

CASE REPORT

Gynecologic Oncology

## Ovarian non-Hodgkin lymphoma with central nervous system involvement: A case report

Brenda K. Nyamu<sup>1\*</sup>, Eunice J. Cheserem<sup>1</sup>

<sup>1</sup>Department of Obstetrics and Gynecology, University of Nairobi, Nairobi, Kenya

\*Correspondence: [bkiende@gmail.com](mailto:bkiende@gmail.com)

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### Abstract

**Background:** Ovarian lymphoma is rare. Extranodal non-Hodgkin lymphoma (NHL) represents 0.5% of malignant genital diseases. Due to its rarity, there are no accepted treatment protocols. Diffuse large B cell lymphoma (DLBCL) has a central nervous system (CNS) involvement rate of 3-5%.

**Case presentation:** A 38-year-old presented to the gynecological oncology clinic with abdominal pain and distension. Her tumor marker CA-125 levels were elevated. She developed right-sided facial nerve palsy and blindness. A diagnosis of aggressive NHL with a differential of DLBCL was made by histopathological and immunohistochemical examination of the

excised ovarian tissue. A head computed tomography scan revealed subtle enhancing opacity suspicious of lymphomatous deposits. She was lost to follow-up after ten months of chemotherapy and cranial radiotherapy.

**Conclusion:** Primary lymphoma of the ovary is rare, and the prognosis is often poorer when compounded with CNS involvement. Because of poor prognosis in the presence of CNS involvement, this case highlights the need for timely diagnosis and management.

**Keywords:** CNS involvement, diffuse large B-cell lymphoma, non-Hodgkin lymphoma, ovarian cancer

### Introduction

Lymphomas are a heterogeneous group of malignant lymph tissue proliferative diseases, which constitute 3-4% of neoplastic processes, 85% being of non-Hodgkin type (1). Twenty-five to forty percent of non-Hodgkin lymphomas (NHL) are extranodal, and 2% involve the female genital system (1). Ovarian lymphomas are rare because no lymphoid tissue is found in the ovaries. They constitute 0.5% of all malignant genital diseases. The diagnosis is more commonly an incidental finding of either a primary or secondary lesion (2). However, they may present as an initial clinical presentation of occult extraovarian diseases or as the manifestation of widely disseminated diseases. The five-year survival rates of primary extranodal and secondary lymphomas are 80% and 33%, respectively (2). Ovarian lymphomas are more

common in the fourth decade of life (3). Due to their rarity, there are no widely accepted treatment protocols. The prognosis is poorer with central nervous system (CNS) involvement, which occurs in 3-5% of cases with a one-year survival rate of 25.3% (4). There is an increased risk of CNS involvement in patients with elevated serum lactate dehydrogenase, serum albumin <35g/l, <60 years of age, retroperitoneal lymph node involvement, and involvement of more than one extranodal site (4). There is a need to diagnose and manage high-risk patients early and accurately. This is a case of ovarian lymphoma discovered incidentally as an ovarian mass complicated by CNS involvement with a poor outcome.

### Case presentation

A 38-year-old presented to the gynecological oncology clinic at Kenyatta National Hospital with a one-month history of per vaginal bleeding and six-month history of intermittent dull pain in the abdominal left upper quadrant associated with gross abdominal distension, and unintentional weight loss. She was a known HIV-seropositive patient and had been on antiretroviral medication for three years before presentation. Physical examination revealed multiple, mobile, firm, irregular abdominopelvic masses, and bilateral breast masses. She did not have palpable lymphadenopathy. A clinical diagnosis of a metastatic ovarian tumor was made. During her treatment course, she developed right-sided facial nerve palsy and blindness. Preoperative abdominal computed tomography (CT) scan revealed a large multilobular soft tissue mass arising from the pelvis that later differentiated to matted retroperitoneal nodes and bilateral inguinal adenopathy with the largest on the right measuring 1.3cm and the left measuring 1.1cm. Her tumor marker CA-125 and lactate dehydrogenase levels were elevated at 356.7U/mL (reference range <35 U/mL) and 1226U/L (reference range 109-245U/L), respectively. A head CT scan revealed subtle enhancing opacity in the right parasellar region and posterior half of the right optic nerve, suspicious of lymphomatous deposits, associated with the enlargement of the superior ophthalmic vein. Genetic screening of the patient revealed a 50% degree of BRCA2 genetic mutation.

Given the suspicious ovarian malignancy, she was scheduled for exploratory laparotomy with bilateral salpingo-oophorectomy and omentectomy (Figure 1-2). An intraoperative assessment revealed retroperitoneal lymphadenopathy. Histopathological examination and immunohistochemistry using CD20, CD79b, CD3, BCL6, BCL2, and Ki67 were performed on the excised tissue. A diagnosis of aggressive non-Hodgkin lymphoma, with a differential of diffuse large B-cell lymphoma, was made. The tumor was staged Ann Arbor IV and the patient received three cycles of the cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) regimen followed by two cycles of etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin (EPOCH). She was also scheduled for ten sessions of cranial radiotherapy. She was lost to follow-up after ten months of treatment and is presumed dead as attempts to trace her were futile, taking into consideration the poor prognosis and lack of improvement in her clinical condition.

## Discussion

Ovarian lymphoma commonly presents with an abdominal or pelvic mass in 3-5% of patients (3),



**Figure 1:** Intraabdominal view of bilateral ovarian masses and atypical omentum.



**Figure 2:** Gross specimen of bilateral ovarian masses with a smooth surface after bilateral salpingo-oophorectomy measuring 25 x 16cm (left ovarian mass), and 26 x 11cm (right ovarian mass).

abdominal pain, weight loss, weakness, dyspnea, vaginal bleeding, ascites, and sometimes constitutional symptoms (5). Atypical presentations include postmenopausal bleeding or changes in bowel habits (6). Diffuse large B-cell lymphoma is the most common subtype of ovarian NHL and the most common subtype of ovarian lymphoma (7) with a CNS involvement rate of 3-5% (4). Here, the patient presented with a six-month history of abdominal pain and distension and later developed CNS symptoms. She presented with three of the five markers of increased risk for CNS involvement, including elevated serum lactate dehydrogenase, retroperitoneal lymph node involvement, and <60 years of age. She also had elevated levels of tumor marker CA-125, although it

has limited specificity in primary gynecologic NHL (8).

Mutations in the BRCA1 and BRCA2 tumor suppressor genes are associated with an increased risk of breast and ovarian cancers (9), which further increased her risk. An immunohistochemical panel consisting of CD45, CD20, CD3, CD10, CD138, ALK-I, BCL-2, and BCL-6 supplemented with prognostic markers is a prime requisite for a patient-centered subtype-specific final diagnosis in lymphomas. Lymphomas are treated with chemotherapy, using a regimen based on the cytological NHL type. Rapidly progressive NHL tumors are highly responsive to chemotherapy, and the first-line therapy includes the CHOP regimen (8). The central nervous system prophylaxis with intravenous systemic methotrexate is recommended as it is associated with decreased CNS recurrence risk and a three-year survival rate of 80% (10). There is lack of beneficial outcomes in cytoreductive surgery and is therefore not recommended (7).

### Conclusion

Primary lymphoma of the ovary is rare, and the prognosis is often poorer when compounded with CNS involvement. In comparison to nodal lymphomas, the poor prognosis could be due to delayed or inaccurate diagnosis. Ovarian lymphoma, therefore, needs to be considered in the differential diagnosis of solid ovarian masses, and early diagnosis is essential. In the event of high-risk patients, early CNS prophylaxis with methotrexate should also be initiated.

### Consent for publication

Informed consent for publication was obtained from the patient.

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### Declarations

### Conflict of interests

The authors declare no conflicts of interest.

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