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Case Report

Primary Ovarian Burkitt Lymphoma: A case report and lessons learnt

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ABSTRACT

Primary ovarian lymphoma though rare but it is well known. We present a case of primary ovarian Burkitt lymphoma in a 19 year old lady that presented with abdominal swelling and pains. Initial abdomino-pelvic ultrasound showed a huge heterogeneous but largely hypo echoic solid mass arising from the pelvis measuring 17.3 x 12.4cm with areas of calcifications. Exploratory laparatomy revealed bilateral ovarian masses necessitating TAH, BSO. Final diagnosis was made after the histology report and immune-staining. She was commenced on R-CHOP regimen post-operatively unfortunately she succumbed to the disease two weeks after the commencement of chemotherapy.

Keywords: primary ovarian Burkitt, Laparatomy, chemotherapy

Introduction

Ovarian malignancy accounts for 15-20% of female genital malignancies and the 4th most common cause of cancer deaths in women.[1] Primary ovarian lymphoma is very rare accounting for 1.5% of all ovarian cancers and 0.5% of Non Hodgkin's lymphoma.[2] We present a case of primary ovarian Burkitt lymphoma presenting as an advanced ovarian cancer managed by surgery and chemotherapy.

Case Report

A 19 year old nullipara presented with a month history of progressive abdominal swelling and a week history of abdominal pain. The pain was described as constant, dull in nature and worse on movement and at night. There was associated high grade fever, nausea, non productive cough and progressive weight loss but no change in bowel habits and no drenching night sweat. She noticed progressive weakness in both lower limbs and subsequent loss of motor function. She had normal menstrual cycle, yet to attain coitarche, no significant co-morbidity.

She was chronically ill-looking, in painful distress, febrile (T38.40c). The abdomen was distended with supra-pubic preponderance with a tender mass of 26 weeks size, smooth and firm. Ascites was clinically demonstrable. There was loss of normal lumbar lordosis with tenderness over the

lumbar region. Abdomino-pelvic ultrasound show a huge heterogenous but largely hypoechoic solid mass arising from the pelvis measuring 17.3 X 12.4 cm with areas of calcifications. Both kidneys were enlarge with multiple well rounded cystic masses bilaterally, the largest measuring 3.1 X 2.4 cm. Chest x-ray was normal and genexpert was negative. She subsequently had exploratory laparatomy with TAH, BSO and infracolic omentectomy. Intraoperative findings were haemorrahagic ascites of 2.5 litres, bilateral ovarian masses measuring 20 X 16 cm and 22 X 14 cm left and right respectively with a combined weight of 3.3 Kg. The left mass ruptured spontaneously. Presence of omental cakes were noted with the largest measuring 5 X 3 cm. The final histopathological diagnosis was NHL-Burkitt variant. CD 10 and CD 20 were positive on immunochemistry CD 3 was negative as well as beta hCG, alpha-fetoprotein (AFP), vimentin and pan cytokeratin.

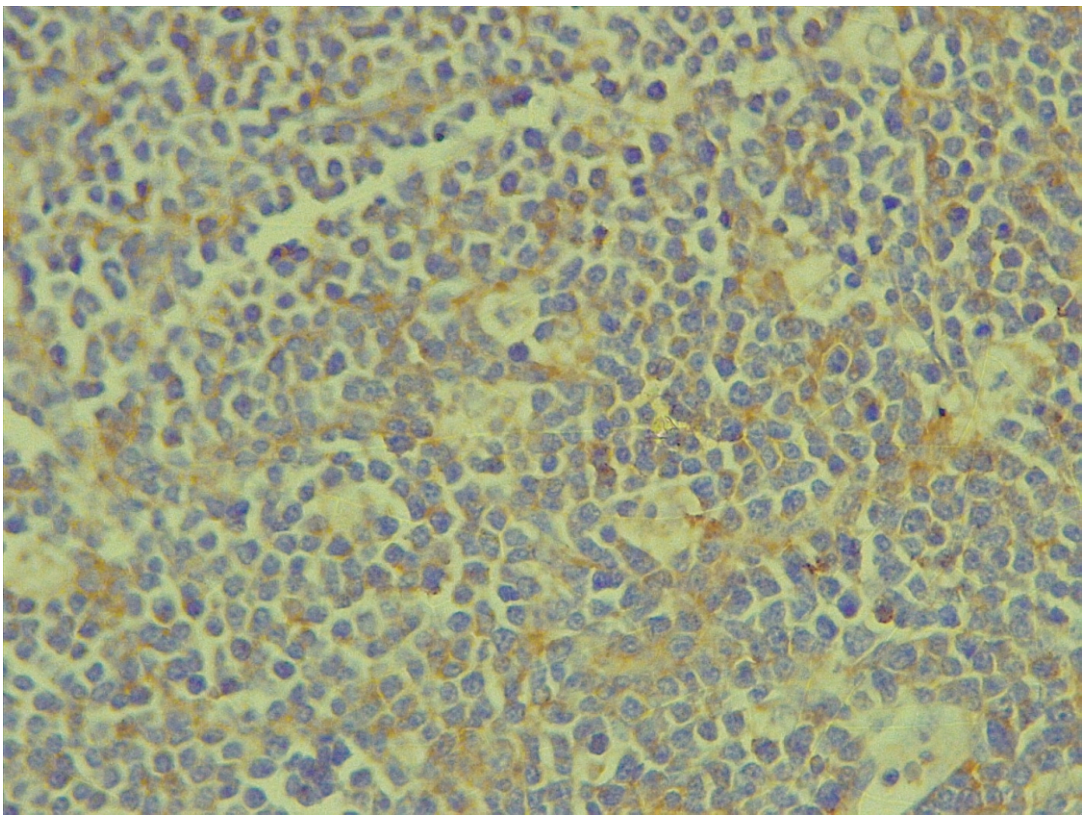

Figure 1 H & E staining classical of Burkitt (diffuse lymphoid infiltrate with starry-sky appearance)

Further evaluation including bone marrow aspiration did not yield any abnormality. She was assigned stage 4EB (Non Hodgkin's lymphoma staging) and was commenced on chemotherapy (R-CHOP) plus intrathecal methrotrexate and cytarabin standard doses, unfortunately she could get only a dose of R-CHOP three weeks after the diagnosis before succumbing to the disease. The challenges faced were largely logistic resulting in delays occasioned by poverty and lack of social support in the management of this patient.

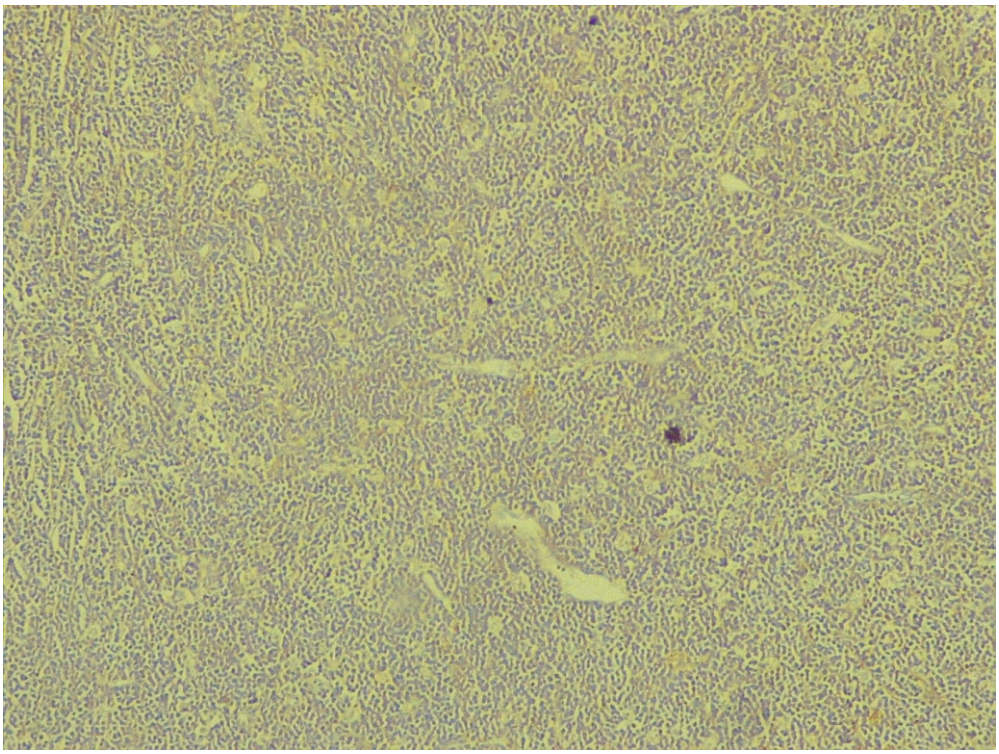

Figure 2 Positive CD20 immunochemistry staining

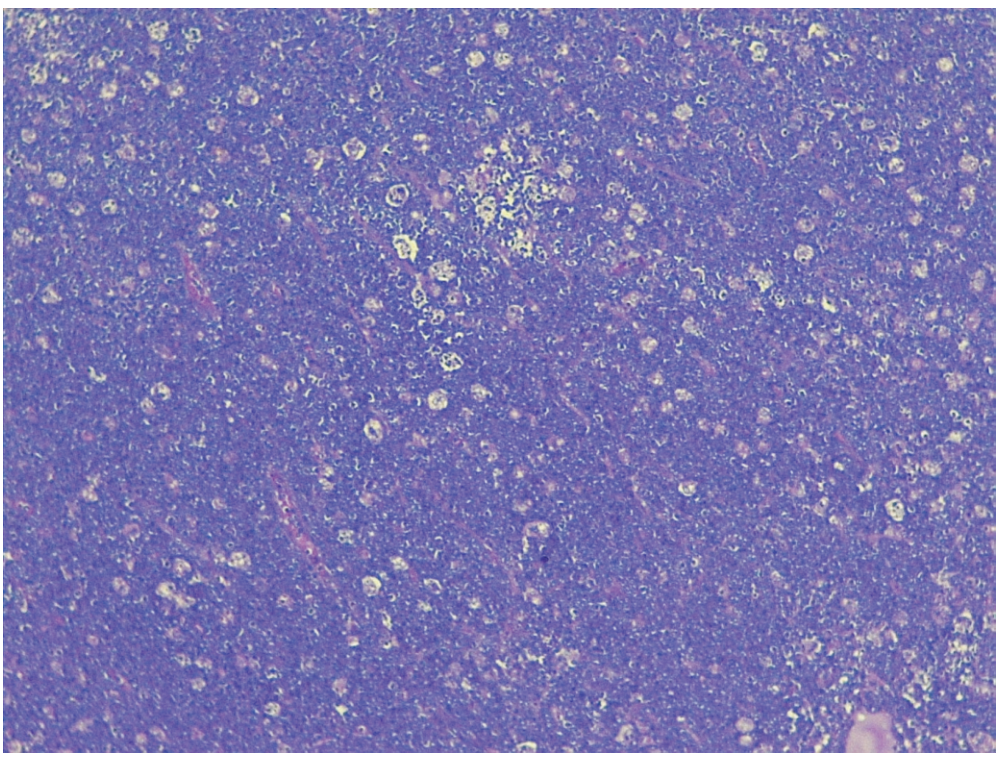

Figure 3 Positive CD10 immunochemistry staining

Discussion

The involvement of the female genital tract in NHL is rare yet well documented. In cases of genital involvement the ovary become one of the common sites.[3] Ovarian lymphoma may be primary or secondary, unlike the secondary lymphoma, primary ovarian lymphoma is rare accounting for 0.5% of Non Hodgkin's lymphoma and 1.5% of ovarian malignancies.[2] The commonest histological subtype in the ovary is the diffuse large B-cell lymphoma followed by Burkitt lymphoma.4 Ovarian lymphoma is thought to arise from lymphoid tissues around the hilum, occasionally from lymphocytes related to the corpus luteum.[5]

In making the diagnosis of primary ovarian lymphoma the following criteria as proposed by Fox et al6 should be considered: (i) The tumour is confined to the ovary or regional lymph nodes or adjacent organs at the time of diagnosis (ii). Absence of abnormal cells in Bone marrow and peripheral blood and (iii) If extra ovarian disease appear later, there must be few months between the time of ovarian and extra ovarian lesions. Our patient met these criteria as both the bone marrow and peripheral blood were free of any abnormal cells as well as absent nodal involvement.

Burkitt lymphoma (BL) is an aggressive disease with potential for very rapid growth. Primary ovarian Burkitt lymphoma may present with rapidly progressive bilateral ovarian swelling usually without ascites. Three subtypes are recognised:[7] endemic BL usually associated with Epstein Barr virus infection, sporadic BL seen mostly among young adults and with extra-nodal manifestation especially in the abdominal cavity and thirdly immunodeficiency associated BL commonly related with HIV/ADIS. Most patient presents with abdominal swelling and pain, other constitutional symptoms like fever, drenching night sweat weight loss and fatigue may be present. Our patient presented with all these features. The haemorrhagic ascites in the patient may be due to the breech in the ovarian capsule noted at surgery.

Plain radiograph no longer play significant role in diagnosis as the features are usually non-specific except for chest involvement. Ultrasound scan may show homogenous hypo echoic solid abdominal mass arising from the pelvis. Computed tomography (CT) is very useful in characterising site of disease and excluding differential diagnosis. With advances in imaging, Positron emission tomography (PET) and PET-CT has become the imaging of choice with very high sensitivity and specificity approaching 100% in identifying both nodal and extra nodal diseases. Immunohistochemistry help to differentiate the types of lymphoma. CD10, CD20 were positive while CD3 HCG and pancytokeratin were negative in our patient. Our patient unfortunately did not have PET-CT scan due to non availability at our facility and lack of finance for referral.

Due to the rarity of POL, diagnosis is usually made after laparatomy for suspected ovarian malignancy. Our patient had laparatomy with TAH, BSO and infra colic omenectomy. Burkitt Lymphoma is highly chemo-sensitive with varying treatment protocols8-12 based on risk assessment. We used R-CHOP (cyclophosphamide, Doxorubicin, Vincristine Rituximab and prednisone) regimen with Intra thecal methrotrexate and cytarabin added. Many authors had reported complete remission of disease with the regimen. Our patient was not that fortunate as she only had a dose of the drugs that was even delayed due to lack of fund and support. She could not afford the intrathecal injections.

The outcome is better in paediatric patients than in adults, similarly primary ovarian Burkitt Lymphoma has a better prognosis than secondary lymphoma.13 Generally it depend on international prognostic index (IPI), these include serum lactate dehydrogenase level (LDH), Ann Arbor staging, number of extra nodal sites, patient age and performance status.[14]

Conclusion/Lessons Learnt

Early diagnosis requires a high index of suspicion. Primary ovarian Burkitt Lymphoma should be suspected in patients presenting with rapidly progressing bilateral ovarian tumour usually without Ascites in a young adult and elevated LDH

level. Chemotherapy should be early and aggressive as the tumour is highly chemo sensitive. Extensive surgery may be counterproductive due to inherent high morbidity and mortality associated with such procedure. Finally, deliberate institutional policy to support cancer patient especially indigent patients should be put in place.

Conflict of interest: None
Support: None

Abbreviations

BL	Burkitt Lymphoma
BSO	Bilateral salpingoopherectomy
LDH	lactate Dehydrogenase
HCG	Human chorionic gonadotrophin
NHL	Non Hodgkin's Lymphoma
POL	primary ovarian lymphoma
TAH	total Abdominal Hysterectomy

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