Case Report

Primary non-Hodgkin's lymphoma of the female genital tract in a 27-year-old female: A rare case report

Pooja Srivastava, Charanjeet Ahluwalia, Sufian Zaheer, Ashish Kumar Mandal

Department of Pathology, VMMC and Safdarjung Hospital, New Delhi, India

ABSTRACT

Primary non-Hodgkin's lymphoma (NHL) of the female genital tract is a rare tumor mainly affecting the elderly age group. A preoperative diagnosis is difficult to reach due to varied clinical presentation and lack of diagnostic features on radiological investigations. We present an unusual case of primary NHL affecting uterus, cervix, and bilateral ovaries in a 27-year-old female.

Key words: Non-Hodgkin's lymphoma, female genital tract, primary

INTRODUCTION

Non-Hodgkin's lymphoma (NHL) can involve extranodal sites. The most common primary extranodal location is the gastrointestinal tract, other sites include bone, testis, salivary gland, thyroid, liver, kidney, and adrenal gland.[1] The uterine cervix is the most common primary site in the genital tract, and the ovary constitutes the most common secondary site of lymphomatous involvement.[2] We present a case of primary lymphoma of the female genital tract with uterus, cervix, and bilateral ovarian involvement in a young female.

CASE REPORT

A 27-year-old nulliparous female came with the complaints of severe abdominal pain and swelling since 20 days. Her menstrual history was regular. Clinical examination revealed an abdominopelvic tender mass in the right iliac fossa about 12 cm × 10 cm in size extending up to the hypogastrium which was firm in consistency with restricted mobility in both directions. There was also a past history of tuberculosis for which she had taken antituberculosis therapy for a year. Ultrasound and contrast-enhanced computerized tomography were done which revealed uterus of size 11.5 cm × 7.9 cm × 7.5 cm with a well-defined hypoechoic lesion of 8 cm × 7.4 cm × 7 cm displacing endometrium suggestive of posterior wall fibroid. Right ovary showed a solid lesion of 14 cm × 13 cm × 10 cm in the right adnexa and left ovary showed a homogenous solid lesion with enhancement in the left adnexa of 11 cm × 9 cm × 8 cm. Minimal ascites was also present. Serum levels of tumor markers were CA 125 63.83 U/ml (normal <35 U/ml), CA 19-9 0.10 U/ml (0–37 U/ml), AFP 1.23 ng/ml (<6.0 ng/mL), and lactate dehydrogenase 799 IU/L (105–333 IU/L). Since she was suffering from acute pain in the abdomen, exploratory laparotomy was done, and total abdominal hysterectomy with bilateral salpingo-oophorectomy and infra colic omentectomy were performed.

On gross examination, the left ovarian mass measured 12 cm × 7 cm × 6 cm and the right ovary measured 12 cm × 11 cm × 8 cm. Capsules of both the ovaries were

Address for correspondence: Dr. Pooja Srivastava, 270 A Hauz Rani Malviya Nagar, New Delhi, India.
E-mail: panacea.2417@gmail.com

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intact and cut surface was fleshy, grayish white homogenous in appearance [Figure 1]. Few hemorrhagic areas were also seen. Uterus measured 13 cm × 4 cm × 5 cm, whereas ectocervix measured 5 cm in length. Cut surface of both uterus and cervix were also gray white and homogenous.

Microscopic examination of ovarian masses, uterus, and cervix revealed sheets of large neoplastic cells that infiltrated deeply through the stroma [Figures 2 and 3]. Cytologically, these cells were of medium to large size with moderate amount of eosinophilic cytoplasm [Figure 4]. Mitosis was conspicuous. On immunohistochemistry, these cells were positive for leukocyte common antigen [Figure 5], CD20, CD10, and BCL2 and negative for CD3, vimentin, and desmin. Since there was the absence of any other nodal or extranodal lymphoid proliferation, a final diagnosis of diffuse large B-cell lymphoma (DLBCL), primary in uterine myometrium with involvement of cervix, and bilateral ovaries was given. The patient was further treated with chemotherapy and was kept on regular follow-up. After 4 months of follow-up, there was no recurrence of disease.

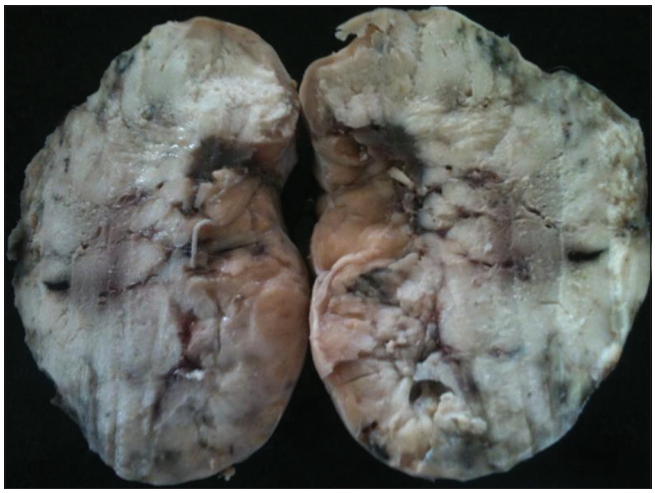

Figure 1: Gross photograph showing fleshy, grayish white homogenous cut surface of the ovary

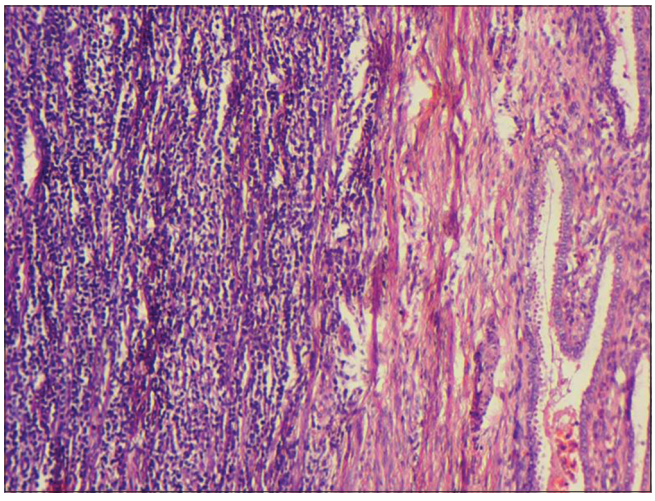

Figure 3: Sheets of neoplastic cells infiltrating into the myometrium (H and E, ×40)

DISCUSSION

Extranodal NHLs are commonly subdivided in literature as primary and secondary.[3] When NHL arises in tissues other than lymph nodes, spleen, Waldeyer's ring, and thymus, these are referred to as primary extranodal NHL.[4] Primary NHL of the female genital tract is a rare disease accounting for around 2% of all extranodal lymphomas.[5] Involvement of uterus and cervix in primary NHL is found in around 0.5% of cases, whereas the involvement of uterine corpus and bilateral ovaries, as seen in the case being presented, is extremely rare.[6] Primary involvement means unequivocal absence of nodal or extranodal disease at the time of presentation or 6 months around diagnosis.[1] Here, in this

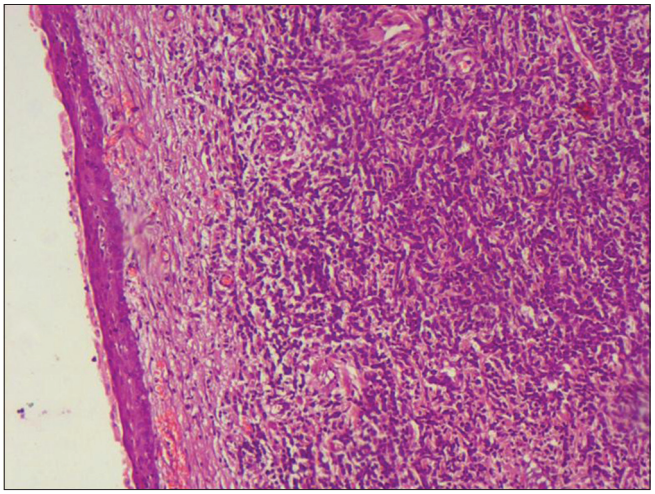

Figure 2: Large neoplastic cells diffusely infiltrating deeply into the stroma of cervix (H and E, ×40)

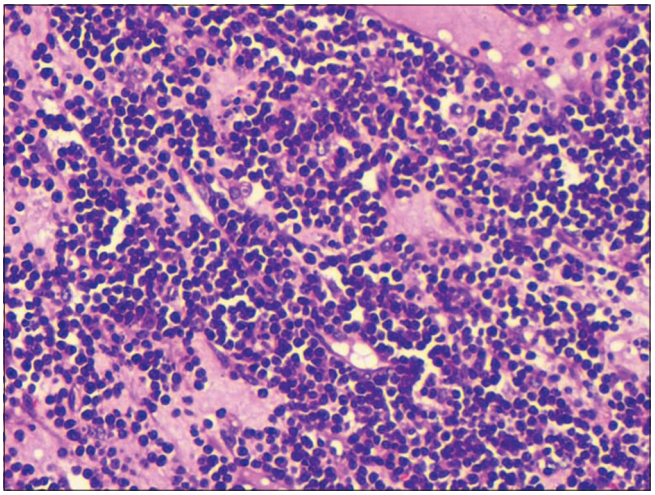

Figure 4: Sheets of medium to large size cells with moderate amount of eosinophilic cytoplasm and coarse chromatin seen to diffusely infiltrate the stroma (H and E, ×100)

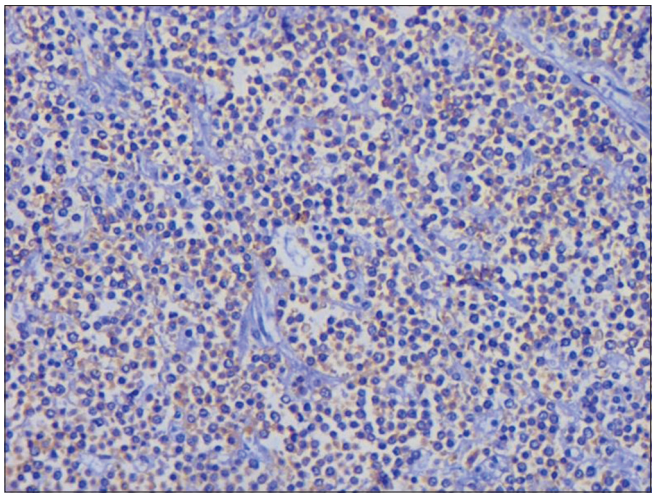

Figure 5: Leukocyte common antigen was strongly positive in the tumor cells (IHC, ×100)

case, it was difficult to say whether the uterine lymphoma is secondary to the ovarian primary lymphoma metastatic to uterine body or a primary uterine lymphoma metastatic to bilateral ovaries. A similar case was reported by Jeong *et al*.[6] where a 67-year-old female presented with lymphoma of myometrium involving bilateral ovaries.

The most common age of occurrence of primary NHL involving female genital tract is around 50 years.[3] However, in this case, the patient was just 27-year-old which further aggravated the diagnostic difficulty, as these tumors are infrequently reported in the younger age group. Generally, the clinical presentation is highly variable with dysfunctional uterine bleeding followed by pelvic pain frequently associated with a palpable mass.[3] Some patients may complain of urinary symptoms because of the extrinsic compression of the bladder, as well as other urinary structures, by an enlarging mass within genital tract.[7] Because of the diverse clinical presentation and infrequent nature of the disease, it presents diagnostic difficulties both for the radiologist and pathologist. Furthermore, as the radiological features are not diagnostic of lymphoma,[1] thus a preoperative diagnosis is difficult to reach.

Even on microscopy, there is a wide range of differential diagnosis of NHL of female genital tract. It includes inflammatory conditions, endometrial stromal tumors, malignant mixed Mullerian tumor, extraosseous Ewing's sarcoma, hematological neoplasm, and melanoma.[1] Reactive inflammatory infiltrates comprise polymorphous population of cells without atypia[8] and these cells are superficial, not showing infiltration in the stroma. In differentiating from endometrial stromal tumors, the presence of monomorphic population of lymphoid cells showing atypia along with lymphoid staining profile on immunohistochemistry and lack of sarcomatoid spindling pattern and prominent arborizing vasculature helps to clinch the diagnosis of lymphoma.[9] Ewing's sarcoma lacks positivity for lymphoid markers except CD99.[3] Hematological neoplasms such as granulocytic sarcoma show the presence of myelocytes and are positive for myeloperoxidase, lysozyme, CD68, and lack lymphoid markers.[10] Poorly differentiated epithelial tumors may cause diagnostic difficulty, but presence of cords, nests, glands, or associations with an *in situ* component help discern their epithelial nature.[3]

Treatment modalities include different combinations of surgery, chemotherapy, and radiotherapy. The prognosis usually remains poor with 5 years survival reported by a recent study on lymphoma involving female genital tract as 39.3%.[11]

CONCLUSION

NHL involving female genital tract is a rare neoplasm and can present not only in elderly but also in young patients. Hence, it should be kept in the differential diagnosis whenever radiological findings are not characteristic and patient presents with multiple lesions involving uterus, cervix, and ovaries. A high degree of suspicion is needed to diagnose these unfamiliar neoplasms for proper patient management.

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Conflicts of interest
There are no conflicts of interest.

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