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Primary Lymphoma of the Uterine Cervix, Report of a Rare Case

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Abstract Primary lymphoma of the female genital tract, including the uterine cervix, is rare. The clinical picture is nonspecific, which complicates the timely diagnosis. Treatment and prognosis are considerably different in the long list of differential diagnoses, including other malignancies and inflammatory processes. Sometimes multiple biopsies and immunohistochemistry are needed for a definite diagnosis. Here we report a 34-year-old female with a mass on the uterine cervix and the final diagnosis of Diffuse Large B-Cell Lymphoma, Non-Germinal Center Type. The pathologist and clinician must be aware of this entity in daily practice, for appropriate diagnosis and treatment.

Keywords: *uterine cervix, extra-nodal lymphoma, primary*

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1. Introduction

Extra-nodal forms of Non-Hodgkin lymphoma (NHL) represent 25-40% of NHL, where only 2% of them concern the female genital system. They develop in the female genital organs primarily, or may secondarily invade them. Extra-nodal NHL genital form is extremely rare and represents 0.5% of all malignant genital diseases. All genital organs may be affected, where most lymphomas are localized in the cervix, uterine body, and ovary. The clinical picture is non-specific which complicates the timely diagnosis. A multidisciplinary approach is used for staging lymph proliferative disease [1].

Primary lymphoma of the female genital tract including the uterine cervix is rare and confusing, due to non-specific symptoms [2,3]. Exophytic mass of cervix or cervical enlargement with abnormal vaginal bleeding, perineal discomfort, dyspareunia, vaginal discharge, abdominal mass, urinary retention, and fixation of the cervix in the pelvic exam are the clinical presentations, easily shown in squamous cell carcinoma of the cervix, other malignancies and inflammatory process [3,4,5,6]. Treatment and prognosis are considerably different in this long list of differential diagnoses [3]. Routine tests including Papanicolaou smear may not detect the abnormal cells [7]. Sometimes multiple biopsies and immunohistochemistry are needed for a definite diagnosis [3,5]. The most prevalent histological subtypes of primary lymphoma of the female genital tract are diffuse large B-cell and follicular lymphoma. Tumors are mainly located in the ovary, cervix, and uterus [2]. Primary lymphoma of the uterine cervix is mainly seen in premenopausal age and most are non-Hodgkin lymphomas [8].

2. Case Presentation

A 34-year-old female was presented with a mass on the uterine cervix in an ultrasound exam. Multiple ultrasound exams of the uterus showed protrusion of a hypoechoic cystic mass with an irregular posterior margin from the posterior wall of the cervix to the posterior cul de sac. The lesion measured about 31x43x77mm, contained debris and thick internal septa. The lesion was extended to the lateral wall of the cervix and was hypovascular in doppler ultrasound. The hue might be suggestive of endometriosis or degenerated fibroma or neoplasm. Rectal wall thickness was increased by 13mm and the radiologist suggested a rectoscopic exam. Complete blood count, thyroid function tests, prolactin, CEA and CA125, alpha-fetoprotein, and CA19-9 of the serum were within normal limits. MRI of the cervix without contrast showed a non-homogeneous mass in the posterior cervical wall with extension to the posterior cul de sac, measured 40x30x25mm with no invasion to the rectum. The radiologist suggested degenerated subserosal fibroma and recommended a trans-rectal biopsy of the mass. The mass was submitted to the pathology with the clinical diagnosis of uterine

leiomyosarcoma. Immunohistochemistry (IHC) was done and showed positivity for CD45, CD20, CD5, and Ki-67 (30% of tumor cells), but SMA, Desmin, CD68, BCL2, CD10 and CD23 were negative. The pathologist suggested malignant high-grade B- cell lymphoma. A second opinion was requested and a repeat with complementary IHC was done at a referral center. Positive markers were Ki-67 (in 80% of tumor nuclei), LCA, Vimentin (in few tumor cells), CD20, CD19, and BCL6 (in the majority of tumor cells). Negative markers were CK, S100, CD34, SMA, Desmin, CD3, CD5, CD10, BCL2, MUM1, C-myc and Cyclin D1. The pathologist reported: "IHC staining is consistent with Diffuse Large B-Cell Lymphoma (DLBCL), Non-Germinal Center Type" (Figure 1). Further follow-up was not available for the pathologist. Written informed consent was obtained from the patient.

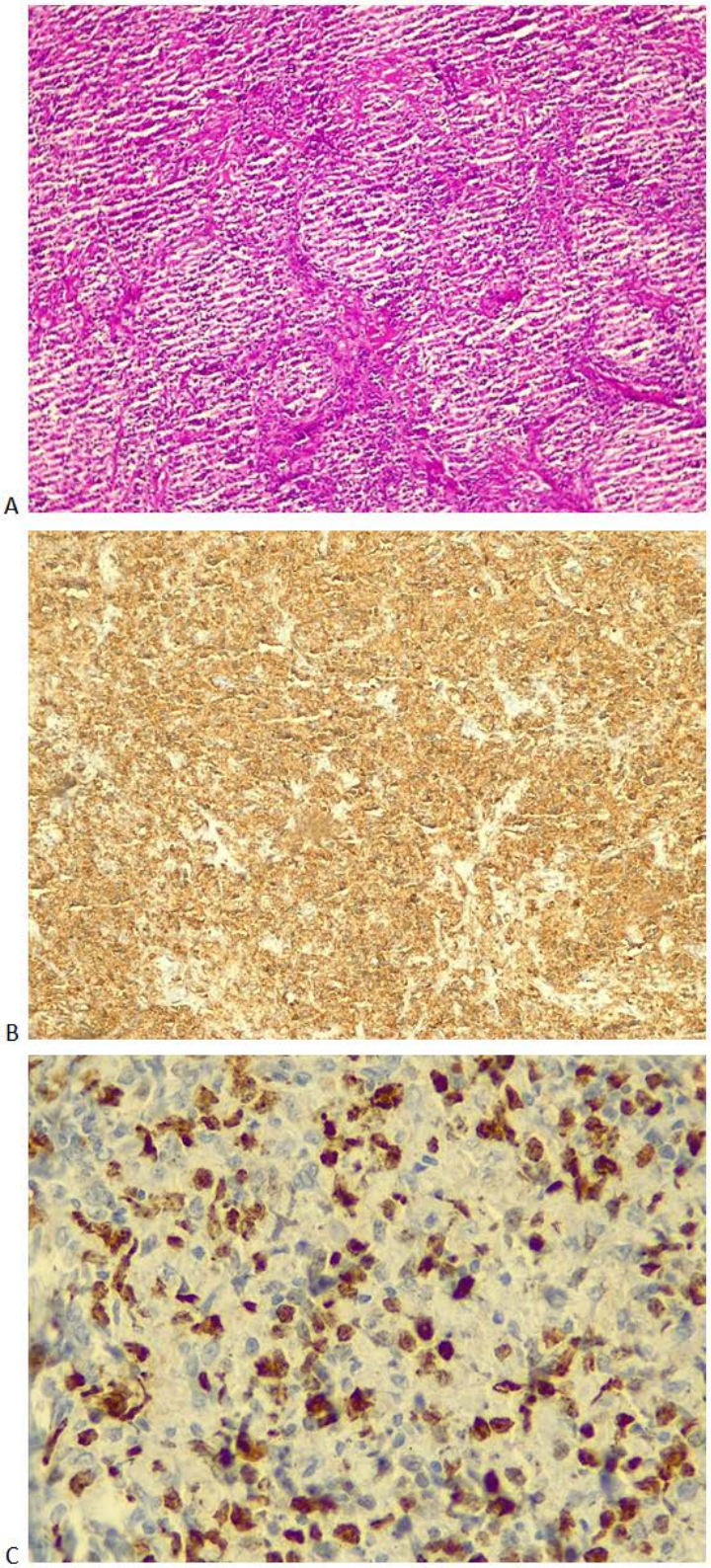

Figure 1. Diffuse Large B-Cell Lymphoma of the uterine cervix. A- Hematoxyline Eosin staining X100 magnification. B- Immunohistochemistry positive CD20 X100 magnification. C- Immunohistochemistry positive Ki-67 X400 magnification

3. Discussion

Primary malignant lymphomas of the genital tract are rare diseases. They account for 1.5-2 % of extra-nodal non-Hodgkin's lymphomas. Only 0.12 % of all non-Hodgkin's lymphomas originate from the uterine cervix. Cervical lymphoma may present in a wide age range (20–80 years); however, 80 % of patients are premenopausal, and cervical lymphoma tends to occur in younger patients compared to other lymphomas. The present case was a 34-year-old woman. The common presenting symptoms are abnormal vaginal bleeding (70 %), perineal discomfort (40 %), persistent vaginal discharge (20 %), and, less frequently, abdominal masses, dyspareunia, and urinary retention [6]. For cervical lymphomas, the median age of presentation is 46 years (range 20-85 years), and the most common histological type is DLBCL (37%). Only 5% of the cases, described in the literature, are follicular NHL [9]. We present a case of DLBCL, however of the non-germinal center type according to the IHC. Malignant lymphoma has a monomorphic appearance, often with cellular necrosis or sclerosis, and immune staining is necessary [5]. Malignant lymphoma is a rare form of malignancy in the uterine cervix. Most of the previously reported cases were B-cell lymphomas, however, Kosari F and his colleagues present a case of primary peripheral T-cell lymphoma, not otherwise specified, in the uterine cervix of a 49-year-old woman with a history of severe vaginal bleeding. The patient was treated with a CHOP (cyclophosphamide, adriamycin, vincristine, and prednisolone) chemotherapy regimen and 28 cycles of radiotherapy. Three months later, she presented with generalized lymphadenopathy and sudden left-eye ptosis as a complication of CNS involvement. She died after 20 months of the initial disease presentation [10]. Cubo AM [3], reported a 51-year-old woman with a primary diffuse large B-cell lymphoma of the cervix. Diagnosis of this tumor was a challenge for the pathologists and clinicians. Four biopsies were needed to find the diagnosis. The patient was treated with combined Rituximab and chemotherapy (R-CHOP) with complete remission after 2-years of follow-up. A cervical leiomyoma was suspected in the case of Li WS [8], similar to the present case, however, the pathology diagnosis was nodular lymphocyte predominant Hodgkin lymphoma.

Table 1 compares characteristics of some previously reported primary uterine cervical lymphomas with the present case.

Table 1. Primary uterine cervical lymphomas

Study (Year)	Clinical presentation	Age(Year)	Tumor size(cm)	Histologic type	IHC positivity
Cubo AM [3] 2017	Postmenopausal vaginal bleeding	51	10	DLBCL	CD20, CD5, BCL-2, BCL-6, CD45, CD23, P53, CD43
Gui W [11] 2019	Postmenopausal vaginal bleeding	65	6	DLBCL	CD20, c-Myc, BCL-2, BCL-6, MUM1, P63
Gui W [11] 2019	Leucorrhea, contact vaginal bleeding	43	4	MALToma	CD20
Gui W [11] 2019	Vaginal bleeding	36	4	DLBCL	CD45, CD20, P63
Roberts ME [7] 2018	Abnormal vaginal bleeding	55	Less than 10	DLBCL	CD20
Regalo A [12] 2016	Pain and swelling of right lower extremity, uterine cervix mass	40	7.9	Non-Hodgkin follicular, large B-cell lymphoma	CD20, CD10, BCL-2, BCL-6
Yang G [4] 2017	Irritative urinary voiding	69	18	B-cell lymphoma	CD45, CD20, PAX5, MUM1
Lavleen S [13] 2014	Abdominal pain for 3 months	34	6	DLBCL	CD20
Lavleen S [13] 2014	Fever and abdominal pain	77	Not mentioned	DLBCL	Not mentioned
Li WS [8] 2015	Abnormal vaginal bleeding for one year	43	5.3	Nodular lymphocyte predominant Hodgkin lymphoma	CD21
Wang GN [5] 2015	Difficulty in micturition	54	4	Nasal type NK/T cell lymphoma	CD3, CD43, CD56, Granzyme-B, TIA-1
Mouhajir N [6] 2014	Menorrhagia and postcoital bleeding	49	Not mentioned	Diffuse B-cell non-Hodgkin lymphoma	CD20,CD45
Anagnostopoulos A [9] 2013	Incidental findings in an ultrasound exam	65	No mass	Low-grade follicular center cell lymphoma	CD20,BCL-2,CD3
Present case	Mass in an ultrasound exam	34	4.3	Diffuse B-cell non-Hodgkin lymphoma, Non-Germinal Center Type	Ki-67 (in 80% of tumor nuclei), LCA, Vimentin (in few tumor cells), CD20, CD19, and BCL6 (in the majority of tumor cells)

4. Conclusion

Primary lymphoma of the uterine cervix is rare and confusing, due to non-specific symptoms. Clinical presentations are easily mistaken for squamous cell carcinoma of the cervix, other malignancies, and inflammatory process. Treatment and prognosis are considerably different in the long list of differential diagnoses. Sometimes multiple biopsies and immunohistochemistry are needed for a definite diagnosis. Pathologists and clinicians must be aware of this entity in daily practice.

Conflict of Interest

None.

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