

Primary diffuse large B cell lymphoma of the cervix presenting as bilateral hydronephrosis

Joel Alcid, Christian Fidler

ABSTRACT

Introduction: Non-Hodgkin lymphoma (NHL) arises most frequently in lymph nodes or other lymphatic tissue. Primary lymphomas of the female reproductive tract are extremely rare, less than 1% of extra nodal NHL and <0.5% of gynecologic malignancies. **Case Report:** We present a case of a 66-year-old postmenopausal female presented with a one month history of fatigue, dysuria and hematuria. A non-contrast computed tomography scan of abdomen and pelvis revealed a large ~7–9 cm mass involving the lower uterine segment/cervix displacing the rectosigmoid colon. Biopsy revealed it to be a CD 20 positive diffuse large B cell type non-Hodgkin lymphoma (NHL). She was diagnosed as stage IE and started on R-CHOP chemotherapy (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone). **Conclusion:** Primary diffuse large B cell lymphoma of the female genital tract is a rare diagnosis and must be diagnosed early due to the aggressiveness of the tumor. There is no standard treatment regimen. Patients are often treated with chemotherapy as the first line of treatment, which has provided promising results.

Keywords: Cervix, Diffuse large B cell lymphoma, Non-Hodgkin's lymphoma, Uterus

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INTRODUCTION

Non-Hodgkin lymphoma (NHL) arises most frequently in lymph nodes or other lymphatic tissue. Primary extra nodal lymphoma accounts for one-third of cases. The most commonly affected extra nodal sites are the gastrointestinal tract and skin, but may affect any organ. The most common extra nodal lymphoid neoplasm is the diffuse large B cell lymphoma, which comprises 25% of NHL. Primary lymphomas of the female reproductive tract are extremely rare, less than 1% of extra nodal NHL and <0.5% of gynecologic malignancies. The cervix is the most commonly affected pelvic site, but may also occur in the ovary, uterus, vagina, vulva, and regional lymph nodes. We present a case of primary diffuse large B cell lymphoma involving the cervix and uterus [1].

CASE REPORT

A 66-year-old postmenopausal woman presented with a one month history of fatigue, dysuria and hematuria. She received multiple courses of antibiotics as an outpatient without improvement. On admission labs, she was found to have a creatinine of 6.92 (baseline ~1.2). A retroperitoneal ultrasound was obtained and revealed

bilateral moderate to severe hydronephrosis. A non-contrast CT abdomen and pelvis revealed a large ~7–9 cm mass involving the lower uterine segment/cervix displacing the rectosigmoid colon, likely representing the site of obstruction (Figure 1–3). An enlarged multi-fibroid uterus was also noted. There was no hepatosplenomegaly or adenopathy. On pelvic examination, a diffuse thickening was noted between the vagina and bladder. There was a firm mass wrapping around the cervix with thickening of the rectovaginal wall. She underwent further examination under anesthesia and multiple biopsies of the cervical mass were obtained. Patient also had bilateral percutaneous nephrostomy tubes placed to treat the obstruction.

Histological examination of the biopsy specimen revealed diffuse large B cell lymphoma (DLBCL). Immunohistochemical staining tested positive for CD 20, MYC, CD 10, and BCL 6. Staging workup with bone marrow aspirate and biopsy was negative for any monoclonal B cell or T cell population. The PET scan revealed mild metabolic activity at the lower uterine segment/cervix and upper vagina. Findings would correspond to a Deauville score of 3–4. Viral serologies for HIV, hepatitis B surface antigen, and hepatitis C were negative. After complete diagnostic workup, she was diagnosed with diffuse large B cell lymphoma stage IE. Patient was started on R-CHOP chemotherapy (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone). After six cycles of chemotherapy, a PET scan for restaging was obtained and revealed a significant decrease in SUV. However, only a minimal decrease in tumor size was visualized. A ~7–8 cm bulky mass remains, and consolidation radiation therapy is being considered.

DISCUSSION

Diffuse large B cell lymphoma is the most common extranodal NHL neoplasm (30–58%). Median age of onset of patients affected by this neoplasm is 40-year-old. Primary lymphoma of the female reproductive tract is extremely rare. Clinical symptoms are usually non-specific and can include vaginal bleeding (70%), bilateral hydronephrosis, perineal discomfort (40%), persistent vaginal discharge (20%), abdominal pain, however, some patients are asymptomatic. The classic B symptoms of lymphoma (fever, night sweats, and weight loss) are rarely seen. Our patient presented outside the median age range, but had the corroborative symptoms of hydronephrosis. Few cases have been reported of DLBCL of the cervix presenting with hydronephrosis [2].

Diagnosing lymphoma of the cervix or uterus can present challenges. A Papanicolaou smear has limited diagnostic value and is usually negative, because these tumors infiltrate the cervical stroma, and the squamous and glandular lining is initially preserved. Deep cervical biopsies are needed along with histopathologic

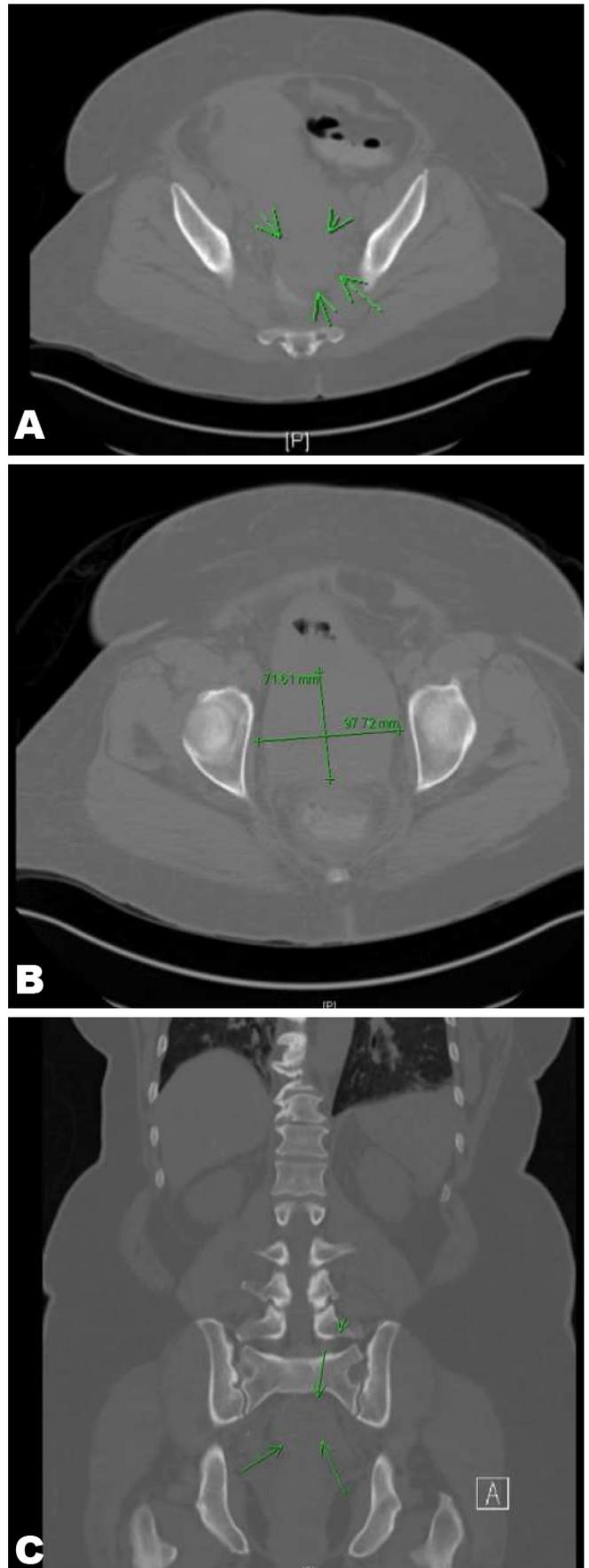


Figure 1: (A–C) Computed tomography scan of abdomen and pelvis. A 7x9 cm mass involving the lower uterine segment and cervix.

evaluation for definitive diagnosis. In a study of women with primary cervical lymphoma, 41% had an abnormal cervical cytology [3, 4].

The study of choice for detection and staging of DLBCL is computed tomography scans. Also included in the workup for staging are bone marrow biopsy and positron emission tomography scan (PET). It is important to distinguish the etiology of the tumor, adenocarcinoma versus squamous cell carcinoma vs lymphoma, as these tumors have different mainstays of treatment. Surgery and radiation are used for adenocarcinoma and squamous cell carcinoma, and chemotherapy/immunotherapy is used for lymphoma [5].

Another important aspect in the diagnosis of lymphoma is immunohistochemical staining, as this can determine the type and subtype of lymphoma. Expression of CD 20, CD 10, BCL 6, and MUM 1 negative are suggestive of germinal center B type lymphoma (DLBCL). Our patient expressed CD 20, CD 10 and BCL 6, consistent with DLBCL [6].

Due to the rarity of this malignancy, optimal treatment has never been standardized and therapy is still under discussion. DLBCL of the cervix/uterus has been managed with chemotherapy, radiation therapy and surgery. The most commonly used and preferred regimen is R-CHOP. With this therapy, it has been reported that up to 70-80% of patients achieve complete remission with six cycles of R-CHOP. Other regimens that have been used are MACOPB (methotrexate, cytarabine, cyclophosphamide, vincristine, prednisolone, and bleomycin, CHOP-bleo (cyclophosphamide, doxorubicin, vincristine, prednisolone, and bleomycin), and ASAP (doxorubicin, methylprednisolone, cytarabine, and cisplatin). For large bulky tumors (>10 cm), residual lesions, or tumors with incomplete response, radiation therapy can be considered. According to a review of cases by Anagnostopoulos et. al. 118 cases were reviewed, and revealed 16.8% had chemotherapy only, 10.9% had radiation therapy only, and 9.2% had surgery only and the remaining patients had multiple modalities of treatment [7, 8].

The overall survival rate of for patients for DLBCL of the cervix, uterus, and vagina is 89% with a relapse free survival rate of 70% [2, 6, 11].

CONCLUSION

Primary diffuse large B cell lymphoma of the female genital tract is a rare diagnosis and must be diagnosed early due to the aggressiveness of the tumor. There is no standard treatment regimen. Patients are often treated with chemotherapy as the first line of treatment, which has provided promising results.

Author Contributions

Joel Alcid – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of version to be published

Christian Fidler – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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