

RESEARCH ARTICLE

Burkitt's Lymphoma Highly Mimicking Advanced Ovarian Cancer: A Case Report and Literature Review

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Citation: Jie Li, Xin yuan Chen, Ling Xi [2022] Burkitt's lymphoma highly mimicking advanced ovarian cancer: A case report and literature review. *J Gynecology Women Health care* 4: 103

Abstract

Primary ovarian lymphoma is a rare manifestation of non-Hodgkin's lymphoma [NHL], which accounts for 0.5% of NHL and 1.5% of ovarian tumors. Here, we present a case of a 30-year-old woman who complained of abdominal distention, diagnosed with primary bilateral ovarian Burkitt's lymphoma [BL] eventually. Ultrasonography showed a bilateral ovarian mass, the left measuring 4.8*4.6cm, the right ovary measuring 8.0*5.9cm, and moderate ascites. Positron emission tomography-computed tomography [PET-CT] scan revealed high metabolism conditions in pelvic mass, cardiocostophrenic angle, thickened peritoneum, and right-side pleural effusion and ascites. During hospitalization, the patient underwent exploratory laparotomy. Pathological examination of the biopsy sample of omentum showed the final diagnosis as BL. Then the patient commenced seven cycles of chemotherapy for one year and received autologous stem cell transplantation [ASCT]. The patient remained disease-free for six years and has kept follow-up since then. Primary ovarian BL is rare, especially for adult women, and its clinical features are highly mimicking ovarian cancer, which has a higher incidence in them. Clinicians need to improve the clinical evaluation for differential diagnosis before surgery. This study offers insights into the differential diagnosis and prognosis factors in primary ovarian Burkitt's lymphoma patients.

Keywords: Burkitt's lymphoma; ovarian cancer; ovary; lymphoma

Abbreviation: BL: Burkitt lymphoma; ASCT: Autologous stem cell transplantation; PET-CT: Positron emission tomography-computed tomography; CNS: the central nervous system; NHL: non-Hodgkin's lymphoma; LDH: lactate dehydrogenase; UA: uric acid; CEA: Carcinoembryonic Antigen; AFP: Alpha-fetoprotein; CA: Cancer Antigen

Introduction

BL as an exuberant, rapidly aggressive, and unique B-cell neoplasm, is the fastest growing human tumor, characterized by a translocation between c-myc and heavy immunoglobulin loci, such as t[8;14] [q24;q32], t[2;8][p12;q24] or t[8;22][q24;q11]. BL is more likely to involve in children, especially boys. Three distinct clinical features of BL can be recognized as endemic, sporadic and immunodeficiency associated.

Primary ovarian lymphoma is a rare manifestation of non-Hodgkin's lymphoma [NHL]. Primary ovarian lymphoma accounts for 0.5% of NHL and 1.5% of ovarian tumors. A possible reason would be that scarce lymphoid tissue exists in the ovary [1]. Ovary has less possibility of being the primary site for BL, which tends to spread to extranodal sites like the central nervous system, bone marrow, mesentery, kidney, breast, testis, and ovary [2]. Ovary involvement occurs mostly in nonendemic form, presented as secondary, as a part of systemic disease. Primary bilateral ovarian BL in adult patients highly mimicking advanced ovarian cancer is unfrequent. Most primary ovarian BL patients' symptoms are not classic "B cells" related, like fever, weight loss, and night sweats. The first complaints about these patients may be gynecology-related symptoms, like pelvis or an abdominal mass, abdominal pain, and distention. The differential diagnosis between ovarian cancer and primary ovarian BL is challengeable. Misdiagnosis may bring unnecessary chemotherapies to these patients, like paclitaxel, cisplatin, carboplatin, and surgeries like hysterectomy, omentectomy, adnexal removal, and lymphadenectomy, which delays their opportunities to receive chemotherapy. In this report, we describe the case of a 30-year-old woman, diagnosed with primary bilateral ovarian BL, treated with chemotherapy and surgery, has kept disease-free for seven years.

Case Presentation

A 30-year-old G2P2 female complained of abdominal distention for half a month to the clinic. She complained of shortness of breath but denied other complaints like nausea, vomiting, diarrhoea, and chest pain. Her medical history was nothing remarkable. The transvaginal ultrasonography showed a bilateral ovarian mass, the left measuring 4.8*4.6cm, the right ovary measuring 8.0*5.9cm, and moderate ascites. A Positron emission tomography-computed tomography [PET-CT] scan revealed high metabolism conditions in pelvic mass, cardiocostophrenic angle, thickened peritoneum, and right-side pleural effusion and ascites. Laboratory test demonstrated serum levels of Cancer Antigen [CA]125 elevated to 2225.30 U/ml and CA199, Carcinoembryonic Antigen [CEA], Alpha-fetoprotein [AFP] was within normal range. Complete blood and chemistry profiles were average. Additionally, uric acid was 805 umol/L.

She was suspected of having ovarian cancer and underwent an exploratory laparotomy. Upon entry into the abdomen, the greater omentum was closely adherent to the peritoneal wall. Scattered friable metastasis was found all over the pelvis, prone to bleed after contact. No visible metastasis could be seen in the liver and the near space. Further exploration was discontinued for massive mass taken over all the pelvis. Pathologic examination of the greater omentum and ascites was consistent with a "starry sky" appearance typical of Burkitt's lymphoma. Histologic staining was positive for CD20, CD79, PAX-5, CD10, CD45, PAX-8, C-myc, BCL-6, and Ki67, and negative for BCL-2, TdT, CD99, CD2, CD3, CK5/6, PR, S-100, PR and cyclin D1. Postoperative examination showed no metastatic disease in the central nervous system [CNS] or bone marrow.

Then the patient began combined chemotherapy, including cyclophosphamide, doxorubicin, vincristine, and dacarbazine. Seven cycles of chemotherapy were continued for almost one year, and after that, the patient received ASCT. After treatment, transvaginal ultrasound showed no tumor-like disease, and a mass in the pelvis was detected. She was then discharged for follow-up once a year. It has now been over seven years since primary ovarian BL was diagnosed. During this time, the patient remained disease-free.

Discussion

As the difficulty in the differential diagnosis between primary ovarian BL and advanced ovarian cancer, we summarised the clinical features and prognosis factors in primary ovarian BL cases reported, as shown. Symptoms and signs in primary ovarian BL are mostly gynecology-related. Most patients' complaints would be abdominal pain, palpable abdominal mass, and abdominal distention for pelvis mass. Some patients' complaints would be fatigue, weakness, shortness of breath, and lack of appetite related to ascites or pleural fluids. "B cells" symptoms are notable in advanced-stage patients, like fever, weight loss, and night sweats. Misdiagnosis may happen to patients with amenorrhea. Even if the woman is pregnant, the diagnosis of primary ovarian BL should not be excluded unless there is no other evidence. In the cases reported, the most common symptom is abdominal pain [3-6].

Primary ovarian lymphoma diagnosis criteria were suggested by Fox et al.: 1 the disease confined to the ovary or spread has occurred to immediately adjacent tissues; 2 absence of disease in blood or bone marrow; 3 nos extra ovarian lymphomatous lesions, if any, should appear at least after few months [7]. As markers to differentiate primary ovarian BL and ovarian cancer, gynecological oncology-related markers help. CA125 elevation with normal HE4, CA199, AFP, and β -HCG could be seen in most primary BL [8]. CA125 elevation could be seen in benign and malignant diseases, like endometriosis, pelvis inflammation disease, pulmonary tuberculosis, tuberculous peritonitis, NHL, acute leukemia, and hepatocellular carcinoma, melanoma, pancreatic tumors [6]. Additionally, high serum CA125 could be an important prognostic factor in NHL, correlating with advanced disease, low response rates, and worse survival. Patients with higher CA125 serum levels suffered from advanced disease, bone marrow involvement, extranodal involvement, poor performance status, the presence of B symptoms, and high serum lactate dehydrogenase [LDH] level [9]. In this way, CA125 measurements may be used for diagnosis, staging, monitoring response to treatment, and follow-up of patients with primary ovarian BL. Increased LDH level helps in diagnosis, staging, monitoring response to treatment, and follow-up likewise. Interestingly, as evidenced in this case, some patients show uric acid [UA] elevation, with the highest one reported as 334 $\mu\text{mol/L}$ [10]. UA is associated with acute kidney failure and tumor lysis syndrome, and an elevated UA level within the first 24 h of hospitalization was associated with a more significant decline in eGFR 1 month after hospital dismissal [11,12]. Great care should be taken to patients with UA elevation.

Most patients underwent surgery for a precise diagnosis [3, 13-14, 15]. During surgery, surgeons may be extremely cautious since most cases indicate that ovarian BL is highly friable, and resection is needed for hemostasis. Grossly, the ovarian mass is grey or off-white, hematogenous, soft pultaceous material [3]. If the tumor is bilateral, the relative non-affected one also needs a biopsy [16].

The mainstay of treatment for BL is multiagent chemotherapy, with a 5-year survival rate approaching 75% [4, 17, 18]. Whether debulking surgery is beneficial for patients remains controversial. However, extensive tumor debulking maybe not be the best solution for ovarian mass, ascites and other clinical complications. During the time between post-operation and chemotherapy, great care should be taken to rapid tumor growth and deteriorated patients' physical condition. After commencing chemotherapy, there could be a significant improvement in patients' condition [19-20]. As evidenced in this case and cases reported above, tumor size will dramatically reduce after chemotherapy [8, 21].

As shown in most primary ovarian BL patients are adult women with a range from 4 to 50 [mean age: 22.05 years]. The ovarian mass tends to be bilateral and huge. Cases reported showed that most patients had a great response to chemotherapy, although BL displays a high proliferation rate [21]. Prognosis factors in BL include advanced age, advanced stage, poor performance status, CNS or bone marrow involvement, anemia, the presence of circulating blasts, and an LDH elevation [2]. Prognosis HIV features may not influence the prognosis of patients undergoing contemporaneous treatment [22]. Patients, both HIV infected or not, could have only pelvis mass-related symptoms in the cases reported before [4].

Pregnancy may be associated with primary ovarian BL since it can impair host immunity. Primary ovarian BL in pregnant women may have a worse prognosis for the increased tumor growth speed and rapid spread. For these patients, pregnancy was terminated since the chemotherapy was toxic to the fetus [23]. In patients wishing for fertility preservation, immediate action must be taken. In ovarian BL patients, enlarged ovary and reduced ovarian reserve made spontaneous conception difficult [24]. A short time in-

terval after chemotherapy may be beneficial if spontaneous pregnancy is attempted. Complete remission of lymphoma may result in a higher possibility for spontaneous conception. Cryopreservation of ovarian tissue with future re-transplantation maybe not be safe in ovarian BL [24].

In this case, we could see that primary ovarian Burkitt's lymphoma mimics ovarian cancer, and its diagnosis mainly relies on pathological examination. CA125, LDH, and UA levels may help in the differential diagnosis. Once diagnosed, further chemotherapy is needed. The primary pelvis mass will disappear as the chemotherapy goes on. For patients with a wish for fertility reservation, spontaneous conception may be attempted in CR status.

Acknowledgment

The patient who is the subject of this case report gave verbal consent to be involved in this case report. All authors of this original manuscript have read and approved the revised version of the paper.

Financial disclosure

None to declare.

Conflict of interest

None to declare.

Informed consent

Not applicable

Data availability

The authors declare that data in this study are available within this article.

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