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Case Report

A rare localization of a common disease: Primary uterine Non-Hodgkin lymphoma mimicking leiomyosarcoma

ABSTRACT

Genitourinary system lymphomas comprise a small part of extra-nodal lymphomas (ENLs). ENLs of uterine origin are extremely rare and are often confused with gynecological malignancies. We present an 80-year-old female patient diagnosed with diffuse large B-cell lymphoma (DLBCL) with a single focus of the uterus. The patient's only complaint was abnormal uterine bleeding. Magnetic resonance imaging revealed an intramural-subserous-submucous multiple mass lesion with minimal contrast enhancement mimicking leiomyosarcoma. Diffuse pathological 18F-fluorodeoxyglucose uptake was detected in the entire uterus corpus and cervix on positron emission tomography/computed tomography (PET/CT) scanning. The pathology of the endocervical and endometrial curettage material obtained was DLBCL; the patient was diagnosed with ENL, and a single focus was the uterus. ENLs should be considered in the differential diagnosis of gynecological malignancies in patients with abnormal uterine bleeding. PET/CT is crucial in showing metabolically active spread areas in these patients.

KEY WORDS: Extra-nodal lymphoma, FDG PET/CT, post-menopausal bleeding, uterus

INTRODUCTION

Extra-nodal lymphoma (ENL) is seen in approximately one-third of all non-Hodgkin lymphomas (NHLs) and has a poor prognosis.[1] Genital system lymphomas constitute less than 1% of ENLs, most of which are diffuse large B-cell lymphoma (DLBCL) sub-types.[2] Primary uterine lymphoma is quite rare and presents with abnormal uterine bleeding.[3-5] In the literature, a limited number of cases of DLBCL are located in the only uterus.[6] We present a patient with ENL with a single focus of the uterus.

CASE REPORT

This case report presented an 80-year-old female patient with only abnormal post-menopausal uterine bleeding and who scanned for this complaint. The patient, who had recurrent bleeding several times, also complained of fatigue in addition to this complaint. The patient had no additional complaints other than fatigue. She had a history of hypertension and type-2 diabetes mellitus. There was no feature in the family history.

No visible mass was observed in the patient's physical examination, who applied to the gynecology clinic with abnormal post-menopausal uterine bleeding. In the lower abdomen, magnetic resonance imaging (MRI) was taken with the preliminary diagnosis of gynecological malignancy. The size of the uterus was large (121 × 76 × 110 cm) on MRI scanning, and additionally, multiple mass lesions with intramural-subserous-submucous minimal contrast enhancement that filled the uterus were detected on abdomen MRI.

Multiple conglomerated hyper-metabolic mass lesions [(maximum standardized uptake value (SUVmax: 32.6)] were detected, filling the entire uterine corpus and extending to the cervix, according to the fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) scan of the patient with suspected leiomyosarcoma. There was no finding to support lymphoid infiltration in any focus other than the uterus on PET/CT scan. Whole-body PET/CT imaging of the patient is shown in Figure 1.

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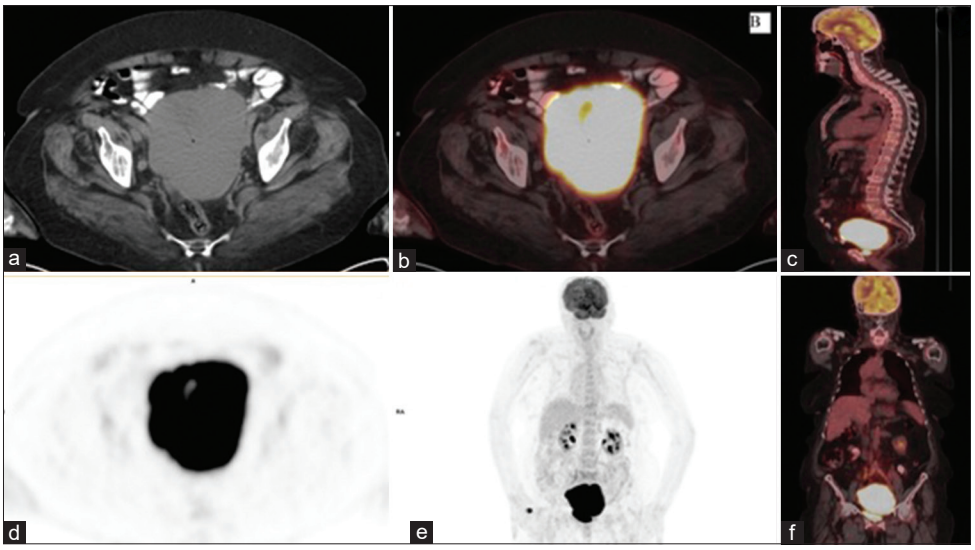

Figure 1: Axial section BT imaging (a), axial (b) and sagittal (c) section fusion images, axial section PET images (d), whole-body maximum intensity projection (MIP) image (e), and coronal section fusion image (f) on FDG PET/CT scan showing diffuse uterine involvement

Endocervical and endometrial curettage was applied to the patient. Microscopic examination of endometrial curettage materials revealed a tumor consisting of atypical lymphoid cells with large hyperchromatic prominent nuclei and diffuse infiltration in the endometrium. In immunohistochemical staining applied to the curettage material, PanSK (-), Sall4 (-), CD45 (+), CD3 (-), CD20 diffuse (+), MUM-1 (-), CD10 weak (+), Bcl-2 diffuse (+), Bcl-6 (+)), C-myc (-), and a high Ki-67 proliferation index (90%) were detected. The patient was diagnosed with DLBCL with the germinal center phenotype with the pathology of the endometrium and cervix curettage material. Images of hematoxylin-eosin (HE) and immunohistochemical staining of endometrial and endocervical curettage specimens are shown in Figure 2.

When the complete blood count (CBC) and biochemistry parameters of the patient were examined, hematocrit 37.3% (reference range: 42–52), sedimentation: 54 mm/h (reference range: 0–20), antistreptolysin-O: 58.1 IU/mL (reference range: <240), lambda light chain: 1950 mg/L (reference range: 900–2100), beta-2 microglobulin: 3.71 mg/L (reference range: 1.09–2.53), lactate dehydrogenase: 781 U/L (reference range: <248), and C-reactive protein: 43.5 mg/L (reference range: <5) were detected. Other CBC and biochemistry parameters were within normal limits.

The chemotherapy protocol was started for the patient diagnosed with primary uterine DLBCL and had a single extra-nodal focus uterus.

DISCUSSION

The incidence of primary internal genital tract lymphomas is rare in clinical practice. Women usually apply to the gynecology department with abnormal uterine bleeding. As stated in the study of Schniederjan and Osunkoya, genitourinary system (GUS) lymphomas give symptoms according to the organ involved.[7] It should be remembered that extra-nodal NHL in women may only give symptoms such as abnormal uterine bleeding. Although mostly single organ involvement is seen in the GUS origin NHLs, more than one organ involvement has also been seen.

18F-Fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT) is an oncological imaging method that provides anatomical and metabolic information about lesions. The contribution of FDG PET/CT in demonstrating malignant lymphomatous infiltration of normal-sized organs or lymph nodes is indisputable in many studies on lymphoma patients. In a study, after FDG PET/CT scan of patients diagnosed with NHL, the stage was increased by approximately 31% and decreased by 1%, leading to a change in treatment modality in approximately 25% of patients.[8] This metabolic imaging method successfully detects unexpected extra-nodal infiltrations that cannot be detected in other conventional imaging methods.[6]

Differentiating primary GUS lymphomas from gynecological malignancies is difficult in the pre-operative period. These patients are primarily operated on with a preliminary diagnosis of gynecological malignancy and are diagnosed with the pathology result of the surgical material.[2] In these patients, hysterectomy is performed for diagnostic purposes and not for treatment, and the patient is exposed to unnecessary surgical intervention.[9] As we understood in this patient, any imaging modality may not distinguish between uterine leiomyosarcoma and primary lymphoma of the uterus. If there is a hyper-metabolic lesion in the uterus on PET/CT imaging presenting with abnormal vaginal bleeding, histopathological verification is needed before the surgery decision. Surgical treatment has no place in the primary uterine DLBCL.

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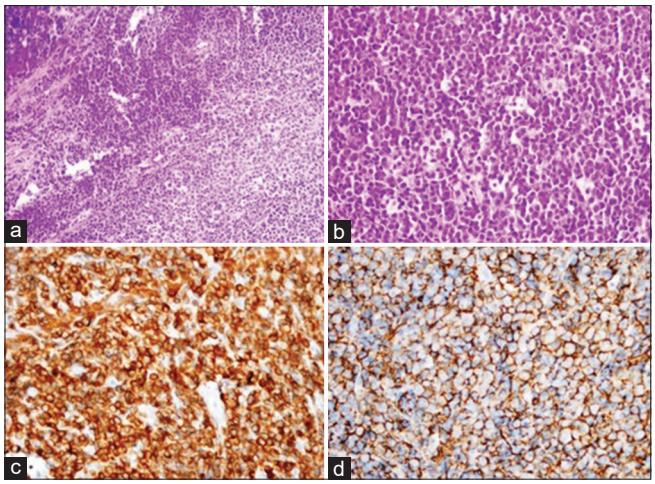

Figure 2: Histopathological specimen shows diffuse infiltrating neoplastic lymphoid cells with HEX200 (a), HEX400 (b), BCL-2 (c), and CD-20 (d)

Diffuse FDG uptake on the only internal genital organ on FDG PET/CT scan without the extensive disease can be seen in ENL. Although the uterus and cervix involvement of DLBCL is quite rare, primary ENLs should also be included in the differential diagnosis of abnormal vaginal bleeding in post-menopausal women. Accurate imaging contributes to the correct diagnosis of these patients and prevents unnecessary surgeries.

Declaration of patient consent

The authors confirm that they have received the patient consent form. On the form, the legal representative gave permission for the patient's images and other clinical information to be reported in the journal. The representative declares that the patient's name will not be published and that every effort will be made to conceal the identity information, but anonymity cannot be guaranteed.

Authors' contributions

SG: Study conception and design, acquisition of data, analysis and interpretation of data, drafting of manuscript, reviewing the paper, advices, and final approval; EK: Aquisition of data, drafting of manuscript, reviewing the paper, advices, and final approval; PK: Aquisition of data, drafting of manuscript, reviewing the paper, advices, and final approval, SE: Aquisition of data, drafting of manuscript, reviewing the paper, advices, and final approval.

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Conflicts of interest

There are no conflicts of interest.

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