

# Primary Ovarian Burkitt's Lymphoma Mimicking an Advanced Ovarian Tumor: A Case Report

## İlerlemiş Bir Ovaryan Tümörü Taklit Eden Primer Ovaryan Burkitt Lenfoma: Bir Vaka Takdimi

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#### Abstract

Primary ovarian Burkitt's lymphoma (BL) is a rarely seen neoplasm and the diagnosis is revealed histologically. A 23-year-old multigravid woman was admitted to our clinic, complaining with abdominal distension, pelvic pain and high body temperature. In this case we present our approach and the management of this pelvic mass was concluded as a BL before the laparotomy. The diagnosis of BL was established from the frozen specimen during exploratory laparotomy. Combined chemotherapy was administered to the surgery treatment. Although rare, BL should be consider in differential diagnosis of a pelvic mass in young patients.

Keywords: **Epstein-Barr Virus Infections; Lymphoma; Ovary.**

#### Özet

Primer ovariyen Burkitt's lenfoma (BL) nadir görülen neoplazmdir ve tanısı histolojik olarak gösterilmektedir. 23 yaşında multigravid kadın abdominal distansiyon, pelvik ağrı ve yüksek ateş şikayetleriyle kliniğimize başvurmuştur. Bu vakada laparotomi öncesi BL olarak düşünülen pelvik kitlenin yönetimini ve bizim yaklaşımımızı sunduk. BL tanısı laparotomi sırasında frozen materyalinden saptanmıştır. Cerrahi tedaviye kombine kemoterapi eklenmiştir. Nadir görülmesine karşın BL genç hastalardaki pelvik kitlenin ayırıcı tanısında göz önünde tutulmalıdır.

Anahtar Kelimeler: **Epstein-Barr Virus Enfeksiyonları; Lenfoma; Over.**

## Introduction

Primary ovarian lymphoma is a rare disorder, accounting for 0.5% of all non-Hodgkin's lymphomas and 1.5% of all ovarian neoplasms. In some autopsy series, ovarian involvement was observed with a frequency as high as 26% in patients with clinically occult or overt lymphomatous malignancy (1). Malignant lymphoma of the ovary can present either as a primary lesion or more frequently as a secondary involvement. Distinguishing primary ovarian lymphoma from secondary involvement is not always easy, especially in the advanced cases. Primary ovarian lymphoma is an extranodal disease and it is uncommon with only few reported cases (2- 4). The diagnosis of ovarian lymphoma is almost invariably not made until the tumor has been examined histologically. Burkitt's lymphoma has characteristic cytogenetics, usually t(8; 14), involving the 'c-myc' oncogene. Epstein- Barr virus (EBV) plays a major role in its pathogenesis. All EBV-associated malignancies have a predominantly latent pattern of viral gene expression. However, many EBV-associated tumors are poor targets for the cellular immune response suggesting that immune evasion strategies might favor tumor development (5). We report a case of primary ovarian Burkitt's lymphoma initially presented with bilateral ovarian masses, diagnosed by combined analysis, and successfully managed with a combination of surgery and adjuvant chemotherapy.

## Case Presentation

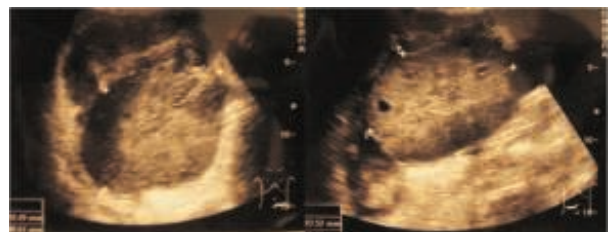
A 23-year-old woman, gravida 1, para 1, was presented with complaints of abdominal distension, pelvic pain and high body temperature. Physical examination revealed abdominal distention due to tumor and ascites. Ultrasonography disclosed bilateral abdominal masses, each measuring 10 cm in diameter with solid and cystic components and a large amount of ascites and pleural effusion (Picture 1). Abdominal computer tomography (CT) additionally showed omental cake appearance but no enlarged lymph nodes. Cytologic evaluation of ascites and pleural effusions were negative for malignancy. Laboratory studies showed elevated levels of serum lactate dehydrogenase (1187 U/L; normal <465U/L) and CA-125 (432 U/mL; normal <35 u/mL) and normal levels of CA19-9 (7.51U/mL); carcinoembryogenic antigen (1.14 U/mL) and alfa-feto-protein (0.9 ng/ml). She had a negative human immunodeficiency virus (HIV) test. Meigs' syndrome, disgerminoma, gastro-intestinal tumor metastasis, pelvic tuberculosis and ovarian lymphoma were included in the differential diagnosis. Bone marrow aspiration, rectosigmoidoscopy and skin tuberculin test

were all within normal limits. EBV EBNA IgG and EBV VCA IgG were positive. Therefore, Burkitt's lymphoma was considered in the pre-operative diagnosis and an exploratory laparotomy was performed.

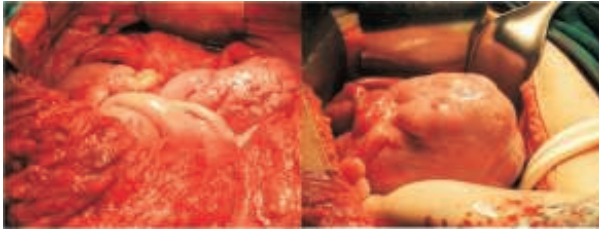
Bilateral ovarian mass in 10 cm diameter were fragile, edematous, and solid-cystic nature and appeared together with 5500 ml of serous ascites during the operation. The Fallopian tubes were elongated and there were diffuse nodular lesions on the surface of the peritoneum. The omentum was diffusely involved by the tumor, with changes showing tumor caking and appendix was fragile and edematous (Picture 2). The liver contained multinodular masses. Spleen and diaphragm were not involved macroscopically. Frozen section of the sample from right ovary revealed malignant lymphoma. Then, total abdominal hysterectomy bilateral salpingo-oophorectomy was performed.

Blastic appearing on microscopic evaluation the tumors composed of a monotonous population of intermediate size, lymphoid cells that showed brisk mitotic activity. Interspersed macrophages imparting a starry-sky pattern were also present. The neoplastic cells expressed CD 20 and showed high proliferative activity with MIB1 (K\_67). The other markers, CD 3, CD 5, and CD 10, were negative. But cytogenetically translocation t(8;14) and t(14;18) were not detected. The clinical stage was IV according to the Ann Arbor system and IIIc according to the International Federation of Gynecology and Obstetrics. The patient began treatment with cyclophosphamide, adriamycin, vincristine, L-asparaginase, and prednisolone for six cycles.

After the chemotherapy the patient was in complete remission without any clinical and laboratory evidence of primary disease. An autologous bone marrow transplantation was performed 3 months after the completion of the treatment.



**Picture 1.** Right and left ovarian ultrasound photographs.



**Picture 2.** Bilateral ovarian masses diffuse omental involvement and diffuse nodular lesions on the surface of the peritoneum.

### Discussion

Burkitt's lymphoma affects mainly children; however, the age range is 3 to 63 years. The median age is 7 for African and Middle Eastern cases and 12 for nonendemic cases (6). The majority of Burkitt's lymphoma cases are male. Jaw and facial bones as well as mesentery and gonad are often involved in endemic cases, whereas the majority of nonendemic cases present with abdominal disease, involving ileum, caecum, mesentery, kidney, liver, and ovary. Immunodeficiency-associated Burkitt's lymphomas are often nodal. Most patients with abdominal Burkitt's lymphoma present with multiple tumors involving many organs including ovaries. Unilateral ovarian involvement is rare in BL cases. Primary ovarian Burkitt's lymphomas are usually bilateral and are usually associated with ascites. African-type Burkitt's lymphoma is frequently associated with EBV. Some nonendemic cases are also EBV-positive (7). Our case was associated with EBV based on EBV EBNA IgG and EBV VCA IgG positive. EBV EBNA IgG and EBV VCA IgG were laboratory tests of EBV latently infected cells. All EBV-associated malignancies have a predominantly latent pattern of viral gene expression. Therefore EBV EBNA IgG and EBV VCA IgG are important markers to discuss the primary ovarian Burkitt's lymphoma for the differential diagnosis like our case. The diagnostic work-up of Burkitt's lymphoma includes histopathologic, immunohistochemical, and genetic studies. Due to its unique morphology and immunohistochemical characteristics Burkitt's lymphoma can easily be distinguished from other non-Hodgkin lymphomas and epithelial/stromal neoplasms of the ovary. The tumors are composed of small noncleaved lymphoblastic cells and show a characteristic starry-sky pattern. Some cases have larger cells and resemble diffuse large B-cell lymphoma. Such cases are called non-Burkitt or Burkitt-like lymphoma (3). In our case, the tumors composed of a monotonous

population of intermediate size, blastic appearing lymphoid cells showed brisk mitotic activity and a starry-sky pattern were also present. According to the latest WHO classification, a tumor that contains cells larger than typical BL cells but has high proliferation rate and shows t(8;14) should also be classified as BL (8). The morphologic impression can be confirmed with immunohistochemistry. Neoplastic lymphoid cells are reactive for CD20 and demonstrate high proliferative activity with MIB1 (Ki67). Other markers, such as CD5, CD21, CD23, CD34, TdT, and CD99, are negative. Detection of c-myc gene-related translocations, i.e., t(8;14), t(14;18), and t(8;22), may also be used to confirm the diagnosis. Burkitt's lymphoma is a rapidly growing tumor and patients usually have high serum LDH levels. With aggressive chemotherapy regimens Burkitt's lymphoma is a potentially curable malignancy. In the differential diagnosis, we thought that ovarian cancer might be existed in this patient because of presentation of pelvic mass, ascites and hydrothorax. However bilateral ovarian fibroma was not expected due to the high level of LDH and bilateral ovarian involvement. We thought that this pelvic mass might be a dysgerminoma. However, ascites was not common in patients with dysgerminoma. Additionally, we thought that this pelvic mass might be metastasis of gastro-intestinal tumor because ascites, hydrothorax and bilaterally ovarian involvement were present. But rectosigmoidoscopy examination of colon was normal. Lastly when we looked at literature, this pelvic mass might be ovarian lymphoma because bilateral ovarian involvement, ascites and hydrothorax were present with EBV EBNA IgG and EBV VCA IgG positivity and tumor markers of CA-125 and LDH levels were high. Therefore we aspirated intraperitoneal fluid and examined the cytology of this fluid. However cytology examination of this fluid was normal. And bone marrow aspiration was studied but the result was normal so a secondary involvement was differentiated. Therefore, prior diagnosis is a primary ovarian Burkitt's lymphoma. Although rare, BL should be kept in mind when isolated ovarian tumors are detected in young patients. And EBV EBNA IgG and EBV VCA IgG may be significant markers in the ovarian Burkitt's lymphoma for differential diagnosis of the ovarian masses.

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