

Case Report

Primary ovarian Burkitt lymphoma: report of a case and review of literature

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Abstract: We report a case of primary ovarian Burkitt lymphoma that occurred in a 25-year-old woman. The patient complained of a mass in the right ovary discerned by physical examination 2 months prior. Ultrasound examination indicated that the right ovary was enlarged and abundant blood flow signals were observed. Right salpingo-oophorectomy was subsequently performed. Histology was characterized by diffuse sheets of monotonous medium-sized lymphoid cells with plentiful mitotic figures and apoptosis. Numerous tingible-body macrophages were found in the ovarian tissue, presenting a starry sky pattern. The tumor cells expressed CD20, CD10, *BCL6*, and *MYC* in the absence of *BCL2*. Ki-67 proliferative index was very high with a proliferation rate of near 100%. *MYC* (8q24) rearrangement was detected by fluorescence in situ hybridization (FISH) with no *BCL2* (18q21) and *BCL6* (3q37) gene rearrangements. Cumulative evidence established primary ovarian Burkitt lymphoma as the final histopathologic diagnosis with clinical stage I (FIGO). The patient received HyperCVAD chemotherapy after surgery and remained complete response (CR) for 18 months. We aim to provide insight into the future treatment of this rare but lethal disease.

Keywords: Primary, ovary, Burkitt lymphoma

Introduction

Burkitt lymphoma (BL) is an aggressive B-cell lymphoma characterized by frequent presence in extranodal sites or as acute leukemia, mostly with high proliferative activity and *MYC* translocation. All organs of the female genital tract can be involved, and ovarian involvement predominates in a majority of the cases [1, 2]. The molecular hallmark of BL is the deregulation of *MYC* expression due to the translocation of *MYC* to an Ig gene locus. Gene expression profiling has defined patterns of molecular signatures for BL [3, 4]. Mutations of *TCF3(E2A)* and *ID3* are seen in 70% of sporadic BL cases documented [5-8]. Mutations of *MYC*, *CCDN3*, *TP53*, *RHOA*, *SMARCA4*, and *ARID1A* are present in 5-40% of BL cases [9]. About 29 cases of primary ovarian Burkitt lymphoma have been reported (Table 1) [10-38]. The present study

reports the clinical data, histologic morphology, immunohistochemistry, molecular characteristics, and treatment in this case in order to provide a further reference about this tumor's diagnosis and treatment.

Case presentation

The patient was a 25-year-old married female who complained of a mass in the right ovary detected by physical examination 2 months ago. She had regular menstruation. The preoperative transvaginal ultrasound displayed that the right ovary was enlarged approximately 93 × 83 × 74 mm, and abundant blood flow signals were also observed (Figure 1A). The uterus, cervix, and left ovary were normal. The endometrium measured about 7 mm in thickness. Cervical cytology was negative for intraepithelial lesion or malignancy (NILM). The patient

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Table 1. Clinical features of 26 primary ovarian Burkitt lymphoma patients

Case number	1st author	Age	Nationality	Symptoms	Ovarian involvement	Stage	Treatment	Follow-up
1	Baloglu [28]	24	Turkey	Secondary amenorrhea, ascites, pleural effusion	Bilateral	IV Ann Arbor	Chemotherapy: Cyclophosphamide, Adriamycin, Vincristine, L-asparaginase, Prednisolone, plus intrathecal Methotrexate	Remission; autologous bone marrow transplantation; death 35 days after transplantation
2	Vang [27]	62	USA	Constitutional symptoms	Unilateral	I Ann Arbor	Chemotherapy, radiotherapy	Remission
3	Hui [34]	13	USA	Abdominal pain	Unilateral	III Murphy staging	Surgery, chemotherapy (COPADM)	Remission
4	Liang [26]	38	Hong Kong, China	Abdominal pain	Unilateral	I Ann Arbor	Chemotherapy, radiotherapy	Disease-free survival (6 months)
5	Taylor [35]	33	USA	Abdominal pain	Bilateral	NA	Surgery, chemotherapy	NA
6	Miyazaki [25]	16	Japan	Abdominal pain, constipation	Unilateral	NA	Surgery, chemotherapy	Died after 171 days
7	Cyriac [24]	13	India	Abdominal pain, fever	Bilateral	IV Ann Arbor	Chemotherapy (LMB 89 protocol)	Disease-free survival (6 months)
8	Crawshaw [23]	28	UK	Abdominal pain, pregnancy	Bilateral	IV Ann Arbor	Chemotherapy	NA
9	Chishima [22]	25	Japan	Abdominal pain	Bilateral	IV Ann Arbor	Surgery, chemotherapy (CHOP)	Disease-free survival (30 months)
10	Monterroso [21]	21	USA	Abdominal pain	NA	NA	Surgery, chemotherapy	Died after a year
11	Ng [20]	20	Malaysia	Abdominal pain	Bilateral	1B FIGO	Surgery, chemotherapy (BFM regime), plus prophylactic intrathecal Methotrexate	Remission
12	Shacham-Abulafia [18]	39	Israel	Night sweats, abdominal pain, dyspnea	Bilateral	IV Ann Arbor	Chemotherapy (R-Hyper-CVAD plus GMALL-BALL/NHL 2002)	Disease-free survival (6 months)
13	Bianchi [19]	57	Italy	Neurological symptoms	Bilateral	IV Ann Arbor	Surgery, chemotherapy (intensive G-mall protocol)	NA
14	Danby [17]	11	USA	Abdominal pain, nausea, vomiting, anorexia, and weight loss	Unilateral	NA	Surgery, chemotherapy (Rituximab, Methotrexate, Cytarabine, intrathecal Hydrocortisone, Cytosine arabinoside, Methotrexate and Cyclophosphamide)	NA
15	Etonyeaku [16]	18	Nigeria	Abdominal pain, lower abdominal swelli	Bilateral	NA	Surgery, chemotherapy (Cyclophosphamide, Vincristine, Methotrexate)	Died after second chemotherapy
16	Gottwald [15]	27	Poland	Abdominal pain, ascites	Bilateral	NA	Surgery, chemotherapy (COP followed by CODXM + IVAC)	Disease-free survival (3 years)
17	Gutierrez [14]	34	Spain	NA	Bilateral	IV Ann Arbor, 3 FIGO	Surgery, chemotherapy	Died after a month
18	Hatami [13]	58	USA	Abdominal masses	Bilateral	IV Ann Arbor	Surgery, chemotherapy (Vincristine, Rituximab, Methotrexate with Leucovorin and intrathecal Methotrexate)	Disease-free survival (42 months)
19	Munoz [12]	30	Spain	Abdominal pain	Bilateral	NA	Surgery, chemotherapy (CODX-M-IVAC plus Rituximab)	NA
20	Khan [11]	6	India	Abdominal pain and masses	Bilateral	IV Ann Arbor	Surgery, chemotherapy (MCP-842 protocol)	NA

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21	Mondal [10]	6	India	Abdominal pain, difficulties with walking	Bilateral	II R Murphy staging	Surgery, chemotherapy (Magrath protocol using CODOX-M regimen)	Remission
22	Gomez [29]	13	Spain	Abdominal pain	Unilateral	IV Ann Arbor	Surgery, chemotherapy (Doxorubicin, Vincristine, Cytarabine, Dexamethasone, Rituximab)	Remission
23	Xiao [30]	19	China	Chest tightness, asthma	Unilateral	IV Ann Arbor	Surgery, chemotherapy (HyperCVAD, MTX plus Ara-c)	Deceased
24	Xiao [30]	43	China	Fatigue, bone pain	Unilateral	I Ann Arbor	Surgery, chemotherapy (R-CHOP, HD-MTX plus VP)	Remission
25	Gravos [31]	21	Greek	Abdominal distension	Unilateral	I Ann Arbor	Chemotherapy	Remission
26	Al-Maghrabi [32]	42	Saudi Arabia	Abdominal pain	Bilateral	I Ann Arbor	Surgery, chemotherapy (R-CODOX, R-IVAC)	Remission
27	Lanjewar [36]	28	India	Weight loss, distension and pain	Unilateral	NA	Surgery, chemotherapy, HAART	Died after 7 months
28	Sergi [37]	24	Italy	Abdominal tenderness, diffuse pain	Unilateral	NA	Chemotherapy	NA
29	Steininger [38]	17	German	Abdominal pain	Unilateral	III Ann Arbor	Surgery, chemotherapy	Remission

Abbreviation: FIGO: International Federation of Gynecology and Obstetrics, NA: Not available.

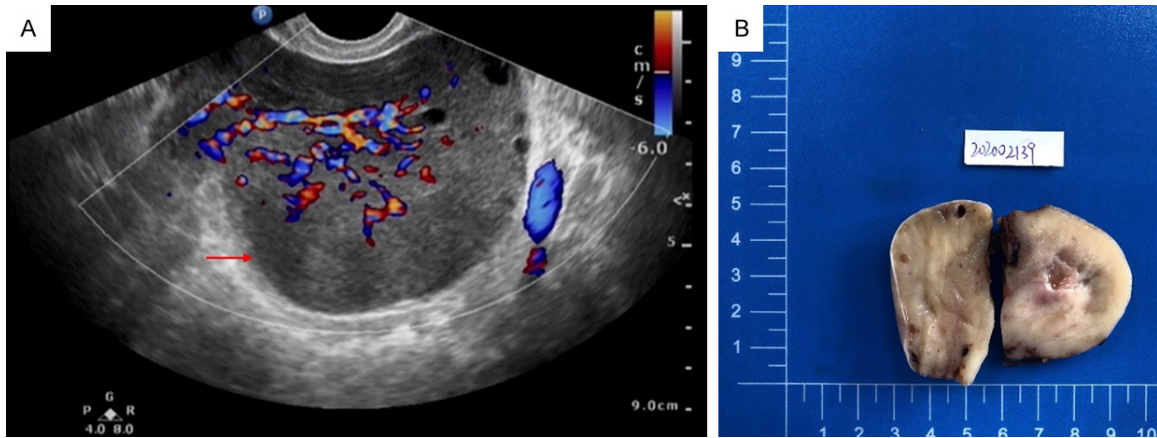


Figure 1. Ultrasound image and gross features of the case. **A.** Preoperative transvaginal ultrasound showed that the right ovary was enlarged, with abundant blood flow signals in the ovarian tissue (marked by red arrow). **B.** The tumor originated from the ovary and was well-circumscribed. A gray-white tender cut surface and scattered hemorrhage was visible.

denied additional aberrant symptoms or signs upon admission. A right salpingo-oophorectomy was subsequently performed.

Gross examination

The removed right ovary measured $11 \times 11 \times 5$ cm in size. The tumor originated from the ovary and it did not spread to the fallopian tube. It was well-circumscribed with a gray-white tender cut surface and scattered hemorrhage was visible (**Figure 1B**). The ipsilateral fallopian tube was grossly unremarkable.

Microscopic examination

Microscopically, the tumor obliterated normal ovarian parenchyma. The involved ovarian tissue was diffusely infiltrated by sheets of monotonous medium-sized neoplastic lymphoid cells and the boundary between the tumor tissue and ovary was unclear (**Figure 2A**). Innumerable neoplastic cells interspersed with scattered reactive tangible-body macrophages engulfing nuclear debris, creating a prominent “starry sky” pattern (**Figure 2B**). When examined at high magnification, the neoplastic cells showed molding and squaring-off of the nuclear membrane and cell membrane, with relatively uniform round or oval nuclei, coarse chromatin, multiple small nucleoli, and an appreciable rim of basophilic cytoplasm. Mitotic figures were numerous as well as a high rate of apoptosis (**Figure 2C and 2D**).

Immunohistochemical findings

Immunohistochemically, tumor cells expressed pan-B-cell antigens (CD20, CD79-a, and PAX5), germinal center markers (CD1 and *BCL6*), and *MYC*. *BCL2*, CD3, CD5, CD138, and TdT were negative. EBV was negative. The proliferation fraction (Ki67 index) was very high, approaching 100% (**Figure 3A-C**).

FISH examination

The $t(8;14)(q24;q32)/IGH-MYC$ was detected by fluorescence in situ hybridization (FISH) (break-apart probes) (**Figure 3D**) whereas *BCL2* (18q21) and *BCL6* (3q37) rearrangements were absent.

The patient underwent bone marrow aspirate evaluation for hematological immunotyping and chromosomes analysis. The results were negative for malignancy and atypical chromosomes. The subsequent PET-CT showed no lesions on other organs. Combined with the clinical visualization, histologic features, immunohistochemical results, cytogenetics, and molecular test, the diagnosis was ascertained as primary ovarian Burkitt lymphoma. The patient received the treatment protocol of HyperCVAD chemotherapy after surgery.

Discussion

Although primary extranodal invasive lymphoma of the ovary is a rare disease, ovarian lym-

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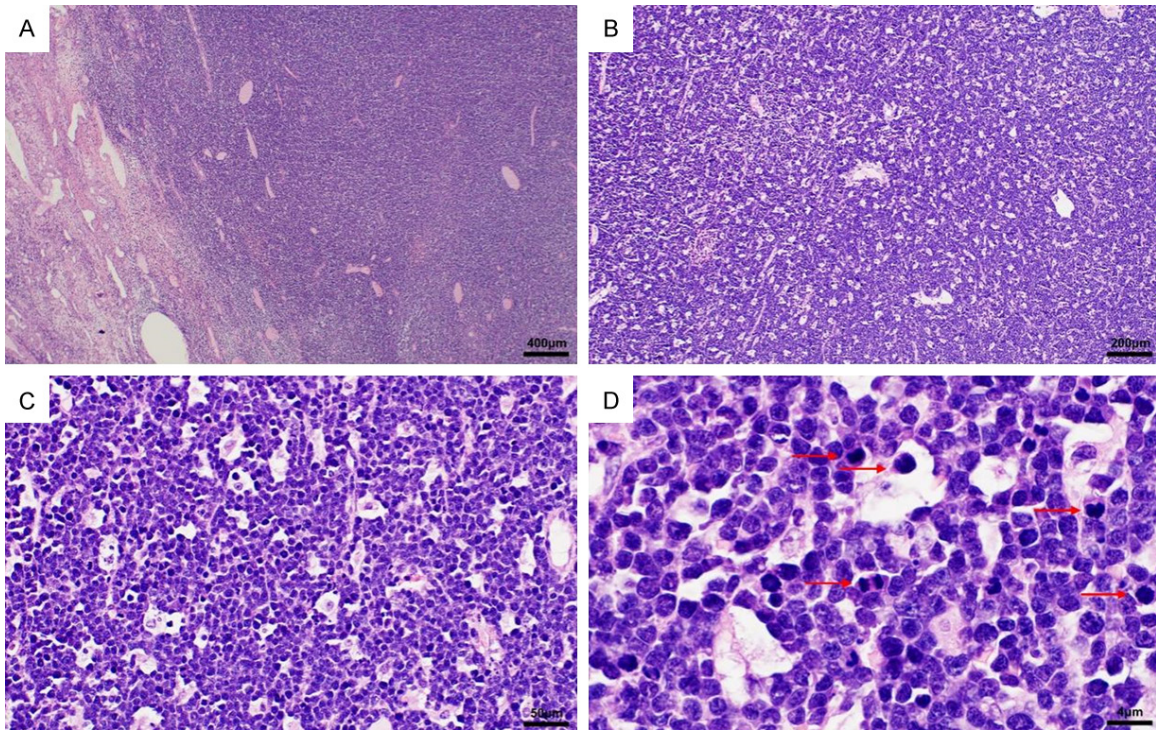


Figure 2. Histology of the ovarian tumor. A. The involved ovarian tissue was diffusely infiltrated by sheets of monotonous medium-sized neoplastic lymphoid cells and the boundary between the tumor tissue and ovary was unclear (magnification: $\times 40$). B. Innumerable neoplastic cells interspersed with scattered reactive tangible-body macrophages engulfing nuclear debris, creating a prominent “starry sky” pattern (magnification: $\times 100$). C and D. The neoplastic cells showed molding and squaring-off of the nuclear membrane and cell membrane, with relatively uniform round or oval nuclei, coarse chromatin, multiple small nucleoli, and an appreciable rim of basophilic cytoplasm. Mitotic figures were numerous as well as a high rate of apoptosis (marked by red arrows). C. Magnification: $\times 400$; D. Magnification: $\times 1000$.

phoma is typically secondary to diffuse systemic disorder. In extranodal lymphoma, primary ovarian lymphoma accounts for 0.5% of extranodal non-Hodgkin's lymphoma and 1% of all ovarian tumors [39]. Several perspectives regarding the origin of primary ovarian lymphoma have been documented: 1. Primary ovarian lymphoma originates from the lymphoid tissues that already exist in the ovary [23]; 2. The blood vessels around the hilum of the ovary or the corpus luteum cells may be tumor-derived cells [10]; 3. Reactive lymphocytes may be secondary to ovarian inflammation (pelvic inflammatory disease and endometriosis) or autoimmune diseases, and following processes of malignant transformation into ovarian primary lymphoma [40]. No specific association was described with EBV. Sporadic BL may coexist with EBV infection [41]. However, tests for EBV infections are also recommended. As reported in this case, the patient has no EBV infection.

Weekes first discovered primary bilateral ovarian Burkitt lymphoma in a 15-year-old girl from Guatemala in 1986 [42]. To our knowledge, about 29 cases of primary ovarian Burkitt lymphoma have been reported in the literature. Among the described cases, 15 were bilateral and 10 were unilateral (1 unavailable). The age of the patients ranged from 6 to 62 years (average, 27 years old). The clinical manifestations of ovarian primary Burkitt lymphoma are unique [43]. Most patients complained of abnormal pain accompanied by additional symptoms including fever, abnormal mass, lower abdominal swelling, ascites, and vaginal bleeding [44, 45]. As for our patient, the young married woman was diagnosed with a mass in the right ovary by physical examination due to infertility. Staging plays an important role in accessing disease progression and determine the individualized treatment of patients. Ann Arbor classification is mostly used in adult BL pa-

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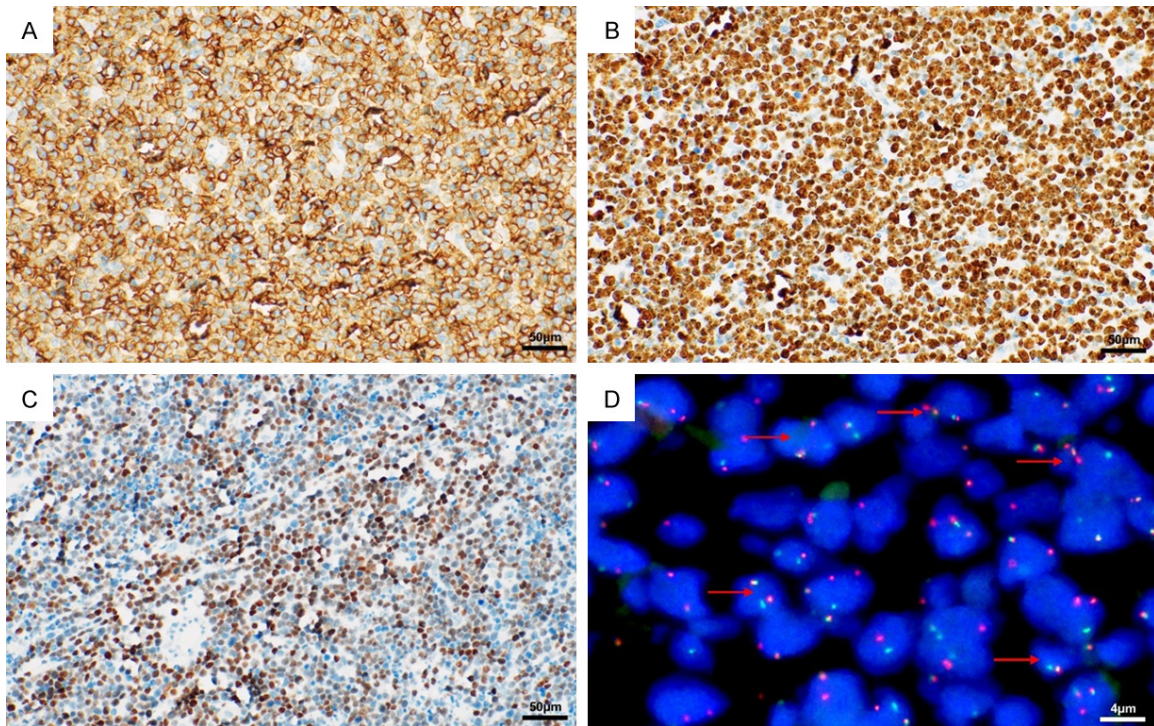


Figure 3. Immunoprofile and FISH examination of tumor cells. A. Positivity for CD20 (magnification: $\times 400$). B. High grade tumor revealed by over 100% cell positivity for Ki67 antibody (magnification: $\times 400$). C. Positivity for BCL-6 (magnification: $\times 400$). D. *MYC* (8q24) rearrangement positive cells include one fusion signal (yellow), one red signal, and one green signal in nuclei. The ratio of *MYC* cleavage signal positive abnormal cells per 100 cells is equal to 61%, which meets the positive diagnostic criteria (magnification: $\times 1000$).

tients, while St. Jude/Murphy applies to pediatric patients. Seven of them are stage I (1 stage 1B, FIGO), 2 are stage II, 11 are stage IV, 6 unavailable. This case presented a primary ovarian Burkitt lymphoma of FIGO staging 1A. The histologic findings of the present case were identical to Burkitt's lymphoma that occurred in other organs, which are characterized by diffuse growth of medium-sized lymphoid cells with round nuclei and agglomerated chromatin [46]. Necrotic debris could be seen in the background, which was swallowed by many tissue cells, forming a typical pattern of the "starry sky".

Primary ovarian Burkitt lymphoma originates from B cells and expresses B cell markers such as CD20, CD79a, PAX5. Cytokeratin and T cell markers are negative. *MYC* represents a family of regulator genes and proto-oncogenes that codes for transcription factors. The *MYC* family consists of three related human genes *c-MYC* (*MYC*), *I-MYC* (*MYCL*), and *n-MYC* (*MYCN*). *c-MYC* (also sometimes referred to as *MYC*) is the first gene discovered in this family

due to its homology with a viral gene *v-MYC*. In cancer, *c-MYC* is often expressed constitutively (persistently), which leads to an increase in the expression of multiple genes. Additionally, some of them are even involved in cell proliferation and contribute to the formation of cancer cells. Common human translocations involving *c-MYC* are critical to the development of most Burkitt lymphomas [13, 47], and the mutation is usually *t* (8;14) (q24; q32) translocation. In this case, CD20 was strongly positive, the result of fluorescence in situ hybridization (FISH) was *MYC* (8q24) rearrangement, the morphology combined with immunohistochemical results and molecular pathologic features were consistent with the diagnosis of Burkitt lymphoma.

The differential diagnosis of primary ovarian Burkitt lymphoma includes secondary ovarian lymphoma, High-grade B-cell lymphoma with double-hit or triple-hit, adult granular cell tumor of the ovary, and small cell neuroendocrine carcinoma (SCNEC). Fox et al. proposed the diagnostic criteria for primary ovarian lym-

phoma in 1988 including: 1. Lymphoma should be confined to the ovary or adjacent lymph nodes or structures at the time of diagnosis; 2. There is no evidence showing the presence of blood or bone marrow disease; 3. Distal involvement should occur at least a few months after ovarian involvement [48]. High-grade B-cell lymphoma with double-hit or triple-hit is a general term for morphologically aggressive B-cell lymphoma with *MYC* rearrangement and additional rearrangement of *BCL2* (double-hit lymphoma, DHL) and/or *BCL6* (triple-hit lymphoma, THL). Adult granular cell tumor of the ovary is frequently encountered in postmenopausal women, with the peak age of onset at 50-55 years old, which represents the most typical clinical ovarian tumor related to estrogen secretion. The nuclei are round, oval, or polygonal, lightly stained, less cytoplasm, and a nuclear groove is visible. Immunohistochemical expression of sex cord-stromal markers is seen such as α -inhibin, calretinin, FOXL2, and CD99. The epithelial markers are negative. Differential diagnosis of the disease also requires an ruling out SCNEC. SCNEC is a high-grade carcinoma composed of small to medium-sized cells with scant cytoplasm and neuroendocrine differentiation. The cytoplasm of the tumor is sparse with a small cell nucleus. Single small nucleolus can be visible and mitotic images are common. Tumor cells can be in a nested pattern, string-shaped and irregular cell clusters, and express neuroendocrine markers such as CgA, Syn, and CD56.

As Burkitt lymphoma is an aggressive B-cell non-Hodgkin's lymphoma with a short and active proliferation cycle, multi-drug combination chemotherapy is considered an optimal treatment protocol for patients with Burkitt lymphoma [49, 50]. The 29 cases previously reported were mostly (20/29) treated with surgery followed by chemotherapy. Surgical treatment plays an important role in providing clinical information, staging, and diagnosis, and patients with confirmed diagnosis should start chemotherapy as early as possible. Although Burkitt lymphoma is highly malignant, combined treatment with multiple chemotherapy regimens can substantially improve the survival rate of Burkitt lymphoma patients [13]. The follow-ups ranged from 0.5 to 3.5 years (average 1.6 years). Four deaths were reported, giving an overall survival rate of 15/19 (78.95%)

(7 unavailable). In our case, the patient underwent right salpingo-oophorectomy and accepted HyperCVAD chemotherapy after surgery. The patient developed complete response (CR) 18 months following the completion of the therapy.

Conclusion

We report a case of a primary ovarian Burkitt lymphoma that occurred in a young woman, confirmed by histological morphology, immunohistochemistry, and FISH. Further studies are of great significance to characterize clinical features of this rare disease, for differential diagnosis and effective therapy.

The reporting of this study conforms to CARE guidelines [51].

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Informed consent has been obtained from patients included in this study.

Disclosure of conflict of interest

None.

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