

Case Report**Pelvic lymphoma: An unusual presentation***Fakhrolmolouk Yassaee<sup>1</sup>, Sedighe Hosseini<sup>2</sup>***Abstract**

Pelvic lymphoma is not a common condition and aggressive recurrence of chronic lymphocytic leukemia (CLL) as a cause is rarely reported. We report a case of lymphoma of left adnexa in a postmenopausal woman, with a three week history of abdominal and left flank pain. Past medical history was consistent with diagnosis of CLL. She had received chemotherapy. Due to abdominal and flank pain and abdominopelvic mass, exploratory laparotomy was done with the impression of ovarian malignancy. A large uterus with adnexal mass and a large tumoral bladder was seen. Biopsy was done from adnexal mass which was compatible with lymphoma. The abdomen was closed and the patient was referred for chemotherapy. Lymphoma usually does not involve the pelvic organs. After laparotomy, her condition deteriorated and she expired.

**KEYWORDS:** Chronic Lymphocytic Leukemia, Aggressive Recurrence, Abdominopelvic Mass.

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**C**hronic lymphocytic leukemia (CLL) is known as a disease occurring after 50 years of age and is twice more common in men than in women.<sup>1</sup> Chronic lymphocytic leukemia (CLL) is characterized by the accumulation of mature -appearing lymphocytes in blood, marrow, lymph nodes, and spleen. The CLL cells are monoclonal B lymphocytes that express CD19, CD5, and CD23, with weak or no expression of surface immunoglobulin (ig), CD20, CD79b, and FMC7.<sup>2</sup> It is the most common adult leukemia, accounting for 24% of all leukemias. The median survival of patients with CLL ranges from 5 to 10 years, but a trend to a longer survival has become evident in recent years, partially due to earlier diagnosis.

Infections and secondary cancers continue to be the principal causes of death in these patients. The rationale for vaccine strategies is to induce host cell-mediated immune responses against autologous malignant cells as a modality to eliminate tumor and provide lasting protection from recurrence.<sup>3</sup> The following report

lights up a case with aggressive recurrent lymphoma.

**Case presentation**

A 67 year old grand multiparous woman was presented to Taleghani Hospital, affiliated to Shaheed Beheshti University of Medical Science, Tehran, Iran, with three week history of abdominal and left flank pain. In the past medical history, she had been diagnosed and treated for chronic lymphocytic leukemia 8 years ago and was currently on chlorambucil and folic acid.

In the preceding month, she developed decreased urine output (oliguria). CT scan showed bilateral hydronephrosis for which nephrostomy was done. In the CT scan, there was a thick and irregular bladder wall, large uterus and a left adnexal mass.

Clinical examination revealed an elderly woman. Her vital signs were normal. A poorly defined mass arose from the pelvis and was compatible with a pregnancy of about 20 week

1- Associate Professor, Department of Obstetrics and Gynecology, School of Medicine, Shaheed Beheshti University of Medical Sciences, Tehran, Iran.

2- Resident, Department of Obstetrics and Gynecology, Shaheed Beheshti Medical Sciences University, Tehran, Iran.

Corresponding Author: Fakhrolmolouk Yassaee

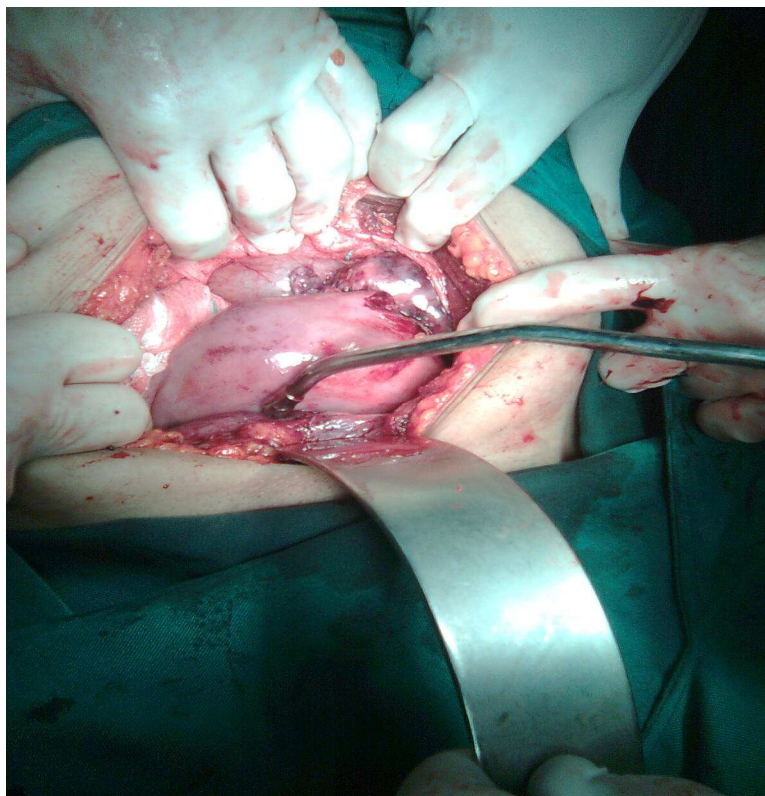
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duration. Sonography showed a hypoechoic mass of 125×86×52 mm in the urinary bladder and a mass of 80×46×30 mm in left adnexa, which was suspected to be ovarian in origin. Three days after admission, she developed left lower extremity edema. Color Doppler sonography showed deep veins thrombosis, and heparin was started. Blood tests revealed a serum calcium of 7.8 mg/dl (normal range: 8.4-10.4 mg/dl), LDH of 1157 mg/dl (normal range up to 248 mg/dl), uric acid of 10.7 mg/dl (normal range of 2.6-6 mg/dl), and ESR of 60 (normal range up to 25). The provisional diagnosis was metastatic ovarian malignancy. So, laparotomy was done to get a biopsy. At operation, through a midline sub-umbilical incision, about 1 liter ascitic fluid was aspirated and sent for cytology. The uterus was large. A large mass of about 8 cm was present in the left adnexa. Bladder wall was thick with a mass of about 8 cm on the wall (Figure 1). Frozen sections from left adnexa revealed lymphoma. So, abdominal wall was closed.

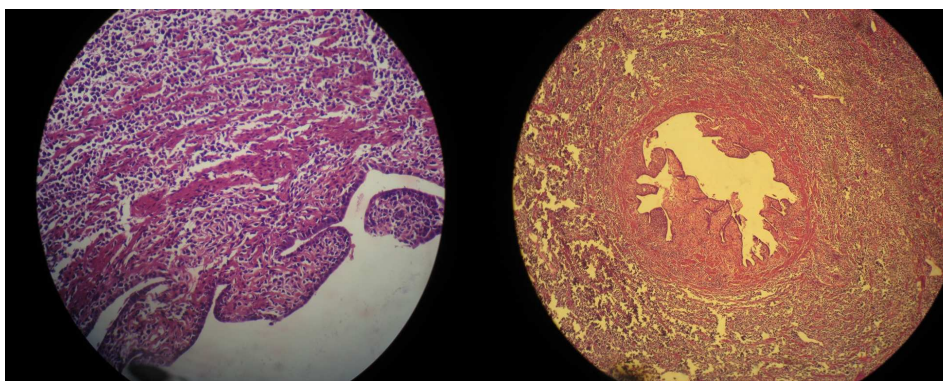
The result of cytology showed monomorphic lymphoid cells, positive for malignancy. Histopathology report revealed tubular fragments of fallopian tube showing presence of malignant neoplasm composed of diffuse infiltration of discohesive cells with hyperchromatic nuclei infiltrating the muscular fibers of fallopian tube wall, suggestive of lymphoma (Figure 2). Immunohistochemically, the cells were positive for LCA and negative for CK, which confirmed the diagnosis of lymphoma. Patient's condition was deteriorated, she became oliguric and septic and died.

### Discussion

The clinical course of chronic lymphocytic leukemia is highly variable.<sup>4</sup> Following diagnosis, some patients have smoldering, asymptomatic disease that may not progress for many years. Others are diagnosed with advanced disease, or early stage disease that rapidly progresses, causes symptoms or bone marrow failure and requires treatment.<sup>3</sup>



**Figure 1.** Macroscopic image of mass.



**Figure 2.** Section of fallopian tube suggestive of lymphoma.

Involvement of other organs in CLL is very rare at first. Some reports described unusual presentations as a first presentation such as hemorrhagic pleural effusion and penile mass with skin lesion.<sup>5,6</sup> Another report by Joulian stated that the involvement of the ovary in lymphomatous process is rare, but the ovary is the common site in the female genital tract to be involved by the hematological malignancies. Involvement of the ovary by malignant lymphoma can be primary or secondary and is discovered incidentally during a workup for abdominal or pelvic complaints. The occurrence of lymphomas primarily arising in the ovaries has long been debated since no lymphoid tissue is found in the ovaries.<sup>7</sup>

A Richter transformation occurs when CLL transforms into another lymphoid malignancy such as polymorphocytic leukemia, diffuse large cell lymphoma, Hodgkin disease, acute leukemia, and multiple myeloma. Richter syndrome refers to the development of a diffuse large B cell lymphoma or its immunoblastic variant in a patient with CLL. The syndrome occurs in 5% of patients, can occur at any time during the disease course, and is characterized by increasing lymphadenopathy, splenomegaly, fever, and weight loss. In addition, these patients frequently have extranodal involvement, with infiltration of kidneys, lungs, and the gastrointestinal tract; most patients die within 1 year. Eighty percent of patients have an increase in LDH. In this patient, LDH increased significantly. It is thus important to obtain a histologic diagnosis before considering therapy.<sup>2</sup>

Primary malignant lymphoma of the female genital tract is an extremely rare clinical presentation and only case reports have described this condition. Some of them present with vaginal bleeding and pelvic mass.<sup>8</sup>

Malignant lymphoma of uterine cervix can be clinically and histopathologically misdiagnosed for the infrequent presentation in this area.<sup>9</sup> Appropriate diagnosis of lymphomas of cervix and uterus is often delayed until the postoperative setting, as clinical and radiographic presentations are nonspecific.<sup>10</sup>

The focus in oncology continues to shift toward therapy based on a clear understanding of genetic abnormalities and their connection with prognosis and response to therapy. Chronic lymphocytic leukemia is a prime example of a malignancy benefiting from not only important new treatment options, also the identification and clarification of numerous prognostic markers.<sup>11-14</sup> Our patient had a good course of disease for 8 years but after that she developed aggressive recurrence of CLL. She presented with abdominal mass and its obstructive effect on pelvocalyceal system, oliguria and deep vein thrombosis. Laparotomy and biopsy revealed a lymphoma.

## Conclusion

Management of lymphoma is medical. This report should once again serve obstetricians and gynecologists to raise their awareness. Although pelvic involvement of lymphoma is a rare phenomenon, it should be considered when there is a pelvic mass in a patient with previous diagnosis of CLL.

## Conflict of Interests

Authors have no conflict of interests.

## Authors' Contributions

FY complete paper is written. SH helped for searching references.

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