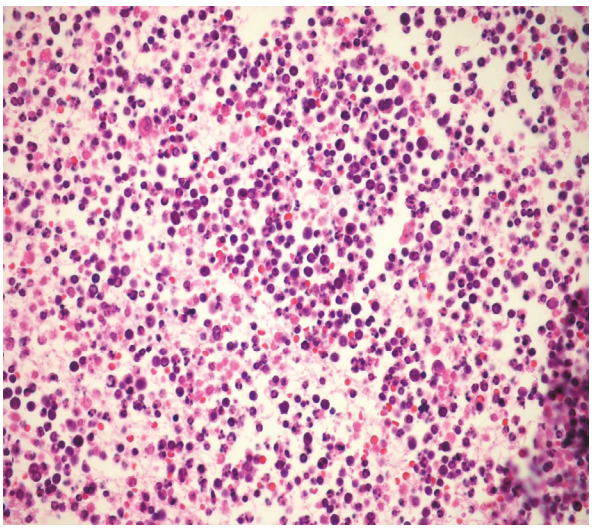

Figure 1. Atypical lymphocytes and apoptotic cells in cellular ascites fluid (H&E, × 200).

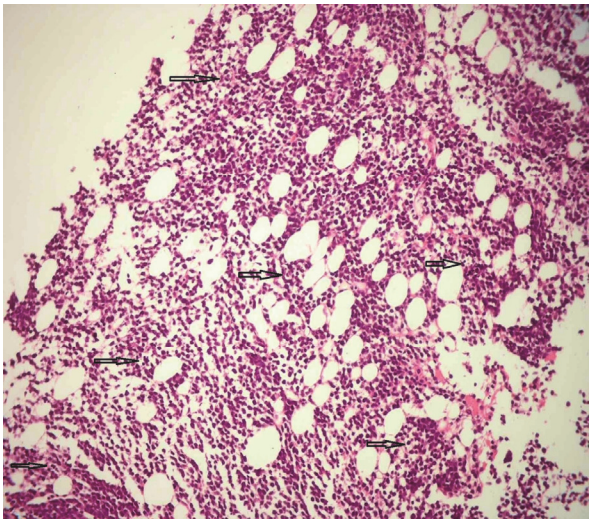

Figure 2. Starry sky view on the bone marrow examination (arrows) (H&E, × 100)

2016;23(4):431-4
DOI:10.5455/jtomc.2016.03.036

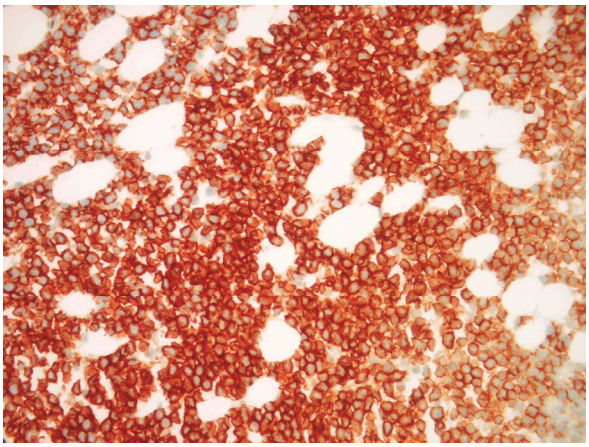

Figure 3. Tumor cells diffuse stained with CD20 antibody (Bone marrow section, streptavidin biotin, x200)

DISCUSSIONS

BL is a disease, with an incidence of approximately 1200 patients per year in the United States (5). Denis Burkitt first described the disease in children in 1958 (6). The tumor cells of classical BL have a characteristic morphology and immune profile. BL cells are mature B cells, positive for CD19, CD20, CD22, and CD79a, and have monotypic surface IgM (5). Furthermore, chromosomal translocations involving c-myc are a molecular hallmark of BL (7). The tumor cells are positive for B cell–associated antigens such as CD19, CD20, CD22, and CD79a (8). However, Burkitt lymphoma may indicate variations. Chu et al. showed in conditions including primary effusion lymphoma and plasmablastic lymphoma, both of which are commonly negative for CD19, CD20, and CD79a (9). Our present case had also pleural effusion and was negative for CD30, CD79a. While the Epstein-Barr virus (EBV) is associated with 98% of endemic BL, it is also seen in 20% of sporadic cases, and 30% to 40% of HIV-associated cases (10).

Clinical presentation and pathobiology of BL varies in different regions of the world. The greatest differences occur in the clinical presentation of African and American BL (11).While BL typically presents as a jaw mass in patients with the endemic variant (African), there is usually an intraabdominal disease in the more common sporadic type (American). Intraabdominal disease usually presents with lymphadenopathy or infiltration of the intestinal lymphoid tissue. Involvement of the omentum and the peritoneum is rare (3). When BL involves the peritoneum, its clinical sign is ascites. To distinguish between BL and the other causes of peritoneal carcinomatosis is difficult, if there is no lymphadenopathy in a patient with ascites. Ascitic fluid analysis is essential for the diagnosis of malignant ascites. The serum-to-ascites albumin gradient (SAAG) accurately identifies the presence of portal hypertension and is more useful than the protein-based exudate/transudate concept. The presence of a gradient of >1.1 g/dL indicates that the patient has portal hypertension with 97 percent accuracy. A gradient <1.1 g/dL indicates that the patient does not have portal hypertension. The gold standard for the diagnosis of malignant ascites is the presence of tumor cells in the ascidic fluid. Immunohistochemical staining combined with conventional cytologic examination increases the diagnostic sensitivity (12). Jahnke K et al. reported two cases of patients initially diagnosed as having diffuse large B-cell lymphoma (DLBCL), in whom the diagnosis had to be changed to BL and Burkitt-like lymphoma (BLL) following a reevaluation by a referral hematopathologist and due to the results of long distance polymerase chain reaction (PCR) of ascites lymphoma cells revealing a translocation involving the c-myc proto-oncogene (1). In the present case, patient had hepatic and peritoneal involvement, but no LAP lymphadenopathy. We performed bone marrow and immunhistochemical examinations for atypical lymphocytic cells were determined on cytologic examination of ascite, and then diagnosed BL. Multiple hypoechoic lesions in the liver were accepted as hepatic involvement of BL. Therefore, we did not need a liver biopsy. Primary hepatic lymphoma is a rare tumor that may present diagnostic difficulties to clinicians. Hepatic lesions can be single or multiple and clinically indistinguishable from any other primary liver tumor or metastatic disease. Primary hepatic lymphoma is defined as lymphoma either confined to the liver or having major liver involvement. Secondary liver involvement by lymphoma is common and can complicate treatment decisions (13).

CONCLUSION

It is difficult to distinguish BL from liver carcinomatosis secondary to an primary neoplasm. Therefore, BL should be kept in mind in young patients presenting with ascites and hepatic mass even without lymphadenopathy or focal gastrointestinal masses.

REFERENCES

1. Jahnke K, Burmeister T, Korfel A, Coupland SE, Thiel E. Long distance polymerase chain reaction of ascites lymphoma cells aids diagnosis establishment of abdominal Burkitt's lymphoma and Burkitt-like lymphoma. Leuk Lymphoma 2005;46(1):83-6.
2. Wong S, Sanchez TR, Swischuk LE, Huang FS. Diffuse peritoneal lymphomatosis: atypical presentation of Burkitt lymphoma. Pediatr Radiol 2009;39(3):274-6.
3. Horger M, Müller-Schimpfle M, Yirkin I, Wehrmann M, Claussen CD. Extensive peritoneal and omental lymphomatosis with raised CA 125 mimicking carcinomatosis: CT and intraoperative findings. Br J Radiol 2004;77(913):71-3.
4. Vade A, Blane CE. Imaging of Burkitt lymphoma in pediatric patients. Pediatr Radiol 1985;15(2):123-6.
5. Perkins AS, Friedberg JW. Burkitt lymphoma in adults. Hematology Am Soc Hematol Educ Program 2008;341-8.
6. Burkitt D. A sarcoma involing the jaws in African children. Br J Surg 1958;46:218-23.
7. Akasaka T, Muramatsu M, Ohno H, Miura I, Tatsumi E, Fukuhara S. Application of long-distance polymerase chain reaction to detection of junctional sequences

2016;23(4):431-4
DOI:10.5455/jtomc.2016.03.036

created by chromosomal translocation in mature Bcell neoplasms. Blood 1996;88:985–94.
8. Kelemen, Katalin. Immunophenotypic variations of Burkitt lymphoma. Am J Clin Pathol 2010;134(1):127-38.
9. Chu PG, Loera S,Huang Q, et al. Lineage determination of CD20– B-cell neoplasms: an immunohistochemical study. Am J Clin Pathol 2006;126:534–44.
10. Brady G, MacArthur GJ, Farrell PJ. Epstein-Barr virus and Burkitt lymphoma. J Clin Pathol 2007;60:1397-402.
11. Kamona AA, El-Khatib MA, Swaidan MY, Jarar MS, Suleiman AJ, Ali HM. Pediatric Burkitt's lymphoma: CT findings. Abdom Imaging 2007;32(3):381-6.
12. Aslam N, Marino CR. Malignant ascites: new concepts in pathophysiology, diagnosis, and management. Arch Intern Med 2001;161(22):2733-7.
13. Salmon JS, Thompson MA, Arildsen RC, Greer JP. Non-Hodgkin's lymphoma involving the liver: clinical and therapeutic considerations. Clin Lymphoma Myeloma 2006;6(4):273-80.