

## Ovarian Burkitt lymphoma: A rare yet clinically significant entity

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### Abstract

Primary ovarian lymphomas are rare tumors presenting as an ovarian mass that can mimic an epithelial ovarian tumor. The average age of onset is 40 years. Burkitt lymphomas, sporadic in the Western world, typically present with abdominal involvement. Biological tests and imaging guide the diagnosis. Prognosis depends on associated bone marrow and central nervous system involvement. Treatment relies on polychemotherapy. Here, we report a case of Burkitt's lymphoma in a young woman simulating advanced bilateral ovarian cancer.

**Keywords:** Cancer; Ovary; Lymphoma; Prognosis; Chemotherapy

### 1. Introduction

Among ovarian lymphomas (1.5% of ovarian cancers), Burkitt lymphomas are even rarer, especially in young women. Their frequency increases in patients with human immunodeficiency virus. Abdominal locations of this type of lymphoma are classical. In sporadic forms, it is mainly an ileocecal tumor. Peripheral lymph node involvement occurs in 10-20% of cases. In endemic forms, digestive involvement is present in only 60% of cases. It is more diffuse, with peritoneal and retroperitoneal involvement (renal, mesenteric, and ovarian). The diagnosis of ovarian locations is often made retrospectively during the pathological examination of the surgical specimen, when surgery is not the standard treatment. The diagnosis of ovarian Burkitt lymphoma necessitates an evaluation for neuromeningeal (40% of cases) and medullary (15 to 20% of cases) involvement, which significantly affects prognosis. Awareness of their clinical presentation is crucial to avoid inappropriate treatment. The prognosis of these lymphomas has improved with advancements in chemotherapy, which remains the standard treatment.

### 2. Case report

This case involves a 48-year-old patient with no significant medical history, who underwent exploratory surgery three months ago due to suspected mucinous ovarian tumor based on findings from a pelvic CT scan. The surgery was performed due to symptoms such as fever, and severe weakness. During laparotomy, peritonitis of appendicular origin with right pyosalpinx was identified.

The surgical intervention encompassed a right salpingectomy, appendectomy, and peritoneal biopsy with subsequent cytological examination, confirming the diagnosis of appendicular peritonitis. Despite this, the patient's clinical condition did not show improvement, leading to referral to our facility. Upon examination, the patient exhibited signs of asthenia, normotension, normal heart rate, absence of fever, along with a weight loss of 10kg and diffuse pelvic tenderness. Further diagnostic investigation included pelvic ultrasound, revealing three heterogeneous hypoechoic pelvic formations, challenging to characterize accurately. An abdominopelvic CT scan revealed inflammatory thickening of the parietal wall of several small bowel loops and the sigmoid colon. The patient underwent a second surgical

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procedure, revealing hemoperitoneum with adhesions obstructing the visualization of the ovaries and pinkish granulations scattered throughout the digestive tract, which were sampled for histopathological examination confirming Burkitt lymphoma. The patient was admitted to the intensive care unit for septic shock and unfortunately succumbed to the condition.

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### 3. Discussion

Ovarian lymphomas can arise from lymphocytic aggregates within the ovaries, either as a physiological process or in response to inflammatory or autoimmune phenomena [1]. Primary ovarian lymphomas represent 0.5% of non-Hodgkin lymphomas and 1.5% of ovarian cancers [2].

The age of onset varies, with a mean age around 40 years [1-11], occurring earlier than epithelial cancers. Bilateral involvement occurs in 41 to 71% of cases [4]. Similar to our patient, presenting symptoms are nonspecific, including pelvic pain, abdominal distension, ascites, digestive obstruction, diffuse peritoneal nodules, and large tumors.

The diagnosis of primary ovarian lymphoma requires an involvement limited to the ovaries, there for they have a favorable prognosis compared to disseminated lymphomas that affect the ovaries, with a remission period of over 60 months after surgical treatment [6].

Burkitt lymphomas represent 3 to 5% of lymphomas in Western countries and are associated with a genetic defect of the c-myc oncogene. In Western populations, Epstein-Barr virus (EBV) genome is only found in episomal form in 10 to 15% of cases. The incidence of Burkitt lymphoma is higher (35 to 40%) in HIV-positive individuals [12]. Our patient tested seronegative for HIV.

Primary ovarian Burkitt's lymphomas predominantly affect young women and commonly manifest bilaterally with associated ascites [5]. Elevated CA 125 and LDH levels are typical in these cases. Differential diagnoses include granulosa tumors, dysacromelic tumors, and metastatic lesions. Pathologically, Burkitt's lymphomas are distinguishable from other non-Hodgkin lymphomas and epithelial ovarian cancers. Imaging techniques aid in the differential diagnosis between these tumor types [13, 14]. Ultrasound typically reveals nonspecific, homogeneous, hypoechoic ovarian lesions with medium-intensity vascularization on Doppler analysis. CT scans show well-defined, hypodense lesions with moderate enhancement post-contrast injection, while MRI depicts homogeneous solid masses with hypointensity in T1 and slight hyperintensity in T2, showing moderate enhancement following gadolinium administration. This is in contrast to epithelial ovarian tumors, which present as heterogeneous masses with cystic and solid components, necrotic areas, and more pronounced enhancement post-contrast injection.

The treatment of Burkitt's lymphoma primarily consists of intensive, short-term multidrug therapy incorporating high-dose methotrexate, cyclophosphamide, and cytarabine. In the literature, surgical intervention has been employed in patients with ovarian lymphoma due to the clinical similarities with epithelial ovarian cancer, although its role in the management of this systemic disease is not well-defined. In our case, preoperative diagnosis was challenging. Burkitt's lymphoma is classified as a high-grade malignancy, and advancements in chemotherapy have significantly improved prognosis, with cure rates ranging from 50 to 80% [1].

The prognosis of ovarian lymphomas is influenced by several factors including clinical stage, disease presentation, histological type, and phenotype. Poor prognostic indicators include medullary and meningeal involvement, positive HIV serology, presence of translocation (14 18), and incomplete remission after chemotherapy. Despite these factors, patients treated solely with chemotherapy for ovarian non-Hodgkin's lymphoma may still retain fertility, particularly if they are under thirty years old at the time of diagnosis [12]. Dao [6] reported a case of triple pregnancy following diffuse intermediate-grade large B-cell lymphoma affecting only one ovary, treated with surgery and chemotherapy. However, the scarcity of such cases may be attributed to older age and bilateral castration in most affected women.

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### 4. Conclusion

Endemic Burkitt's lymphoma with ovarian involvement is frequently diagnosed belatedly, contributing to its typically grim prognosis. Enhancing its management and prognosis necessitates an emphasis on early and accurate diagnosis, highlighting the importance of vigilant clinical surveillance and timely intervention.

## Compliance with ethical standards

### *Disclosure of conflict of interest*

No conflict of interest to be disclosed.

### *Statement of ethical approval*

The present research work does not contain any studies performed on animals/humans subjects by any of the authors.

### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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