CASE REPORT

PRIMARY OVARIAN BURKITTS LYMPHOMA

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Primary ovarian Burkitt lymphoma (BL) is a very rare and aggressive malignancy. We report an 18-year-old female patient who presented with a large, tender abdomen, and highly de-ranged renal and liver functions. Ultrasonography showed hepatosplenomegaly, mild ascites, dilated biliary channels and a heterogeneous pelvic mass of size ~15×10×6.4 cm. Immunohistochemical (IHC) staining of the biopsy sample excised from the left ovary demonstrated reactivity for CD20 and CD10, and negativity for CD3, Bcl-2 and TdT. The C-myc translocation was positive in 60% of tumour cells. Moreover, the proliferation index was ~90%. These features were consistent with BL. After haemodialysis, the patient was planned for multiagent chemotherapy, including cyclophosphamide, doxorubicin, vincristine and prednisone. This case supports the hypothesis that primary ovarian BL is an aggressive malignancy that appears to respond promisingly to multi-agent chemotherapy.

Keywords: Bcl-2; Burkitt lymphoma; chemotherapy; CHOP; Non-Hodgkin lymphoma

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INTRODUCTION

Primary ovarian Burkitt lymphoma (BL)- a class of non-Hodgkin malignant neoplasms- is a very rare malignancy (i.e., <01%[1]), predominantly seen in young female patients. The BL presents a very aggressive character irrespective of the anatomical site, accentuating its prompt diagnosis. Epstein-Barr virus (EBV) is speculated as a possible cause of BL.[1] In the context of differential diagnosis, the three BL subtypes by histopathology features are endemic BL, sporadic BL and immunodeficiency-associated BL. The immunohistochemistry (IHC) features of the BL include C-myc translocation, very high proliferation index (i.e., Ki-67 ~ 90%), over-expression of B-cell antigens (e.g., CD22, CD19, CD79a, CD20), expression of Bcl-6 in tandem with CD10 and negativity of Bcl-2, CD5 and CD23 stains.[2] Clinically, patients with ovarian BL have abdominal tumours causing pain and discomfort, as well as impaired renal function, abnormal metabolism, extra-nodal involvement and invasion of contiguous organs. Albeit surgery is not the first-line treatment in the management of ovarian BL, the intervention is critical in the differential diagnosis (through immune-histological examination) and staging.[3] Moreover, intensive multi-agent chemotherapy regimens have been advocated for higher efficacy in patients with ovarian BL.[4] Although the outcomes of aggressive treatment are very optimistic, treatment-related toxicity continues to be a major challenge in the treatment of ovarian BL.[5] Here, we present the case of an 18-year-old female patient diagnosed with primary BL in the left ovary.

CASE REPORT

An 18-year-old female patient presented to the outpatient oncology clinic with lower abdominal pain, anorexia, fever and weight loss for the past two months. Clinical examination revealed a pale patient with a large, tender abdomen. The Eastern Cooperative Oncology Group performance status of the patient was three (ECOG-III) at the time of presentation.

A preliminary workup revealed a creatinine level of 12 milligrams per decilitre (mg/dL), urea of 150 mg/dL, lactate dehydrogenase (LDH) of 1200 units per litter (U/L) and completely de-ranged serum glutamic pyruvic transaminase (SGPT) of 600 U/L. The ultrasound of the abdomen showed a heterogeneous, hypoechoic pelvic mass of size ~15×10×6.4 cm, with increased vascularity. The ultrasonography also demonstrated hepatosplenomegaly, mild ascites and dilated biliary channels.

An immunohistopathological study of the biopsy sample (size ~5.3×2×2 cm) excised from the left ovary was performed, which demonstrated intermediate-size tumour cells organized in sheets. The pattern of neoplastic cells was a starry sky imparted by tangible body macrophages. The neoplastic cells had abundant basophilic cytoplasm, non-cleaved rounded nuclei with coarse chromatin and distinct nucleoli. Histochemical staining demonstrated expression of CD20 and CD10, and negativity for CD3, Bcl-2 and TdT, the C-myc translocation was positive in 60% of tumour cells in well-fixed areas. Moreover, the proliferation index (i.e., Ki-67) was ~90%. Figure-1 shows details of the immunohistopathological study. These features favoured B-cell Non-Hodgkin lymphoma of high-grade, consistent with BL.

Contrast-enhanced computed tomography (CT) showed renal parenchymal disease with moderate ascites, distended loops of the gut, mild obstructive biliary pattern and splenomegaly (Figure-2). These clinical and radiological workups rendered the disease stage IIIE. It may be noted that the highly compromised functions of the liver and renal system made it impossible to include positron emission tomography (PET) imaging in the staging workup.

Before chemotherapy, the patient underwent haemodialysis two times a week for one month (total of eight cycles). The patient was planned for multi-agent chemotherapy (i.e., CHOP), including doxorubicin; 50 mg/m2, cyclophosphamide; 750 mg/m2, prednisone;100 mg/m2 and vincristine; 1.4 mg/m2. Presently, only one cycle of the abovementioned chemotherapy has been administered, where the disease response was very good, clinically. The performance status of the patient improved from ECOG III to ECOG II.

It may be noted that this study was approved by the Ethical Review Committee of the Institute of Radiotherapy and Nuclear Medicine (IRNUM). Moreover, the patient provided written informed consent for publishing this case report.

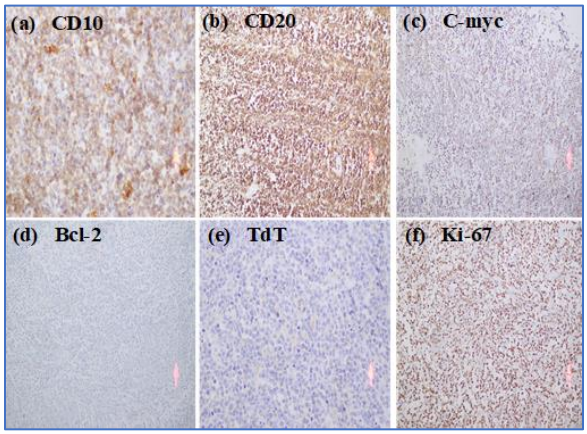

Figure-1: Illustrative images of immunohistopathology of the biopsy sample from the left ovary. Positivity of (a) CD20 (b) CD 10 (c) C-myc translocation, negativity of (d) Bcl-2 and (e) TdT and high proliferative index of Ki-67.

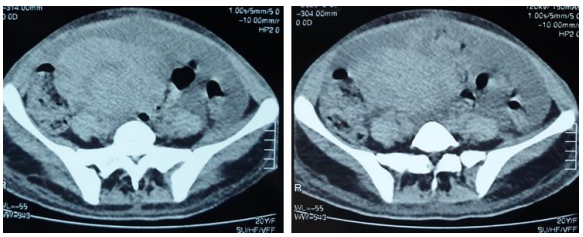

Figure-2: Illustrative images of computed tomographic study, showing renal parenchymal disease.

DISCUSSION

Burkitt's lymphoma (BL), with the primary occurrence of the disease in the ovary, is a rare clinical manifestation, accounting for less than 0.5% of NHL.[6] Primary ovarian BL usually occurs at a young age and is aggressive with a short proliferation cycle, enabling regional lymph node involvement and a greater possibility of early spread to distant sites. The case presented here followed the same pattern of age and aggression.

Lymphoma being a disease of the lymphatic system implicates the ovary as a secondary site. Expression of B cell markers (e.g., CD10, CD19, CD20 and CD22) by the primary ovarian BL indicates that it originates from the B cells; however, the consensus about its origin is illusory. In this direction, the following perspectives have been documented: a) Primary ovarian lymphoma may grow out of the lymphoid tissue of the ovary[7] or ovarian teratoma[8]; b) Lymphocytes present in the blood vessels surrounding the hilum and, in the corpus, luteum of the ovary may provide the primary site for the ovarian lymphoma[9]; c) Ovarian inflammation triggering reactive lymphocytes may undergo malignant transformation into primary ovarian lymphoma[10].

Chemotherapy is considered as the primary intervention in the treatment of ovarian lymphomas.[11] Specifically, the CHOP regimen (i.e., doxorubicin, cyclophosphamide, prednisone and vincristine) is predominantly advocated.[12,13]. However, the CHOP regimen is sometimes integrated with immunotherapy (e.g., rituximab: R-CHOP). Due to financial constraints, our patient received the CHOP chemotherapy only and showed a good response to the first cycle of chemotherapy, as the symptoms were relieved.

CONCLUSION

A young patient with primary ovarian Burkitt lymphoma (BL) was presented. The patient showed a promising response to a single cycle of chemotherapy (i.e., CHOP), as evidenced by an improvement in performance status from ECOG III to ECOG II. This case highlights the aggressive nature of primary ovarian BL and a favourable response to chemotherapy.

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Competing Interests

The authors declare no Conflict of Interest.

Ethical Committee Approval

This study was approved by the Ethical Review Committee of the Institute of Radiotherapy and Nuclear Medicine (IRNUM).

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