Case Report

A Case of Primary Ovarian B-cell Non- Hodgkin's Lymphoma

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Abstract

Primary ovarian lymphoma presenting with ovarian mass as a primary disease is extremely rare. We report here a case of primary ovarian B-cell non-Hodgkin's lymphoma with bilateral involvement which was managed by surgery and chemotherapy. A 35 years old woman was admitted with signs and symptoms suggestive of ovarian cancer. The diagnosis of malignant lymphoma was established after bilateral adnexectomy, histological and immunohistochemical study of the excised tissue. The stage was I ES according to Ann-Arbor staging system. The patient was planned for R-CHOP chemotherapy, but due to unaffordbility patient was treated with 06 cycles of standard CHOP regimen and had completed her treatment. She has now been on follow-up and is without disease from 15 months of treatment. **Key Words:** NHL lymphoma, extranodal lymphoma, ovarian tumor.

INTRODUCTION

Non-Hodgkin's lymphomas (NHL) are heterogeneous group of lymphoproliferatve disorders originating in B and T- lymphocytes or both. NHL accounts for 4 - 5 % of new cases and 3% of cancer related deaths[1]. Number of classification systems has been developed for classification of NHL. The most widely used is that of World Health Organization (WHO) classification which is evolved from Revised European-American classification for lymphoid neoplasm (REAL classification) proposed by lymphoma study group. It classifies NHL on the basis of morphological, immunological and genetic characterization of the tumor. By giving different International Prognostic Index (IPI) scores for NHL, low risk (0 or 1), low intermediate risk (2), high intermediate risk (3), high risk (4 or 5) disease states are described. Extra nodal NHL occurs in 25% of cases, while malignant lymphomas occurring in female genital tract are very rare, accounting for only 1% or less cases[2]. But primary ovarian lymphomas are unusual[3]. We present a case of ovarian non-Hodgkin's lymphoma, presenting like ovarian cancer, which was managed by surgery and chemotherapy.

CASE REPORT

A 35 years old woman presented to gynecologist with one year history of abdominal pain with distension, constipation, and loss of appetite. She had no relevant past and medical or family history. Physical examination per abdomen revealed a huge mass of 20cm, in right side of abdomen with lower edge of mass unremarkable, mass was non-tender with smooth surface, with no ascites. Per rectal examination showed rectal mucosa is free and uterus was felt separately from rectal mucosa. Neither Liver nor spleen was palpable, no adenopathy were noted. Pre-operative ultrasound showed a large solid heterogeneous mass in right adnexal region measuring 20.5 x 12.1 cm in size, and metastatic deposits on spleen with moderate ascites. Serum tumor markers were positive for serum CA-125: 610 IU/ml (normal range < 35 IU/ml), serum lactate dehydrogenase (LDH): 710 IU/L (normal range 200-500), and other tumor markers serum alpha feto-protien (AFP) and serum Beta-hcg were normal. An exploratory laparotomy was done through a large abdominal vertical midline incision so as to establish

the diagnosis followed by total abdominal hysterectomy with bilateral salpingo-oophorectomy and omentectomy. Specimens for histopathological assessments consisted of uterus with bilateral adnexa, sections from endo and ecto cervix which were tumor free with chronic non-specific inflammation. Section from fallopian tubes identified which were unremarkable. A section from endomyometrium was also seen and exhibited autolyzed endometrium and myometrium showed non-specific pathologic change. Cut surface of both ovaries showed solid gray white tumor with autolysis, which were composed of sheets of intermediate and large sized cells and moderate amount of cytoplasm, pleomorphic vesicular nuclei with irregular contours, vesicular chromatin, and prominent nucleoli. The tumor was negative for glycogen as seen on special stain (PAS +/-D). The sections were stained with following Antibodies using Envision System.

LCA :	Positive
CD 20:	Positive
CD 30:	Negative
Mib-1:	Positive in 65-70% cells.

Pathologic assessments were consistent with non-hodgkin's lymphoma with morphological and immunohistochemical features favoring diffuse large B- cell lymphoma.

Further investigations like Bone marrow aspiration cytology showed all cell lines: Erythroid series were active and showed normoblastic features, myeloid series showed all stages of maturation and differentiation. Adequate megakaryocytes were present, Blast cells 3%, Plasma cells 4%. Overall features showed no evidence of bone marrow involvement by lymphoma. Post-operative ultrasound done after 02 months which showed splenomegaly with multiple rounded irregular hypoechoic masses suggestive of splenic metastasis. Uterus and ovaries were not visualized. A large irregular solid hypoechoic and heterogeneous mass was seen which could be due to recurrent growth. Patient was prescribed six cycles of standard CHOP regimen (cyclophosphamide 750 mg/m² d1, doxorubicine 50 mg/m² d1, vincristine 1.4 mg/m² d1, and prednisolone 100 mg/m² d1-5). She had completed all six cycles and 15 months after surgery, she remained disease free.

Figure-1 (before treatment)

Ultrasonographic study showed huge pelvic Mass

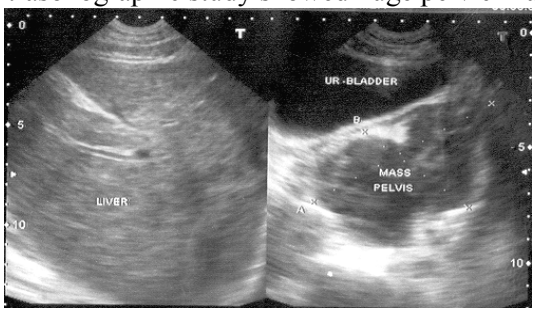


Figure-2 (before treatment)

Showed multiple metastatic deposits on Spleen

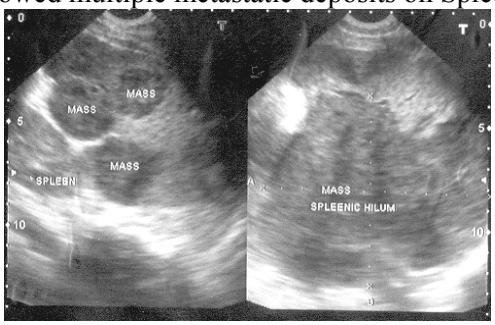


Figure 3 (after treatment)

Ultrasonographic study showed no any growth per abdomen

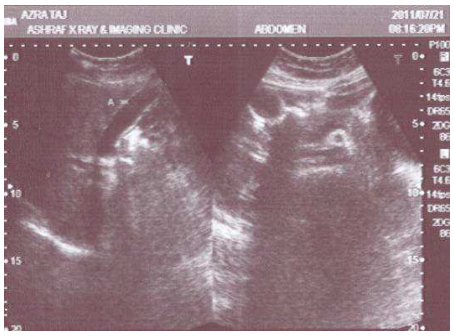
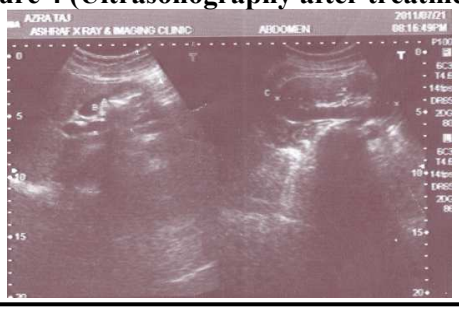


Figure 4 (Ultrasonography after treatment)



DISCUSSION

Lymphoma is a rare tumor of ovary and its presence most commonly represents involvement in overt systemic disease, almost always of non-Hodgkin's type[3]. The diffuse large B-cell lymphoma appears to be the most common type of primary ovarian non-Hodgkin's lymphoma[4]. Non-Hodgkin's lymphoma may involve lymph nodes in almost any area of the body but may also present in extra nodal sites, presumably arising from lymphoid tissue widely distributed throughout the body. Patients presenting with extra nodal lymphoma usually have localized disease, the symptoms relate to site of involvement. GIT is most commonly involved site for extranodal lymphoma accounts for 25% to 35% of cases followed closely by Waldeyer's ring and other head and neck sites (18% – 28%) and skin[5]. While presentation in ovary as primary lymphoma is even rarer. Determined from literature, Patients with primary ovarian lymphoma have a mean age of 35 years (range 06 - 74 years) and most of Patients present with pelvic or abdominal pain and mass. The presence of bilateral involvement is an uncommon feature; the mean tumor size is 10.5cm [6]. Fox et al[7] suggested three criteria for diagnosing primary ovarian lymphoma a: Tumor is confined to ovary, regional lymph nodes or adjunctive organs at the time of diagnosis, as in our case. b: The peripheral blood and bone marrow should not contain any abnormal Cells, as it is the case in our Patient c: The lymphomateous lesions that occur at the sites remote from ovary, at least several months should have elapsed between appearance of ovarian and extra ovarian lesions [7]. In present case, there was no obvious lymphadenopathy at the time of diagnosis and during a period of one year follow up. Peripheral blood examination revealed no any atypical cells. Malignant lymphoma in ovary may be confused with other primary ovarian tumors. Involvement of the fallopian tubes and broad ligaments are more common in lymphomas than in most of the tumors in differential diagnosis. However in this case both of the fallopian tubes were free from any tumor infiltration. The differential diagnoses of ovarian lymphoma are dysgerminoma, granulocytic sarcoma, undifferentiated carcinoma and metastatic breast carcinoma Dysgerminoma is most important one and may mimic lymphoma both macroscopically and microscopically [8]. Komoto et al [9], suggested that CT is the mainstay of lymphoma staging in the chest, abdomen, and pelvis as well as in other nodal lymphomas. Bone marrow biopsy is also mandatory for staging. Positron emission tomography (PET) with 18F-flourodeoxyglucose, (FDG) has been reported as a useful method for staging and for assessment of the therapeutic response [9].However for ovarian lymphoma there are variety of diagnostic imaging tools can be useful. The US pattern of lesions were aspecific, homogenous and hypoechoic; color Doppler US showed mild vascularization. CT showed clear cut lesions, hypodense and with mild contrast enhancement in all cases. MRI showed homogenous masses which were moderately hypointense on T1-weighted images and slightly hyperintense on T2-weighted images. GD-T1 weighted images showed mild enhancement [3]. Patients with localized disease to one ovary usually do well with unilateral surgical resection followed by systemic chemotherapy[10]. The use of chemotherapy is based on the principle that ovarian lymphoma must be considered as a localized manifestation of systemic disease [7]. The prognosis of such patients is much better than that of patients with obvious systemic disease [10]. Signorelli et al, suggested that in primary ovarian lymphoma stages I-II, a conservative management based on exclusive chemotherapy may be attempted in selected patients desiring pregnancy. The protocol of chemotherapy used in diffuse, large B-cell histology is the standard CHOP regimen. The outcome of these patients, treated with appropriate chemotherapy, appears to be similar to that of patients with other nodal non-Hodgkin's lymphomas [11].

Our patient was treated with total abdominal hysterectomy and bilateral salpingo-oophorectomy and Omentectomy followed by six cycles of CHOP.

Primary ovarian non-Hodgkin's lymphoma is very rare, patients usually present with pelvic complaints, but in

some cases may be detected as an incidental finding. The neoplasm may be microscopic or very large and are usually unilateral, either B-cell or T-cell NHL can arise from ovary but B-cell tumors are common.

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