

Case Report

B-Cell Lymphoblastic Lymphoma in Relapse Presenting as a Uterine Mass: A Case Report and Review of Literature

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Keywords

Lymphoblastic lymphoma · Leiomyosarcoma · Uterine mass

Abstract

B-cell lymphoblastic lymphoma (LBL) is a highly aggressive malignant proliferation of lymphoblasts of B-origin grouped with acute lymphoblastic leukemia. Multiple studies demonstrated the various sites of involvement in adult LBL. The involvement of the uterus as a site of relapse for such disease is rare. We herein report the case of relapsed B-cell LBL mimicking endometrial sarcoma. The patient is a 56-year-old female patient known to have B-cell LBL on chemotherapy. She presented with abdominal pain and fever. Positron emission tomography-computed tomography showed the presence of a uterine mass with bilateral iliac lymph node involvement. She underwent surgery with mass removal and pathology showed relapsed B-cell LBL.

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Introduction

Lymphoblastic leukemias/lymphomas are neoplasms of precursor T cells and B cells or lymphoblasts. A diagnosis of lymphoblastic lymphoma (LBL) is made when the disease is limited to primary nodal and extranodal sites with minimal involvement of bone marrow (<20% lymphoblasts in marrow). Combination chemotherapy is a standard treatment to LBL, but relapse is common. The involvement of the uterus as a site of relapse for such disease is rare. We herein report the case of relapsed B-cell LBL mimicking endometrial sarcoma.

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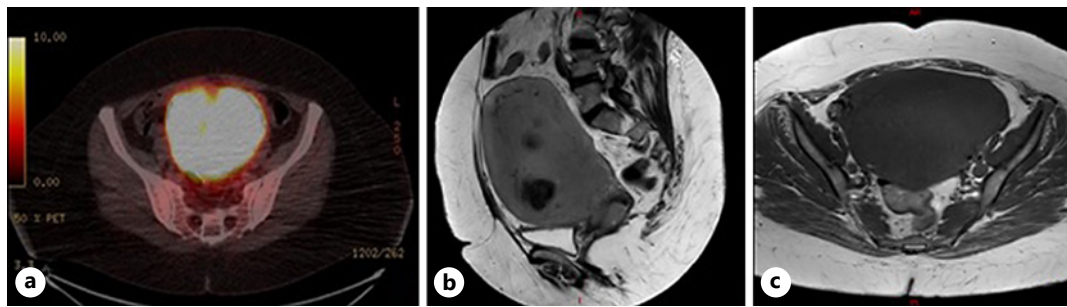


Fig. 1. **a** PET-CT scan showing diffuse increase uptake in the uterus. **b** A sagittal plane of a MRI with gadolinium showing the mass of 15 cm described as possible leiomyosarcoma of the uterine cavity and a second mass in the anterior aspect of the cervix measuring 3.8 cm with 2 suspicious lymph nodes of 1.3 and 1.2 cm noted in the right and left iliac chains respectively. **c** A transverse plane showing dense adherence of the mass to the sigmoid. PET, positron emission tomography-computed tomography.

Case Presentation

It is a case of a 56-year-old lady, Gravida 3 Para 3, in menopause, not on hormonal replacement therapy who presented with bilateral flank pain, constipation, and obstipation. The patient has a history of B-cell acute LBL initially diagnosed 1 year ago on a clavicular lesion biopsy after she presented with a history of osteitis resistant to antibiotics. Bone marrow at diagnosis showed no involvement by lymphoma. After 2 cycles of R-Hyper CVAD (rituximab, cyclophosphamide, vincristine, adriamycin, and dexamethasone) a positron emission tomography (PET) scan was performed and showed complete remission. After receiving a total of 8 cycles of chemotherapy, a control PET scan showed an increase in the size of the uterus measuring 12.5 cm in the medial-lateral aspect with increased FDG uptake of the uterus and the cervix (shown in Fig. 1a). A bone marrow exam was performed and showed 50–60% cellular marrow with maturing trilineage hematopoiesis and <1% blasts. At this moment, the patient was asymptomatic and the physical examination revealed a palpable pelvic mass with a non-tender abdomen. She has no vaginal bleeding or discharge. She has no family history of gynecologic cancer. Pap smear was normal. MRI with gadolinium was done for better characterization of the mass and showed further increase in the size of the uterine mass in 1 month reaching 15 cm described as possible leiomyosarcoma of the uterine cavity. A second mass was noted in the anterior aspect of the cervix measuring 3.8 cm with 2 suspicious lymph nodes of 1.3 and 1.2 cm noted in the right and left iliac chains, respectively (shown in Fig. 1b, c).

After 1 month, she presented to the emergency room with acute and persistent flank pain and obstipation for 24 h, a computed tomography scanner of the abdomen and pelvis with intravenous contrast was performed and showed the same findings of a heterogeneous uterine mass, an enlarged uterus compressing the sigmoid colon associated with colonic dilatation. There was evidence of bilateral hydronephrosis with large bowel obstruction. Multiple bilateral enlarged lymph nodes were noted in the external iliac chain, the largest measuring 1.8 × 1.6 cm.

Taking into consideration the aspect of the mass seen on MRI described as possible leiomyosarcoma and the patient's conditions, she was admitted for surgical removal. Bilateral double J was inserted, and then she underwent an emergent exploratory laparotomy: a 15 cm uterine mass was adherent to the sigmoid. Total abdominal hysterectomy with bilateral salpingo-oophorectomy with sigmoidectomy and a colorectal anastomosis (posterior pelvic

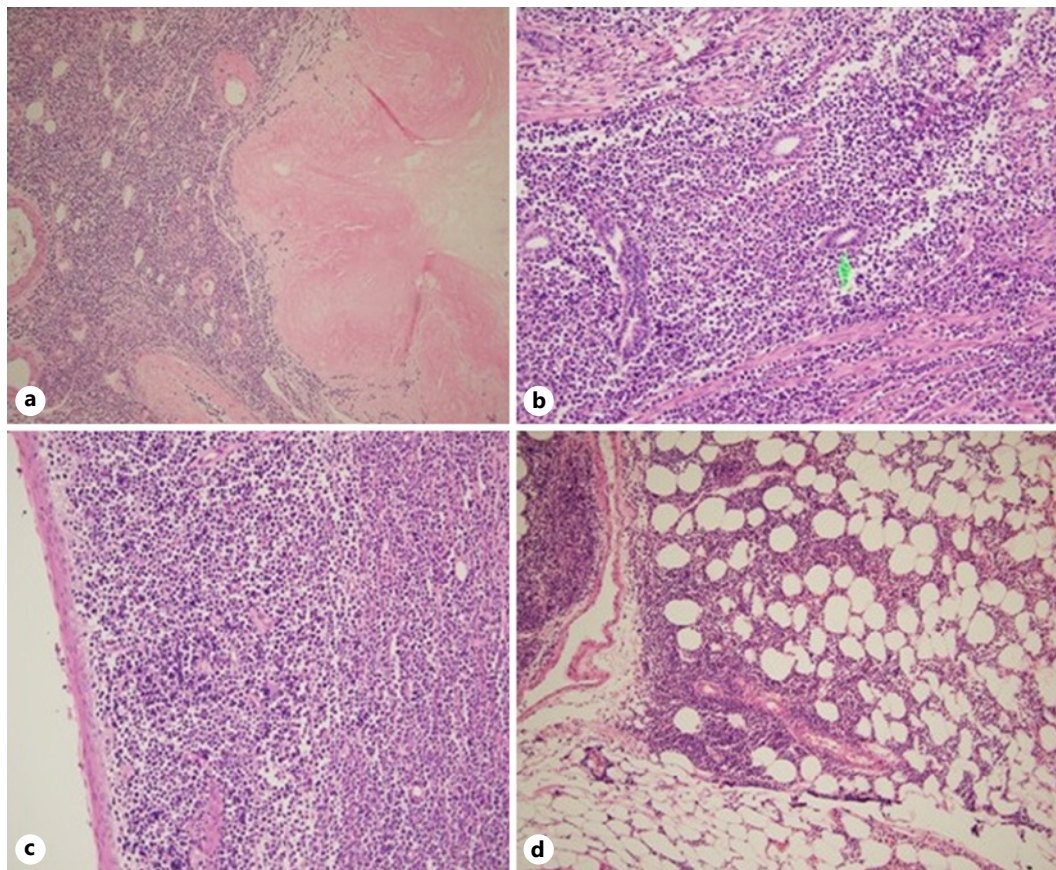


Fig. 2. Hematoxylin-eosin stain microscopy of the neoplastic lymphocytic proliferation. **a** Section from the left ovary exhibiting an infiltrated corpus albicans ($\times 10$ magnification). **b** Endometrial gland diffusely infiltrated by neoplastic lymphocytes ($\times 20$ magnification). Infiltration of the cervix ($\times 20$ magnification) (**c**) and of the mesocolon ($\times 10$ magnification) (**d**).

exenteration) was done en bloc. The specimen was sent for frozen evaluation and showed a relapsed B-cell LBL in the uterine. Based on this result, pelvic lymphadenectomy was not performed as it was not indicated.

Surgical pathology reported a diffuse infiltration of the uterus, the cervix, the ovaries, the fallopian tubes, the parametrium, and the mesocolon by neoplastic lymphocytes disclosing hyperchromatic irregular nuclei. These neoplastic cells infiltrated massively most of the leiomyomas. Immunohistochemistry showed that the neoplastic lymphocytes were diffusely positive for CD45, CD79a, PAX5, Bcl2, and TdT, while negative for cytokeratin C, Desmin, CD3, and CD20. The diagnosis of B cell (CD20 negative) infiltrating the female genital tract was rendered (shown in Fig. 2, 3). The postoperative period was without complications and the patient was discharged on day 8. She is planned to receive gemcitabine and oxaliplatin chemotherapy for a total of 6 cycles.

The authors state that they have followed the principles outlined in the Declaration of Helsinki for all human or animal experimental investigations. In addition, written informed consent was obtained from the patient for publication of this case report and any accompanying images.

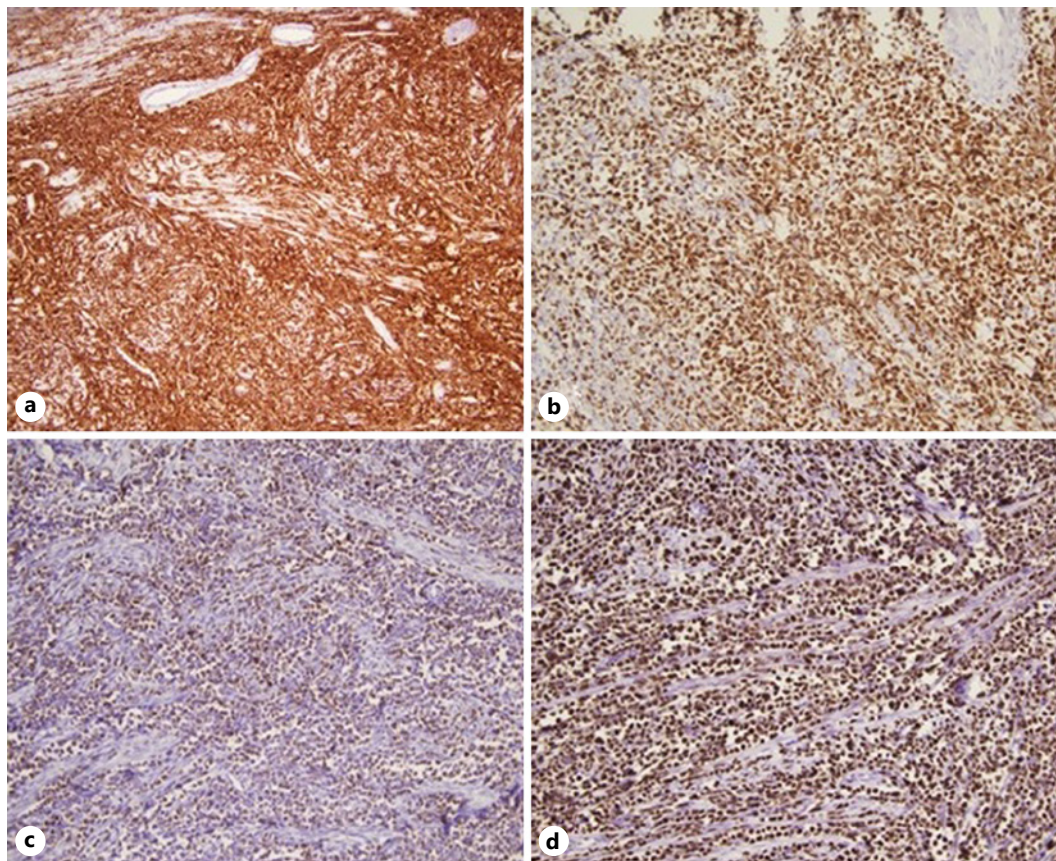


Fig. 3. Immunohistochemistry of the B-cell LBL. The neoplastic cells are positive for CD45 (a), TdT (b), CD79a (c), and PAX5 (d). LBL, lymphoblastic lymphoma.

Discussion

In this case report, we report a case of B-cell LBL involving the uterus presenting similarly to locally advanced endometrial leiomyosarcoma. Multiple studies have demonstrated the distribution of extramedullary sites involved among patients with LBL. These sites were mainly the pleura, pericardium, central nervous system, liver and spleen, and to a lesser extent the skin and kidneys [1]. However, the involvement of the female genital tract is rare and accounts for <1% of extranodal lymphoma and/or leukemia [2].

Patients with hematological malignancies involving the genital tract usually present with symptoms of vaginal bleeding, abdominal pain due to a pelvic mass along with other constitutional symptoms such as B symptoms (weight loss, night sweats, and fever). Our patient presented with nonspecific diffuse abdominal pain and obstipation without any other signs or symptoms with imaging showing a uterine mass.

The appearance of the mass on imaging favored leiomyosarcoma (LMS). LMS is usually solitary, heterogeneous, and poorly demarcated masses on MRI [3]. They frequently show areas of high intensity on T1-weighted images corresponding to necrosis. T2-weighted images show intermediate to high signal. After intravenous gadolinium contrast, LMS demonstrates early and heterogeneous enhancement [3–5]. PET scan is sensitive to detect tumors with a high rate of proliferation. It is sensitive for staging high-grade non-Hodgkin lymphoma [6]. However, in the setting of a single uterine mass, PET scan is not sensitive to differentiate between LBL and LMS as both entities will be highly FDG avid. Taking into consideration the

history of the patient and despite the rare involvement of the genital tract by hematological malignancies and the typical features of leiomyosarcoma on MRI, one should always suspect a relapse of the primary disease.

In this patient, our decision to perform surgery was based on the high specificity and diagnostic accuracy of the MRI in detecting uterine sarcomas [7]. In this situation, according to the national comprehensive cancer network NCCN guidelines, preoperative imaging and biopsy may help to identify uterine sarcomas although biopsy sensitivity is less than for endometrial cancer. The suspicion of malignant mesenchymal sarcoma is a contraindication of fragmentation and morcellation. Hence, a mass biopsy was not done during the laparotomy, and a surgical removal en bloc was performed. Pathological findings of the surgical specimen showed relapse B-LBL. Histopathological features of LBL show usually a diffuse proliferation of lymphoblasts comparable to acute lymphoblastic leukemia cells [8]. Immunohistochemistry will confirm the B-cell origin.

In summary, B-cell LBL is aggressive hematological neoplasms and extramedullary relapses of the disease are frequent. When the patient is symptomatic, imaging is necessary as relapses may occur at multiple sites without bone marrow involvement. Interpretation of radiologic findings should always be correlated to the clinical presentation as these findings alone are nonspecific. In the context of known hematological malignancy namely LBL, any soft tissue mass can represent infection, secondary neoplasms, or relapse of primary disease [9].

Statement of Ethics

The authors state that they have followed the principles outlined in the Declaration of Helsinki for all human or animal experimental investigations. In addition, written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

All the authors have contributed equally in the literature search and redaction of this article.

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