Non-Hodgekin's lymphoma of female genital tract: a case report

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Abstract

Non-Hodgkin lymphoma (NHL) causes many deaths worldwide with increasing incidence. Non-Hodgkin's lymphoma (NHL) may involve the gynecologic tract, and the ovary to be one of the commoner anatomic sites as reported. Ovarian involvement by NHL is usually secondary, occurring as a part of systemic disease. The diagnosis is often made incidentally while investigating for gynecological symptoms. We report an interesting case of NHL in a 35 year old female, who primarily presented with acute abdomen disease with secondary ovarian and cervical involvement and occult extra-ovarian disease.

Keywords: Cervix, Ovary, Lymphoma, NHL.

Introduction

Non-Hodgkin's lymphoma (NHL) may involve the gynecologic tract, with the ovary as a more common anatomic sites (1). It has been seen that Non-Hodgkin's lymphoma usually involves the ovaries secondarily (2). Localized, presumably primary NHL of the ovary is rare (3). Primary ovarian lymphoma accounts for 0.5% of NHL and 1.5% of ovarian tumors. The occurrence of lymphomas primarily arising in the ovaries has long been debated since no lymphoid tissue found in the ovaries and the cells of origin are considered to be lymphocytes present in the ovary, surrounding vessels and the hilum (4) We present a case of ovarian lymphoma presenting as acute abdominal pain due to ruptured ovarian mass and an occult extra-ovarian disease.

Case report

A 35-year-old woman, with three children

and last child born six year old, presented with complaints of increasing pain in the abdomen and dyspepsia of 3 months' duration, fowl smelling vaginal discharge for one week and dizziness for one day. Previously, there was history of weight loss, anorexia, night sweats and low grade fever for 3 months. She had consultation from various doctors regarding lethargy and weakness. Her bone marrow biopsy which was done earlier for evaluation of anemia had no abnormality.

Physical examination revealed a tender mass of 22 weeks size in the left iliac fossa, which appeared to be arising from the pelvis and extending up to the umbilicus. On speculum examination, cervix was hypertrophied with irregular margins with fowl smelling vaginal discharge. Per vaginal examination revealed a mass in the pouch of Douglas pushing the uterus to the right side.

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Urgent baseline laboratory investigations along with tumor markers including CA-125, Lacatate dehydrogenase, serum beta HCG and alpha fetoprotein were analyzed. An abdomino-pelvic ultrasound revealed a left ovarian mass 12.1 x 10.4 cm with moderate vascularity and a right ovarian mass 6.7 x 4.9 cm with high vascularity along with free fluid in peritoneal cavity reaching up to paracolic gutters. Both CA-125 and Lactate dehydrogenase levels were high.

At the time of consultation, patient had a fainting attack, and went into shock. Resuscitation was done and patient rushed to E.R. Since it was an emergency case, there was no time for pre-op and she was arranged for needed frozen section at the time of operation.

Since she suffered from acute pain in abdomen, exploratory laparotomy was done which revealed presence of three litres blood in peritoneal cavity with ruptured left ovarian cyst 10 x 8 cm with solid component and right ovarian mass of 5 x 5 cm with intact capsule and irregular surface. Total abdominal hysterectomy with bilateral salpingo-oophorectomy and omenectomy were performed.

On gross examination, the left ovarian mass measured 9.0 x 6.0 x 4.0 cm. Right ovary measured 3.5 x 2.0 x 1.0 cm with intact capsule. Cut surface of both ovaries showed fleshy, grayish white homogenous cut surface. Separate multiple nodules were seen. Uterus weighed 120 grams and measured 10 cm from fundus to cervix, while ectocervix measured 3.5 x 2.5 cm. Cervix was shiny white with a greysih white nodular area measuring 3.5 x 2.5 x 1.0 cm on posterior aspect of the cervix.

Microscopic examination of ovarian masses and cervix revealed tissue being replaced by lymphoid atypical infiltrate comprising medium to large size lymphoid cells with pleomorphic, hyperchromatic nuclei, indiscrict nucleoli and scanty cytoplasm. At places, starry-sky appearance was seen. Abundant mitosis and apoptosis was also noted. (Figs I and II)

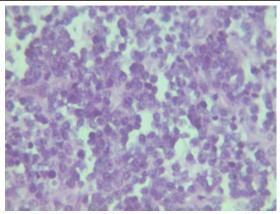


Fig. 1. The ovarian tissue with lymphoma (High magnification).

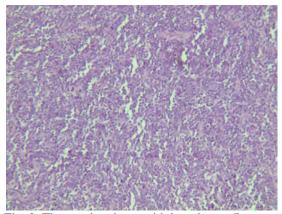


Fig. 2. The ovarian tissue with lymphoma (Low Magnification)

Immunohistochemically, the cells were positive for LCA and negative for cytokeratin, which confirmed the diagnosis of lymphoma. Sections from the omentum revealed focal collections of these lymphoid indicating diffuse non-Hodgkin's lymphoma (probably diffuse large-cell lymphoma) involving the ovary.

The patient was evaluated postoperatively with an abdominal CT scan, which revealed multiple enlarged lymph nodes involving the common iliac, internal iliac and external iliac groups. Bone marrow aspirates and trephine biopsy confirmed marrow involvement. A diagnosis of stage IV (Ann Arbor staging) non-Hodgkin's lymphoma with secondary ovarian involvement with an initial clinical presentation of occult extra-ovarian disease was made.

A chemotherapy protocol comprising 6 cycles of CHOP regime was administered

to the patient, which responded well to the therapy.

Haematoxylin- and eosin-stained section showing a diffuse population of medium to large sized cells with scattered macrophages

Discussion

Diffuse large B-cell lymphoma is the most common subtype of NHL. It can occasionally present in extranodal sites like gastro-intestinal tract, thyroid, skin, female genital organs etc (5). About 33% of NHL arise in tissues other than the lymph nodes, spleen, Waldeyer's ring and thymus and these are referred to as primary extranodal NHL (6). In our case, both ovaries and cervix were involved as part of the generalized disease. Our patient had a very unusual presentation with acute pain in abdomen and discharge per vaginum, while the usual symptoms of lymphoma like fever, weight loss and peripheral lymphadenopathy were appeared in onset for 3 months.

The ovarian involvement in malignant lymphoma can be primary or secondary. The secondary involvement has been described of being of 2 types (1): It can present as an initial clinical presentation of occult extra-ovarian disease or (2) it can be as a manifestation of widely disseminated disease (1). It becomes prudent to differentiate between the two because the primary extra-nodal lymphomas run a less aggressive course and have a 5-year survival rate of 80% as compared to the malignant lymphomas, which have a 5-year survival rate of only 33%. (8) Lymphomas of the ovary have been seen to occur at any age, but mostly women in their 40s are affected (7,8).

There has been much controversy in defining what constitutes a primary ovarian lymphoma. It has been proposed that there should be presence of an ovarian mass, confined to one or both ovaries. The lymphoma should be considered as an ovarian primary even if only microscopic involvement of the contiguous lymph nodes is detected. Also, the peripheral blood and the

bone marrow should not contain any abnormal cells and months should have elapsed between the appearance of the ovarian and the extraovarian masses (9).

Paladugu et al (10) proposed that there should be a disease-free interval of at least 60 months after the oophorectomy.

Since the clinical presentation of our patient had no evidence of generalized lymphadenopathy preoperatively and intraoperatively, a diagnosis of primary ovarian lymphoma was considered. However, on subsequent CT evaluation and multiple abdominal lymph node involvement, a diagnosis of ovarian lymphoma with an initial ovarian presentation of occult extra-ovarian disease was considered, and the patient treated accordingly.

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