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

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

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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 lymphadenopathy 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: Ascites; Hepatic Involvement; Burkitt Lymphoma.

Öz


Assiti, lenfadenopatisiz karaciğer veya fokal gastrointestinal kiti ile başvuran genç hastalarda Burkitt lenfoması düşünülmesi gerekiyor. Burkitt lenfoma; lenfadenopati veya fokal gastrointestinal kitieller olmadan bile asit ve karaciğerde kitle ile başvuran genç hastalarda düşünülmelidir.

Anahtar Kelimeler: Assit; Hepatik Tutulum; Burkitt Lenfoma.
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 lymphadenopathy was present. Complete blood count showed; white blood cells (WBC): 13,300/mm³, 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 dehydrogenase (LDH): 2574 U/L, and uric acid: 7.4 mg/dl. Viral markers were negative for HBsAg, anti-HCV, anti-HIV, Epstein–Barr virus (EBV). Cansino 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 hypoechoic 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 thoracic lymphadenopathy was revealed on the CT. Esophagogastroduodenoscopy was normal. Paracentesis was performed. Macroscopic appearance of paracenteses fluid had intensive content and hemorrhagical. Ascite tests indicated WBC: 130,300/mm³, 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/m²/day (1. day), daunorubycine 45 mg/m²/day (1-3. days), vincristine 1.4 mg/m²/day (1, 8, 15, 22. days), and prednisolone 60 mg/m²/day (1-14. days) was initiated. CNS prophylaxis was done with intrathecal installations of cytosine arabinoside (30-45 mg/m² day 7) and methotrexate (12.5 mg day 10). After 14 days, abdominal ultrasonography revealed minimal ascites, and no lesion on the liver.
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% 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 ascitic fluid. Immunohistochemical staining combined with conventional cyologic 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 immunohistochemical 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