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Primary Uterine Cervix B-Cell Lymphoma: A Case Report and Treatment Debates

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ABSTRACT

Non-Hodgkin lymphomas are a heterogeneous group of lymphoproliferative disorders with various behaviors and responses to treatment. As a primary extranodal NHL, the disease must be confined to one location, and bone marrow should not be involved. Primary uterine cervix lymphoma is a rare malignancy as well as a rare site of extranodal lymphoma. Because of the rarity of the disease, there is no standard treatment guideline for women with primary uterine cervix lymphoma. Patients mostly present with abnormal uterine bleeding, vaginal discharge or pelvic pain. Typically, a pap smear may not show the malignant cells in the specimen. When the diagnosis is made, management may be debated due to its rarity and lack of standard treatment. Surgery, chemotherapy, chemo-immunotherapy and radiotherapy, either alone or in combination, are the treatment options. Most patients respond well to chemotherapy and radiation therapy. The prognosis is usually favorable. Here, we report a case of primary cervical lymphoma in a 50-year-old woman who presented with abnormal vaginal bleeding. She was treated with radiotherapy and chemo-immunotherapy and remained disease-free after nine months of treatment.

Keywords: Extranodal Lymphoma, Uterine Cervix Lymphoma, Chemotherapy, Radiotherapy

Introduction

Primary cervical lymphoma is a rare malignancy. It is estimated that less than 1 % of extranodal lymphoma cases appear in the uterine cervix **(1)**. Diagnosis is challenging as the pap test often fails to detect the pathologic cells **(2)**, and it could be easily misdiagnosed as squamous cell carcinoma **(3)**. Treatment is debatable because of its rarity. A high rate of treatment success has been achieved with a combination of surgery, chemotherapy, chemoimmunotherapy and radiotherapy **(4)**. Here, we report a case of primary uterine cervix lymphoma in a 50-year-old woman who presented with abnormal vaginal bleeding.

Case Presentation

The patient is a 50-year-old woman presenting with abnormal vaginal bleeding since one month ago, whose bleeding was becoming worse and passing big clots during the last week of presentation. She went to a gynecologist for further evaluation. In a physical examination, the cervix was enlarged, and a lobulated mass was seen on the cervix.

The patient was referred to a gynecologic oncologist for colposcopy and exam under anesthesia, cervical biopsy and dilation and curettage.

Transvaginal sonography showed an enlarged uterus of 114*54*53 mm with a normal endometrial thickness of 5 mm. A solid hypoechoic mass containing vascularity measuring 68*30 mm was noted at the anterior part of the cervix, extending to the fornix with the differential diagnosis of cervical tumor (carcinoma or myoma) **(5)**. For more evaluation, an MRI was recommended **(6)**. MRI of the abdomen and pelvis with and without contrast showed a lobulated border soft tissue mass with heterogeneous enhancement involving the posterior vaginal fornix, invasion to the posterior wall of the urinary bladder, and stranding in the right parametrial fat, suggesting at least SIIb cervical cancer with measures of 55 mm in sup-inf. diameter (Figure 1).

No pelvic adenopathy or extrapelvic pathologic findings were reported.

A tumor was biopsied. Pathology **(7)** showed a high-grade, undifferentiated tumor (Figure 2). Immunohistochemistry (IHC) is important in the differential diagnosis of uterine cervix tumors. IHC evidence was in favor of large B-cell lymphoma, germinal center type, CD20 positive, CD3 positive in small lymphocytes, CD10 positive, PAX-5 positive, MLM-1 negative, BCL-6 negative, CMYC negative, BCL2 negative, and Ki67 50% **(7)**.

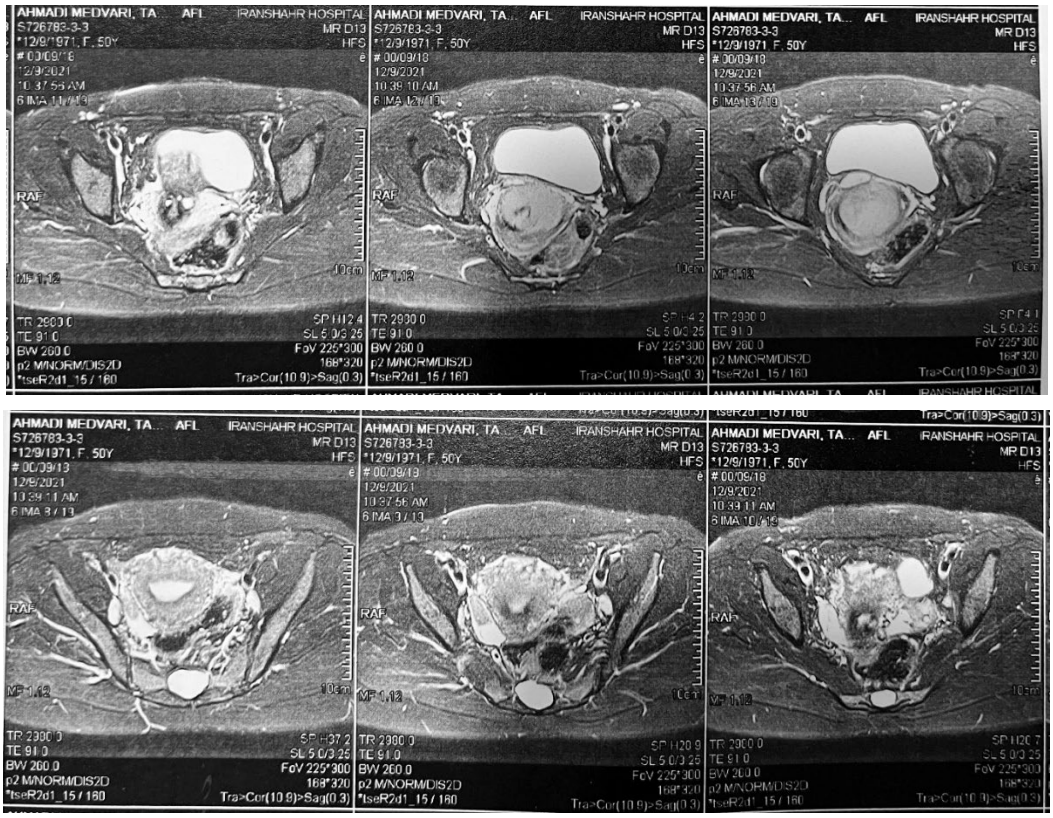

Figure 1. **Pelvic MRI showed an enlarged uterus**

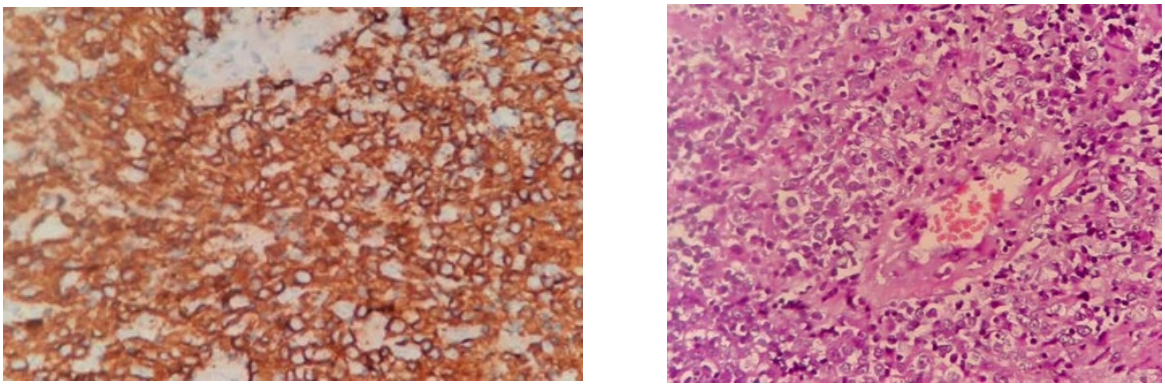

Figure 2. **Pathology**

As a staging workup and to detect other involved sites, FDG-PETCT was done. The results of PET CT showed an FDG avid soft tissue mass within the uterine cervix compatible with the patient's biopsy, a few faintly avid pelvic lymph nodes suspicious for lymphomatous involvement, and no pathologic enlarged or FDG avid lymph node nor metabolic evidence of lymphoma in the remainder of the imaged portion of the body. Finally, the patient was diagnosed with stage I extranodal primary cervical lymphoma **(8)** (Figure 3 and 4).

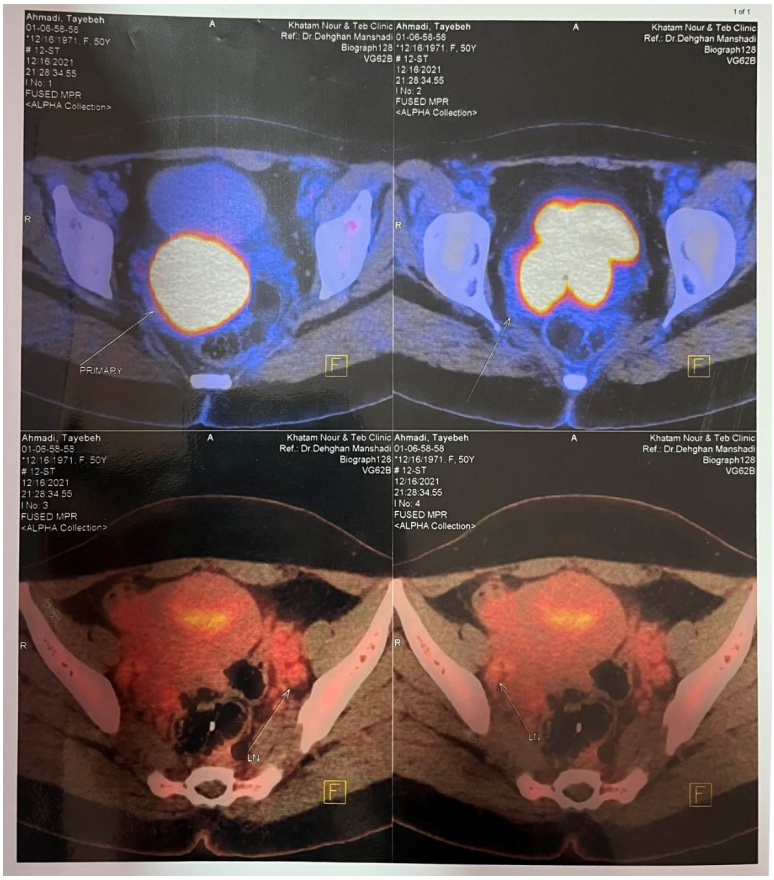

Figure 3. **The results of PET CT**

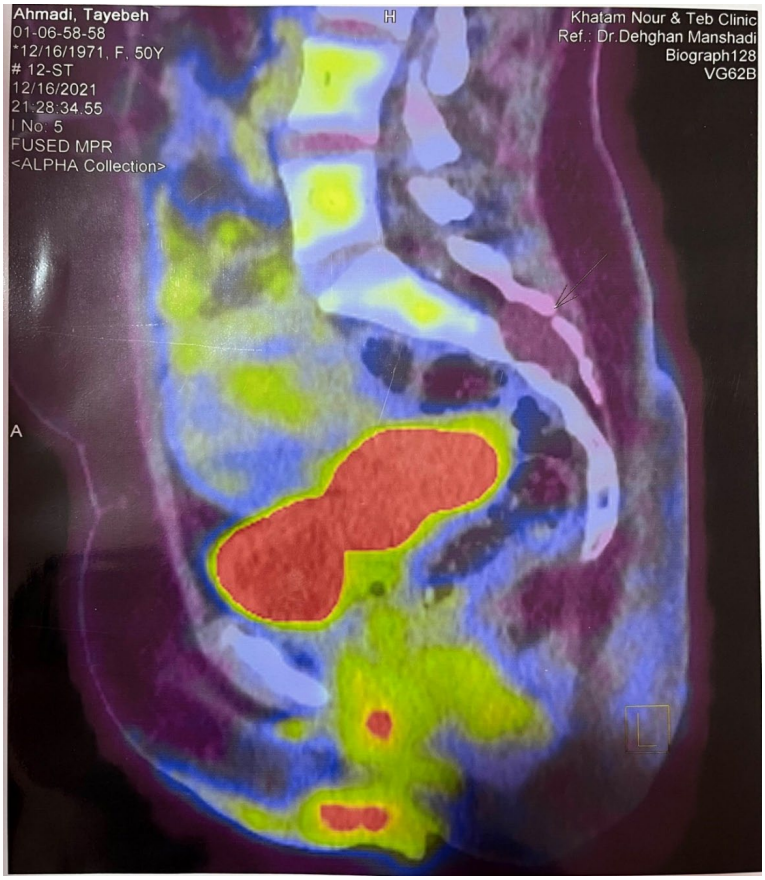

Figure 4. **The results of PET CT**

Treatment began with whole pelvis irradiation of 50 Gy and was continued with six cycles of immune-chemotherapy R-chopp (Rituximab, Cyclophosphamide, Adriamycin, Vincristine and Prednisone) every 3 weeks. She didn't have surgery in her treatment protocol.

At the end of treatment, she went on a surveillance protocol with a physical exam and MRI every 3 months following treatment **(7)**.

An MRI was done 3 months following the end of radiotherapy and showed a normal-sized uterus (80 mm), a thin endometrium (1 mm), normal myometrial signal intensity, mild thickness on the right side of the vaginal fornixes and vaginal wall, as well as mild soft tissue fullness in the fat plane in the night posterior aspect of the urinary bladder, and no pelvic side wall adenopathy. The physical exam was normal, with no tumor seen or palpated on the cervix **(8)**. The following MRI six months later didn't show any pathologic findings (Figure 5).

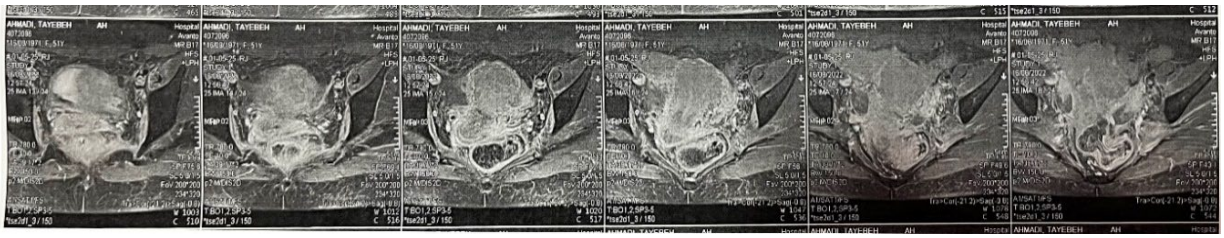

Figure 5. MRI post chemotherapy and radiotherapy

Discussion

Non-Hodgkin lymphomas are a heterogeneous group of lymphoproliferative disorders with various behaviors and responses to treatment. As a primary extranodal NHL the disease must be confined to one location, and bone marrow should not be involved. One-third of lymphomas are extranodal in origin **(9)**.

Primary uterine cervix lymphoma is rare, and less than 1% of extranodal lymphoma originates in the female genital organs.

The median age at presentation is in the fourth or fifth decade of life, although it can present at 20-80 years of age. The presenting symptom is usually an irregular bleeding, vaginal discharge or dyspareunia. Classic lymphoma signs such as fatigue, night sweats and weight loss are not usually seen.

Differential diagnosis includes poorly differentiated squamous cell carcinoma, sarcoma and chronic inflammation. When a diagnosis is made, PET-CT is helpful as a staging tool and is also used to identify additional information about the disease such as bone marrow and visceral involvement **(10)**.

The Ann Arbor staging and the FIGO are used for staging. In women diagnosed with primary cervical lymphoma, staging is the best predictor of survival, and if diagnosed at early stages, the prognosis is excellent.

The treatment approach is not standard due to its rarity. However, complete remission is reported with a combination of chemotherapy and radiation. In most cases reported by other authors, surgery was not chosen for treatment. Commonly, R-chopp is the chemotherapy protocol used.

Post-treatment surveillance is not clearly defined. Physical and pathologic examinations, as well as MRI, are reasonable options.

Conclusion

In conclusion, we described a case of primary uterine cervix lymphoma and discussed the clinical and pathologic characteristics as well as the treatment options for these patients. Based on the literature, most of these patients present at an early stage, respond well to treatment, and have long-term survival.

Acknowledgments

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None.

Conflict of Interest

The authors declare that they have no conflicts of interest.

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