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Primary Ovarian Diffuse Large B-Cell Lymphoma in a 34 year old Lady: A Case Report

Dr. Shibina Sasidharan[1], Dr. Lila Rani Vijayaraghavan[2]

[1]Junior Resident, Dept of Pathology, Govt Medical College, Thiruvananthapuram, Kerala, India

[2]Head of The Department, Dept of Pathology, Govt Medical College, Thiruvananthapuram, Kerala, India

ABSTRACT

Involvement of the ovary by malignant lymphoma is well known as the late manifestation of disseminated disease. Primary ovarian lymphoma(POL) is extremely rare , accounting for 0.5 % of all NHLs and 1.5% of all malignant neoplasms[1] .

We here in describe a case of Primary ovarian Large B cell Lymphoma involving bilateral ovaries in a 34 year old female.Preoperative CT imaging showed a bilateral heterogenous soft tissue masses with cystic areas involving both ovaries. She underwent bilateral Ovarian cystectomy. Current diagnosis is based on histopathological immunohistochemical analysis. Patient died within one month of diagnosis.

KEYWORDS: Primary Ovarian Lymphoma, Primary Ovarian non-Hodgkin Lymphoma, Non-Hodgkin Lymphoma, Diffuse Large B-Cell Lymphoma

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1. INTRODUCTION

Among patients with disseminated Lymphoma, the ovary is relatively common site of involvement. However, primary ovarian lymphoma is an exceptional occurrence[2]. Among which Diffuse large B-Cell Lymphoma(DLBCL) is the most common type. Accounting for 20% of Primary Ovarian Lymphoma[3] . Exact origin of the primary ovarian lymphoma is unknown, however it is believed that it is arising de novo in the ovary from the lymphoid aggregates found in normal ovarian tissue[4]. Fox et al. have suggested three criteria for the diagnosis of primary ovarian lymphoma : (1) tumor confined to the ovary, regional lymph nodes or immediate adjacent organs at the time of the diagnosis, (2) bone marrow and peripheral blood have not contained any abnormal cells, and (3) if extraovarian disease appear later, there must be a few months between the time of ovarian and extra-ovarian lesions[5].

In this article, we present a case of primary ovarian large B-cell lymphoma in a 34 year old female, who was diagnosed based on histopathological and immunohistochemical findings.

2. CASE REPORT

A 34 year old female, who is a known case of Type 2 Diabetes Mellitus & Systemic Hypertension, presented with h/o abdominal distension of 1 month duration. No other illness in the past. General physical examination was normal. Per abdomen, ascites (+). Per vaginal examination was normal. Routine hematology, peripheral smear and chemistry showed no abnormalities. CA 125 -31.3, AFP-3.20, CEA – 3.3 ; all within normal limits.

CT Abdomen :Heterogenously enhancing soft tissue density masses with cystic areas involving both ovaries.Gross ascites. Possibility of bilateral malignant ovarian neoplasm.

She underwent Staging laparotomy and debulking surgery.

Gross:

Right ovary :Irregular nodular mass mea 9x7.5x4.5 cm. C/s grey white fleshy with multiple cystic and haemorrhagic spaces at periphery. Cystic spaces filled with solidified gelatinous material.

Left ovary :Nodular mass mea 10x4x5.5 cm. C/s grey white solid area and cystic spaces, filled with solidified gelatinous material.

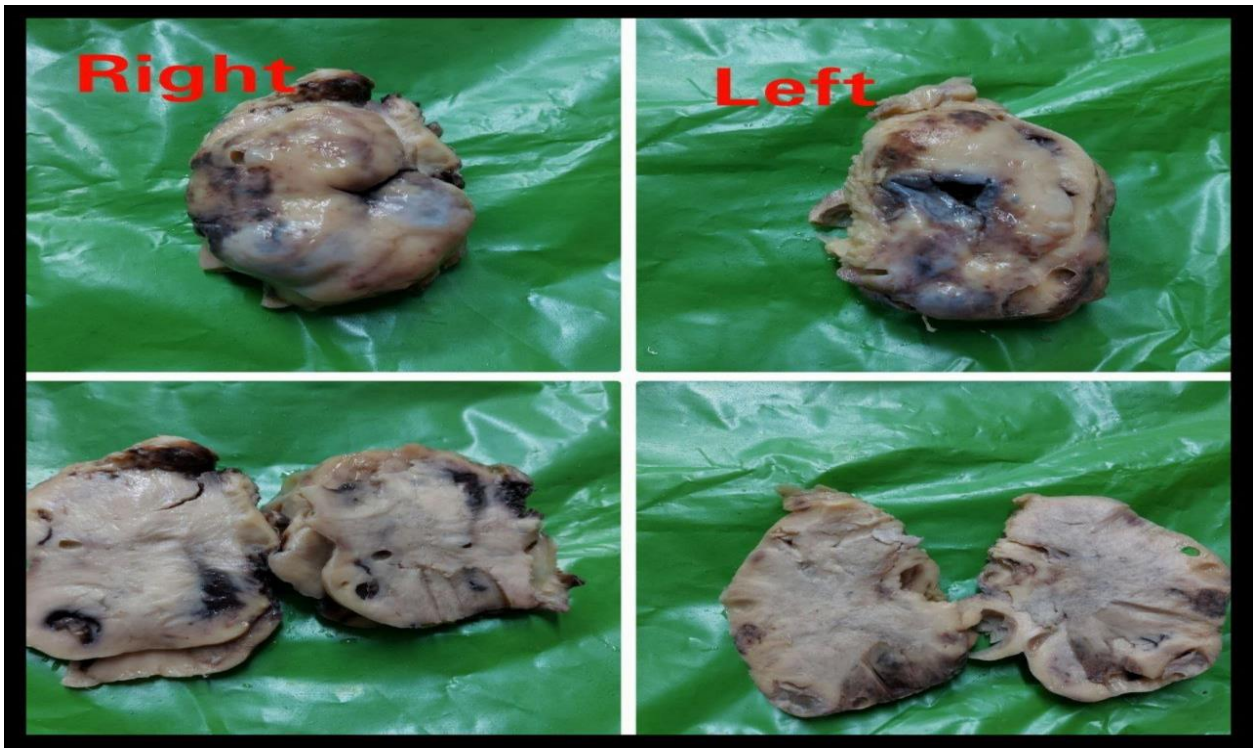

Figure 1 : Gross images showing outer surface and cut section of the enlarged right and left ovary, gray-white fleshy with multiple cystic and hemorrhagic spaces.

Microscopy:

Histology of the specimen revealed diffuse growth pattern of monomorphic tumor cells with large round to irregular nuclei, prominent nucleoli.

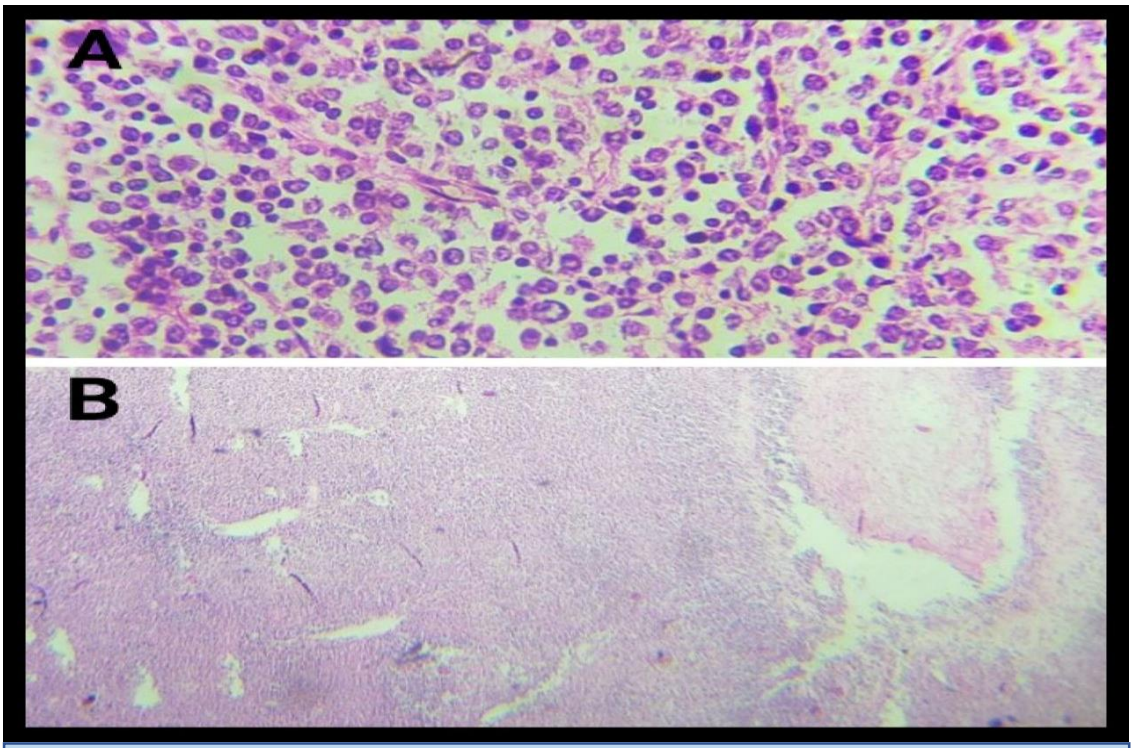

Figure 2 : (A) Sheets of tumor cells with high N:C ratio,round to irregular nuclei,coarse chromatin,prominent nucleoli,and scant cytoplasm.

(B) Tumor cells infiltrating to the ovarian tissue , show diffuse growth pattern.

Immunohistochemistry :

Paraffin immunostaining studies showed positivity of neoplastic cells for B cell CD20, BCL2, eucocyte common antigen(LCA) antibody. (Figure 3)

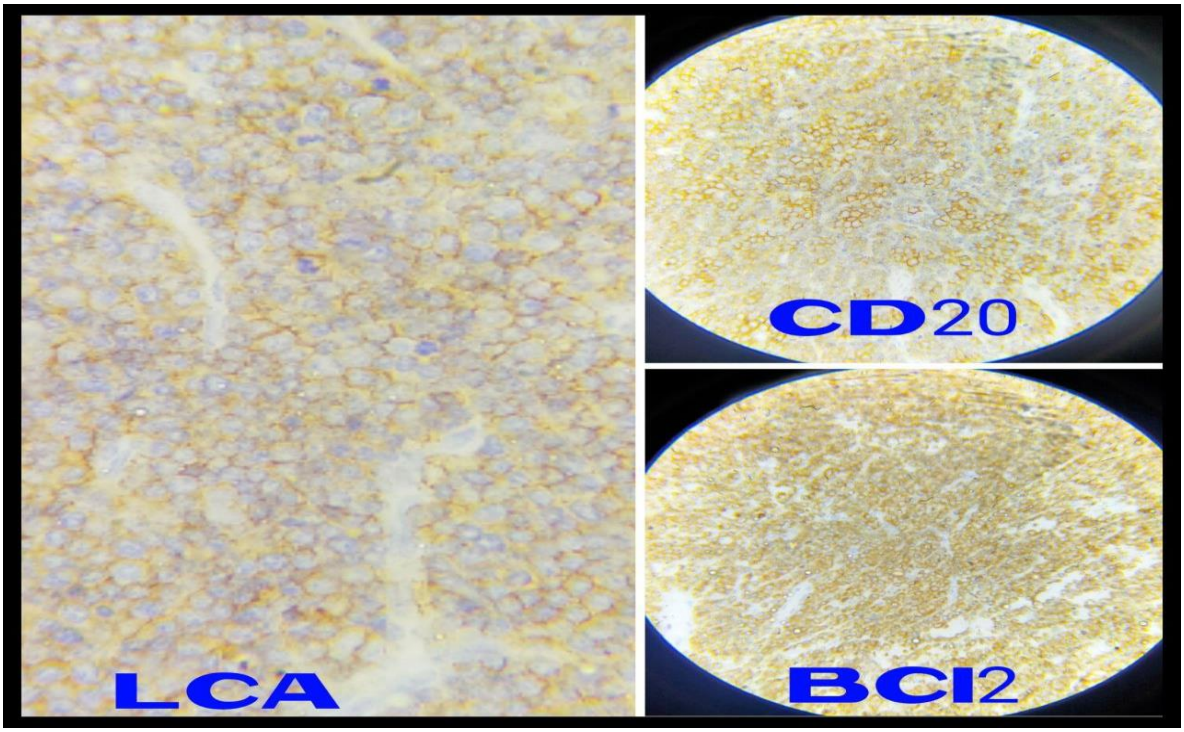

Figure 3 : Paraffin immunostaining studies showed positivity of LCA ,CD20,BCl2

No immunoreactivity was noted with EMA, OCT4, CK.

3. DISCUSSION

NHL is rarely involves female genital tract. This can be seen either as a late presentation of disseminated disease or as a primary NHL of the female genital tract, with the former being relatively common as compared with the latter[5 ,6]. There are a limited number of case reports describing POL. The distinction between POL and secondary ovarian involvement is of considerable importance as primary extranodal lymphomas have a less aggressive course and a better 5-year survival rate as compared with secondary disseminated disease [6].

A thorough literature search reveals that majority of the patients with POL are young with median age of 45 years[3]. Some of the patients may present with constitutional symptoms such as unexplained fever, weight loss, and night sweats, and may also have enlarged abdominal lymph nodes and/or hepatosplenomegaly. Bilateral ovarian involvement has been frequently reported in the literature [7,8]. Ultrasonographically the lesions are seen as hypoechoic mildly vascularized lesions and on CT these are seen as hypodense lesions exhibiting only mild enhancement with contrast[9].

Establishing a clinical diagnosis of POL is extremely difficult owing not only to its exceptional occurrence but also to the variable and nonspecific clinical presentations. Furthermore, establishing an accurate histopathological diagnosis is also challenging, due to the overlapping morphological features. Immunohistochemistry, thus, plays an important role in arriving at a final diagnosis, as in the index case.

The differential diagnosis of solid ovarian tumors includes, dysgerminoma, teratoma, metastasis and definitive diagnosis can be only confirmed by pathologic examination of the tumor tissue. Definite pathologic diagnosis was performed after immunostaining.

The presence of positive staining for leukocyte common antigen distinguishes malignant lymphoma from nonlymphoid neoplasms. In our case tumor cells were positive for LCA and B Cell marker CD20.

Disease stage was based on the Musshoff modification of the classical Ann Arbor staging system for extra-nodal lymphomas [10]. Stage I were patients with disease limited in single extra nodal site , stage II were patients with localised involvement of extra lymphatic organ or site on the same side of the diaphragm , stage III Involvement of organ on both sides of the diaphragm and stage IV were patients with disseminated disease.

Index patient had bilateral ovarian involvement and on evaluation no other primary site was detected. And she died shortly after the diagnosis indicating the aggressiveness of the disease.

4. CONCLUSION

In summary, primary ovarian non-Hodgkin's Lymphoma is rare. Among which primary ovarian DLBCL is most common and has a high proliferative activity and poor prognosis. Establishing an early and accurate clinical as well as histopathological diagnosis, although challenging, is essential for the appropriate management of these patients and to avoid unnecessary surgeries. Furthermore, it is important to exclude secondary involvement of the ovary by disseminated NHL in these patients, as the two entities carry significantly different prognosis.

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