

East African Medical Journal Vol. 92 No. 3 March 2015

TUBO-OVARIAN PRESENTATION OF BURKITT'S LYMPHOMA: CASE REPORT

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SUMMARY

Burkitt's lymphoma rarely presents as a primary of the ovary. High index of suspicion is required to avoid delay of definitive management. There are a few case reports presented on ovarian Burkitt's lymphoma. We present a case of a 23 year old, para 1+1 HIV negative patient who presented to the Kenyatta National Hospital with a one month history of progressive abdominal swelling, fatigue, lower limb swelling, nausea and vomiting. Abdominal examination, revealed bilateral adnexal masses confirmed by ultrasonography. She underwent emergency laparotomy following a diagnosis of bilateral ovarian masses with torsion. Surgical specimen showed tubo-ovarian tissue with sheets of lymphoid cells of small to intermediate size, with numerous tangible body macrophages depicting a starry sky appearance. Immunohistochemistry demonstrated a strongly positive CD20, a positive CD 10, a 90-95% positive Ki67, a positive Bcl6 and a negative pan-CK. A definitive diagnosis of tubo-ovarian Burkitt's was made. The patient unfortunately succumbed before commencement of chemotherapy. Autopsy, concluded the cause of death to be widely disseminated Burkitt's lymphoma, with a most likely tubo-ovarian primary and intestinal obstruction. Burkitt's lymphoma should be considered as a differential diagnosis in ovarian masses for timely diagnosis and management.

INTRODUCTION

Burkitt's lymphoma is a common form of non-Hodgkin's lymphoma (NHL) which originates from the B cells. NHL may involve the reproductive system with the ovary as the most common site of involvement. Burkitt's lymphoma accounts for 0.5% of all NHL and 1% of all ovarian neoplasms, making it a rare primary presentation of the ovary. Secondary ovarian involvement is more likely compared to primary NHL. Secondary ovarian NHL is bilateral in half the cases and usually the ovaries are of normal size or slightly enlarged. There usually associated ascites in primary involvement(1-3). The most common symptoms of primary ovarian Burkitt's lymphoma are pelvic pain, abnormal vaginal bleeding, progressive abdominal mass and sometimes it presents with non-specific symptoms (3). The risk factors include infection with Human Immunodeficiency Virus (HIV), Epstein Barr Virus, Human T-Cell Lymphotropic Virus type 1 and

malaria(4). There are few documented cases of tubo-ovarian presentation of Burkitt's lymphoma. Due to this, clinicians may miss out this rare diagnosis, hence a need for a high index of suspicion.

CASE REPORT

We describe a case of 23 year old, Para 1+1 HIV negative female patient with unknown Epstein Barr virus antibody status. She presented at the Kenyatta National Hospital Accident and Emergency Department with a one month history of progressive abdominal swelling, fatigue, lower limb swelling, nausea and vomiting. The abdominal swelling was associated with abdominal pain, early satiety and a feeling of fullness.

On physical examination, she was sick looking, wasted, very pale and dyspnoeic. There was no associated jaundice or cyanosis. Abdominal examination revealed, bilateral adnexal masses associated with marked tenderness. An abdomino-

pelvic ultrasound, showed large multi-septated cysts in both adnexae and fatty infiltrates of the liver. Both plain chest radiograph and a lower limb doppler ultrasound were normal.

A diagnosis of bilateral ovarian masses with torsion was made and the patient underwent explorative laparotomy. Intra-operatively, straw colored ascitic fluid with bilateral ovarian cysts measuring 8 cm by 9 cm were found. A left salpingo-oophorectomy and drainage of cystic fluid from the right ovary was done. The right ovary was preserved. The specimen was taken for histology and immunohistochemistry.

Histology showed tubo-ovarian tissue with tumour composed of sheets of lymphoid cells of small to intermediate size, with numerous tangible body macrophages depicting a starry sky appearance. Mitotic figures, some of which were atypical were appreciated together with areas of coagulative necrosis. A histological conclusion of features in keeping with tubo-ovarian Burkitt's lymphoma was made and immunohistochemistry recommended to confirm diagnosis. Immunohistochemistry demonstrated a strongly positive CD20, a positive CD 10, a 90-95% positive Ki67, a positive Bcl6 and a negative pan-CK. A definitive diagnosis of tubo-ovarian Burkitt's was made. The patient was worked up for chemotherapy.

The patient unfortunately succumbed before chemotherapy treatment was commenced. The post mortem revealed multiple soft and enlarged mediastinal, para hilar, para aortic and para iliac lymph nodes. The stomach was distended with multiple sub-mucosal tumour nodules. The small intestine had adhesions with proximal dilatations and tumour nodules. The genitourinary system showed a left salpingo-oophorectomy scar with adhesions and a grossly normal uterus. The right tube and ovary were not infiltrated by tumour. Sections of the lymph nodes showed sheets of monotonous lymphoid cells consistent with Burkitt's lymphoma. Autopsy histology of sections from the tumour nodules, para aortic lymph nodes and parailiac lymph nodes showed a sheet of monotonous population of lymphoid cells of intermediate size, uniform and round with scanty basophilic cytoplasm and coarse chromatin often with 1-3 nucleoli, features in keeping with a lymphoproliferative neoplasm consistent with Burkitt's lymphoma.

Image 1, shows the gross appearance of stomach and small intestine autopsy specimen with sub-mucosal tumour nodules. Images 2-5 show histology slides of ovarian, stomach, small gut and parailiac node autopsy tissues. Images 6-9 are immunohistochemistry slides of the surgical specimen.

Image 1: Gross specimen of stomach and small intestine autopsy with sub-mucosal tumor nodules in a case of tubo-ovarian Burkitts Lymphoma



Stomach



Small intestine

Image 2: Autopsy histology of ovarian tissue in a case of tubo-ovarian Burkitts Lymphoma

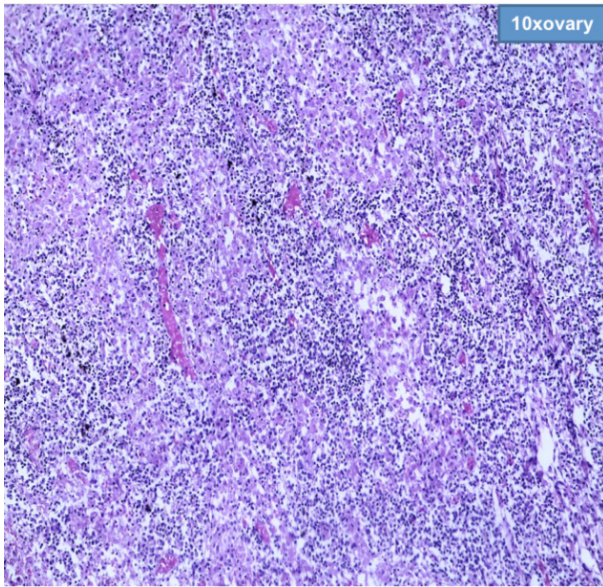


Image 5: Autopsy histology of para iliac lymph node in a case of tubo-ovarian Burkitts Lymphoma

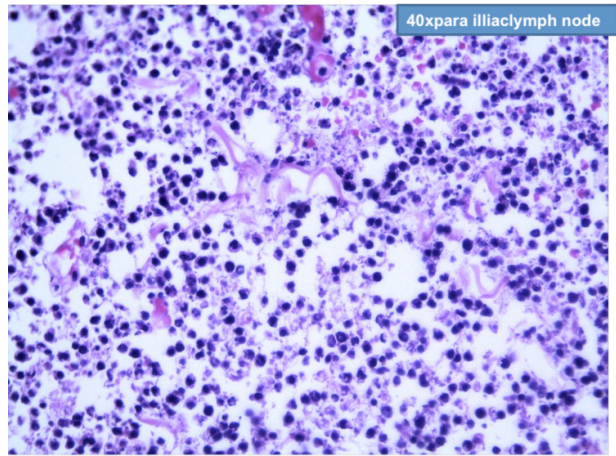


Image 3: Autopsy histology of stomach tissue in a case of tubo-ovarian Burkitts Lymphoma

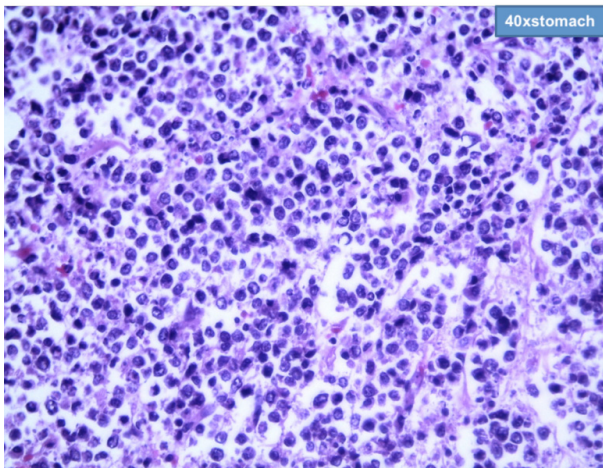


Image 6: Strongly positive CD20 immunohistochemistry on surgical specimen in a case of tubo-ovarian Burkitts lymphoma

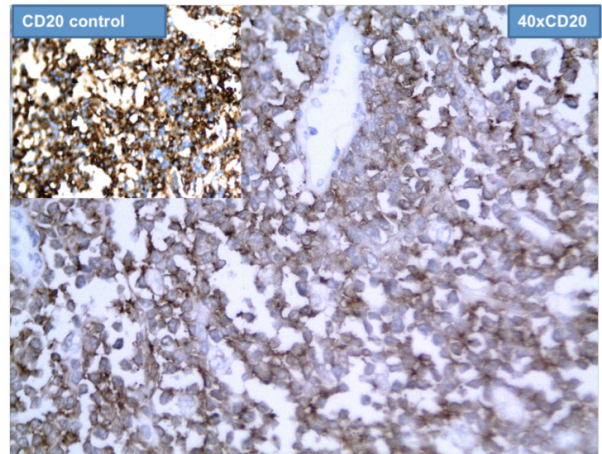


Image 4: Autopsy histology of small gut tissue in a case of tubo-ovarian Burkitts Lymphoma

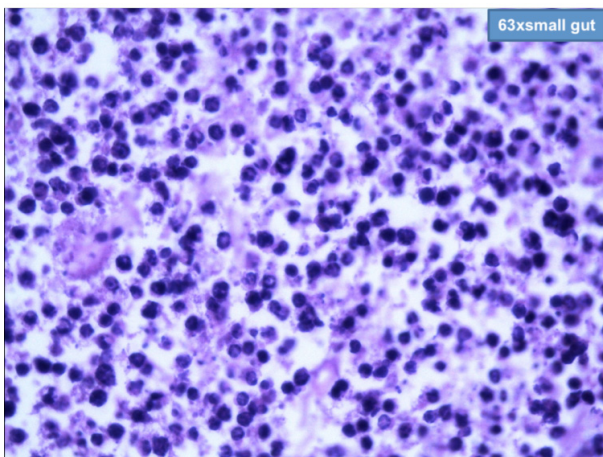


Image 7: Positive CD10 immunohistochemistry on surgical specimen in a case of tubo-ovarian Burkitts lymphoma

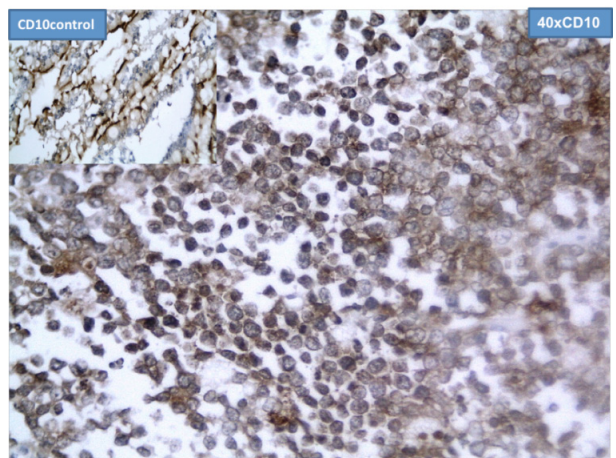


Image 8: Positive Bcl6 immunohistochemistry on surgical specimen in a case of tubo-ovarian Burkitt's lymphoma

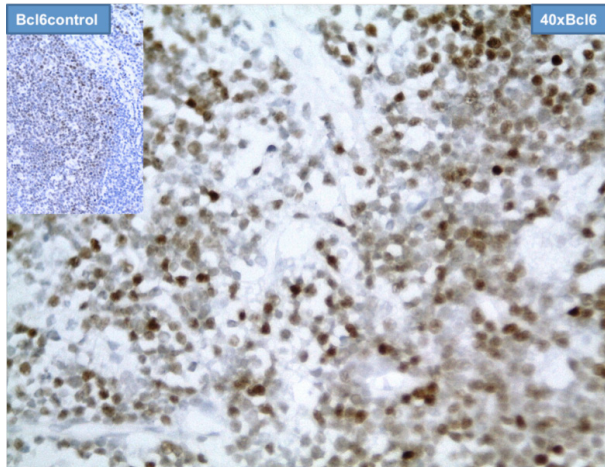
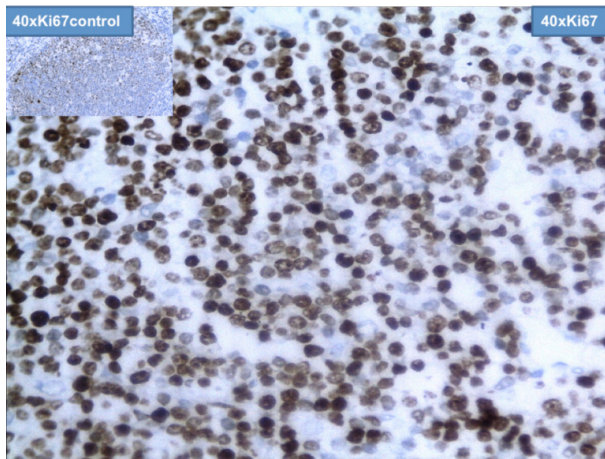


Image 9: Positive Ki67 immunohistochemistry on surgical specimen in a case of tubo-ovarian Burkitt's lymphoma



The cause of death was concluded to be widely disseminated Burkitt's lymphoma with a most likely tubo-ovarian primary and intestinal obstruction due to intestinal adhesions.

DISCUSSION

Tubo-ovarian Burkitt's lymphoma is a rare presentation of adnexal masses. Its diagnosis is often made incidentally after surgical specimen histology followed by immunohistochemistry. Symptoms and signs are non-specific and often mimic those of an acute gynecologic surgical emergency such as ovarian torsion, as was with this case (5). A high index of suspicion is therefore needed for prompt definitive management. This case has also underscored the importance of autopsy especially in cases such as

this. In this case autopsy revealed involvement of the stomach and intestines, which we believe its complications were the cause of death that impeded a chance to offer definitive treatment.

The treatment of choice is combination chemotherapy. The role of surgery is still under debate (5). With timely diagnosis and treatment, the prognosis of ovarian lymphomas has been shown to be good, with B cell lymphomas such as Burkitt's faring better than T cell lymphomas (1, 5).

CONCLUSION

Burkitt's lymphoma should be considered as a differential diagnosis in ovarian masses for timely diagnosis and management.

Informed consent: Written informed consent was obtained from the next of kin for publication of this case report and any accompanying images.

ACKNOWLEDGEMENTS

We acknowledge all the staff in the Departments of Obstetrics and Gynaecology, University of Nairobi, Department of Reproductive Health, Kenyatta National Hospital and Department of Human Pathology, University of Nairobi.

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