**Burkitt’s Lymphoma Mimicking a Primary Gynecologic Tumor**

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**SUMMARY**

**Objective:** Burkitt’s lymphoma (BL) occurs mostly in children; bilateral ovarian involvement mimicking a gynecologic malignancy in adults is extremely rare. Here, we report a patient with BL mimicking a gynecologic tumor. **Case Report:** A 50-year-old Taiwanese woman presented with the complaint of persistent lower abdominal distension with dull pain, easy satiety, and progressively increasing abdominal girth for 2 weeks. Amenorrhea was also noted for about 2 months, and her review of systems was negative for the common “B” symptoms associated with lymphoma. At our hospital, imaging studies revealed a huge pelvic mass (10.8 × 8.7 cm), suggesting a large subserous myoma or an ovarian tumor. Under the impression of pelvic mass, she underwent exploratory laparotomy. Primary ovarian sex-cord malignancy with cecum involvement was impressed by the primitive intra-operative frozen section report. Subsequently, an optimal cytoreductive operation with right hemicolecotomy was performed. However, final histopathologic report was an extranodal multifocal BL. **Conclusion:** Although extranodal BL in ovaries is a rare condition, it should be noted in the differential diagnosis of pelvic gynecologic malignancies. [*Taiwanese J Obstet Gynecol* 2006;45(2):162–166]

**Key Words:** Burkitt’s lymphoma, pelvic mass

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**Introduction**

Burkitt’s lymphoma (BL) was first described by Burkitt in 1958 [1]. It is a highly aggressive non-Hodgkin lymphoma, often presenting in extranodal sites or as an acute leukemia [1]. Adult patients with sporadic BL often present with extranodal disease, and the abdomen is the most frequent site of involvement [2]. However, malignant lymphomas involving the female genital tract are rare, and <1% of patients initially present with ovarian enlargement [3]. Although ovarian lymphoma is a disease with poor prognosis, it is reported that successful results are possible with chemotherapy and surgery [4]. Nevertheless, an extranodal BL presenting with signs and symptoms mimicking a primary gynecologic tumor is extremely rare. In this article, we present a case of BL mimicking a gynecologic malignancy. In addition, the patient did not have the common “B” symptoms associated with lymphoma, such as fever, weight loss, night sweats, and fatigue.

**Case Report**

A 50-year-old Taiwanese woman visited her primary care physician with the major complaints of persistent lower abdominal bloating sensation with dull pain, easy satiety, and progressively increasing abdominal girth for 2 weeks. Amenorrhea was also noted for about 2 months, and she was transferred to our outpatient clinic due to a pelvic mass detected by ultrasonography.

At our hospital, the review of systems for this patient was negative for fever, chills, weight loss, night sweats, nausea, vomiting, and bladder symptoms. Her annual Pap smears were all normal and her past history was unremarkable. Physical examination revealed a low abdominal pelvic mass, approximately 20 weeks’
gestation in size, with firm consistency. The mass was fixed to the uterus, and there was no cul-de-sac nodularity. A pelvic high-resolution real-time transvaginal ultrasound revealed a large homogeneous pelvic mass (10.8×8.7 cm), extending to the uterus, with mild enlarged left ovary and minimal cul-de-sac fluid (Figure 1). Power Doppler ultrasonography showed no blood flow signals in the mass. Besides, all the routine blood tests and biochemical markers were normal before surgery. Initially, a pelvic tumor of subserous myoma or of ovarian origin was suspected.

She underwent an exploratory laparotomy. A large fragile yellowish right ovarian tumor, 15×15 cm in size, with adhesion to the uterus, was noted during surgery. Cecal area involvement was also noted. The primitive report of frozen section showed an ovarian malignancy of sex cord-stromal origin. Therefore, she received optimal debulking surgery, including total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, excision of the peritoneal tumor, pelvic and para-aortic lymph node sampling, as well as right hemicolectomy.

Figure 1. (A) Transabdominal ultrasound detected a large mixed echogenic pelvic mass of irregular shape (10.8×8.7 cm). (B) Transvaginal ultrasound revealed a huge solid pelvic mass, extending from the right anterior wall of the uterus, with homogeneous echogenicity and some irregular anechoic areas within the mass. Minimal cul-de-sac fluid was also noted.

Figure 2. Gross findings. (A) Large fragile yellowish irregular surface tumor (about 15 cm in diameter) originating from the right ovary. (B) Smaller yellowish fragile tumor about 5 cm in diameter originating from the left ovary. In contrast, the uterus and cervix are normal. (C) Tumor about 8 cm in diameter originating from the junction of the terminal ileum and ascending colon.
The final pathologic report revealed a multifocal BL, involving bilateral ovaries, cervix, peritoneal cavity, terminal ileum, and ascending colon (Figure 2). All lymph nodes dissected were negative for tumor metastasis. Microscopically, a typical “starry sky” appearance was clearly noted (Figure 3A). With advanced immunohistochemical staining, tumor cell markers revealed LCA(+), CD3(–), CD20(+), CD10(+), Bcl-2(–), Bcl-6(+), and CD5(–) (Figure 3B). The Ki-67 proliferative index of tumor cells was > 90% (Figure 3C). In addition, Epstein–Barr virus (EBV) glycoprotein immunoglobulin G (IgG) was positive (1:320). Moreover, EBV-encoded small nonpolyadenylated RNA-1 (EBER-1) in situ hybridization showed a positive result (Figure 3D). Nevertheless, bone marrow biopsy was negative for tumor involvement.

The patient recovered very well after the operation and was discharged from our hospital 13 days later. She was referred to medical oncology for further chemotherapy.

**Discussion**

BL, an endemic disease of central Africa, is a highly undifferentiated B-cell lymphoma [1]. BL mainly affects children. However, the affected ages range from 3 to 63 years old. As BL has a remarkably high growth rate with a doubling time of 24 hours, early diagnosis and immediate treatment are very important.

Cases from other parts of the world share the same histopathology as that of African origin. Although they have the same histology and biologic behaviors, to date, the epidemiologic features, clinical presentations, and genetic alterations separate BL into three main categories: (i) endemic (African), (ii) sporadic (non-endemic/non-African), and (iii) immunodeficiency-associated BL. The endemic form shows frequent involvement of the jaw and kidneys. In contrast, most patients with sporadic BL present with multiple abdominal tumors involving many organs [1,5,6]. Immunodeficiency-associated BL is often nodal and frequently observed.
in patients with human immunodeficiency virus infection [7,8].

BL symptoms vary, with abdominal pain, nausea, vomiting, bowel obstruction, gastrointestinal bleeding, or syndromes mimicking acute appendicitis or intussusception. At the time of diagnosis, patients usually already have bulky masses, high lactate dehydrogenase and uric acid levels. Bone marrow and central nervous system (CNS) involvement have been reported in 30–38% and 13–17% of adults, respectively [9,10]. Although ascites is common, unilateral ovarian involvement is extremely rare in BL. Because of hypervascularity and increased weight of the ovaries, there is a tendency to partial torsion and intermittent lower quadrant pain [7]. However, in our case, there was no bone marrow and CNS involvement, and no ovarian torsion.

In imaging studies, lymphomatous involvement of ovaries in BL is usually seen as unilateral or bilateral masses. Nevertheless, there are no specific imaging characteristics that can differentiate BL from other solid neoplasms [11]. Pelvic high-resolution real-time transvaginal ultrasound may depict a nonspecific hyperechoic mass. Color or power Doppler ultrasound shows only mild vascularization. In our case, no blood flow was visualized on power Doppler.

Although BL can be distinguished from other tumors based on its unique morphology and immunohistochemical characteristics, a definitive diagnosis based on frozen section only is usually very difficult. The frozen section of BL is usually misinterpreted as sex cord-stromal group of tumors due to its sheet or cord patterns and its rarity, just as in our case.

The final diagnostic work-up of BL might include histopathologic and immunohistochemical staining. In histopathology, medium-sized cells, with abundant basophilic cytoplasm (often containing lipid vacuoles), round nuclei with clumped chromatin, and multiple nucleoli with a diffuse, monotonous pattern of infiltration, are characteristics of classic BL. A “starry sky” appearance has been described in BL because of its abundant proliferative rate, frequent apoptosis, and numerous macrophages containing ingested apoptotic tumor cells [1]. All the above-mentioned histopathologic characteristics of our case are shown in Figure 3A.

The histopathologic impression of BL can be further confirmed with immunohistochemistry. BL cells may express surface IgM, Bcl-6, CD10, CD19, CD20, CD22 and CD79a, and may also demonstrate high-proliferative activity with Ki67 [2]. The Ki-67 proliferative index of tumor cells was > 90% (Figure 3C).

Although EBV is strongly implicated in the pathogenesis of African BL, only 20% of EBV involvement is detected in the non-African variants or sporadic BL samples [8]. EBER in situ hybridization was conducted to determine the presence of EBV in neoplastic cells. Besides, the inhibition of viral proteins (EBNA-1 and EBNA-2, in particular) can inhibit cell growth and lead to apoptosis of EBV-immortalized cells in vitro [2]. The EBV studies in our case further confirmed the role of EBV in the pathogenesis of BL. EBER-1 in situ hybridization revealed a positive result (Figure 3D) and the EBV glycoprotein IgG was also positive (1:320).

It is still controversial as to whether BL therapy should incorporate debulking surgery, because there is highly effective chemotherapy and an increased rate of mortality and morbidity associated with surgery [2]. However, Dao reported that successful cure of BL is possible with chemotherapy and surgery together [4]. Therefore, our BL patient undergoing operation is justified. For further chemotherapy, the patient was transferred to the Department of Medical Oncology.

In conclusion, an extranodal BL presenting with signs and symptoms mimicking a primary gynecologic tumor is extremely rare. In this article, we present a case of BL mimicking a gynecologic malignancy before surgery. In addition, the patient did not have the common “B” symptoms associated with lymphoma, such as fever, weight loss, night sweats, and fatigue. Although extranodal BL in ovaries is a rare condition, it should be included in the differential diagnosis of pelvic gynecologic tumors.

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References