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CASE REPORT

Primary lymphoma of a cervix - a devil in disguise

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Abstract

Primary Lymphoma of the female genital tract a rare disease. Primary cervical lymphoma accounts for .008% of malignant cervical tumours. The symptoms and signs are nonspecific and very common. Papanicolaou cervical smear is not too much helpful in screening. Standard management protocol is not available for this disease. Here, we discussed a case of primary NHL of cervix presenting with abnormal bleeding per vaginum.

Keywords: primary cervical lymphoma, NHL -Non-Hodgkin's lymphoma, pap- smear, IHC – immunohistochemistry.

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Introduction

Lymphoma of the female genital tract is a rare disease [1]. Cervical involvement in non-Hodgkin's lymphoma (NHL) is common as a part of multiorgan disease but primary involvement of cervix is very uncommon. Because of rarity of disease, the clinical course, management and prognosis of primary gynaecological lymphoma is not well defined in literature. Here, we present a case of primary NHL of cervix presenting with abnormal bleeding per vaginum.

Case Report

A 47-year-old, P2 L2, diabetic and hypertensive women reported to Cancer Hospital and Research Institute in Gwalior after hysterectomy which was performed elsewhere. The histopathology of resected specimen revealed NHL of cervix.

On evaluation of old records, she had a history of irregular bleeding per vaginum for 6 months, which was not relieved with medical management. On preoperative examination, cervix was normal, uterus was bulky and retroflexed; bilateral fornices were normal. Her preoperative pap smear was normal and ultrasound showed a small intramural fibroid at fundus. She had no history of prolonged fever, weight loss, and night sweats. There was no history of similar condition in the family. She had no significant past and personal history.

She underwent total abdominal hysterectomy and bilateral salpingo-oophrectomy for fibroid uterus. Histopathology of surgical specimen revealed high-grade B cell lymphoma of cervix. This was confirmed on immunohistochemistry (IHC) as leucocyte common antigen (LCA) and CD20 showed strong and diffuse membranous positivity respectively while CK was negative in neoplastic cells.

Postoperative CT scan, one month after surgery, showed a large heterogenous mass in the uterine bed with infiltration of posterior wall of urinary bladder, rectum, sigmoid colon, and both distal ureters causing bilateral mild hydronephrosis. Her metastatic workup with CT thorax, bone marrow cytology, and peripheral smear were all normal. The disease was staged as primary cervical NHL stage II E using the Ann Arbor system. She was treated with 3 cycles of Rituximab, Cyclophosphamide, Doxorubicin, and Vincristine. Following 3 cycle of chemotherapy, she developed hydrocephalous with weakness of right upper and lower limb with neutropenia and deraigned renal function test. Despite treatment, her condition deteriorated and she expired after 16 weeks of primary surgery.

Discussion

Primary cervical lymphoma accounts for 0.008% of malignant cervical tumours [1]. The age of patient at diagnosis for cervical NHL ranges from 27 to 80 years (median 44years) [2]. The most frequent symptoms are pre or postmenopausal abnormal bleeding per vaginum, vaginal discharge, and abdominal pain. These symptoms are non-specific and common in gynaecological practice.

Physical examination reveals a diffusely enlarged or barrel-shaped cervix, often with absence of erosive or exophytic lesion. Therefore, it is difficult to differentiate NHL from benign lesions, like cervical fibroids, cervical inflammation, or Nabothian cysts clinically [3] (Figures 1 and 2).

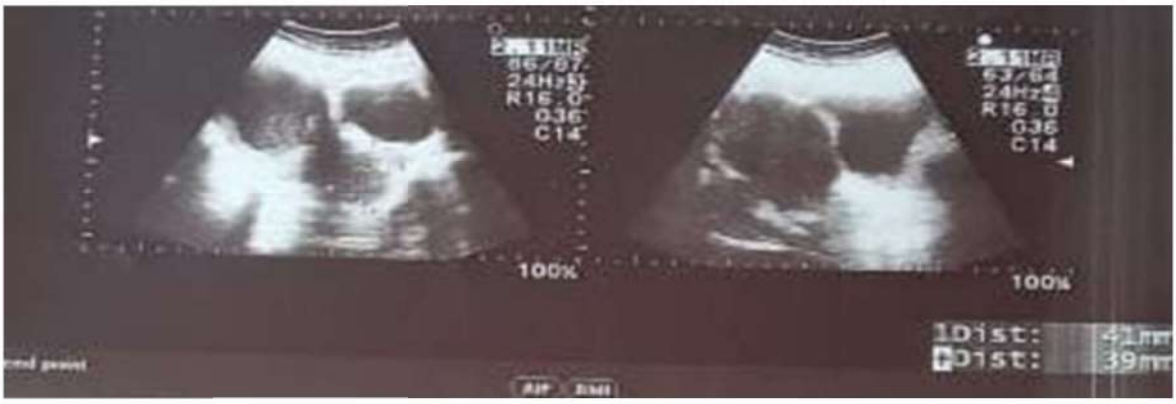

Figure 1. Uterus is anteverted & anteflexed appears bulky with small 2cm size hypo echoic area at fundus represents intra mural fibroid. Cervix appears bulky without a definite growth or mass lesion. Peri-uterine fat planes with adjacent structures are grossly normal

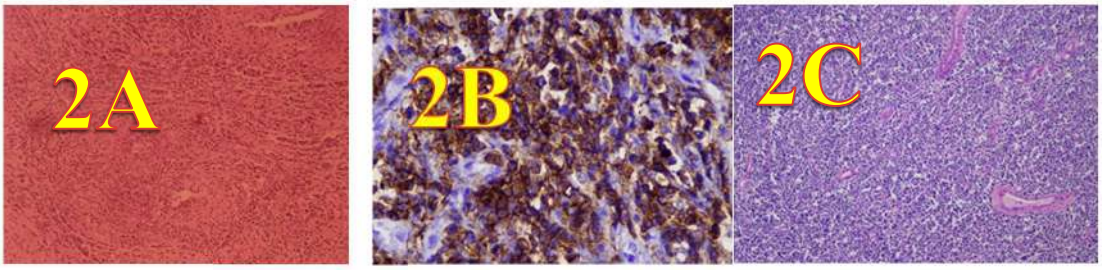

Figure 2. 2A: H&E (200x) - Large atypical lymphoid cells arranged in diffuse sheets. 2B & 2C: Immunohistochemistry (400x)- Tumour cells throwing diffusely positivity with LCA & CD20 respectively

Table 1. Interpretation of immunohistochemical stains in large lymphoid cells lymphoma.

CD45+(LCA)	Marker for leukocyte common antigen found in hematopoietic cells, thus indicating lymphoma.
S100+, PNL2+	Marker for melanoma.
Cytokeratin+	Marker for carcinoma.
CD20+	Marker for B-cell lymphoma.
CD10+, Bc12-, Ki67>99%	Immunohistochemical stain.
CD10+/-, Bc12+	Favours diffuse large B-cell lymphoma.
CD3+, CD20-	Marker for T-cell lymphoma.

Pap smear is not helpful in the screening and diagnosis of cervical lymphomas. As they usually arise from the cervical stroma rather than the mucosa, thus lacking the surface abnormalities. There is no gold standard among imaging techniques for diagnosing pelvic lymphoma. A high index of suspicion and use of ultrasound, MRI, and CT can suggest the diagnosis. PET CT may be performed for staging and metastatic assessment [4].

The importance of immunohistochemistry (IHC) in the diagnosis of primary malignant cervical lymphomas is well recognised (1). Currently, there is no specific IHC marker for lymphoma. Use of immunohistochemical stains, listed in Table 1, is necessary to establish a definite diagnosis when initial biopsy and usual stains are nonconclusive [3].

There is no consensus regarding the best treatment for Primary NHL of female genital tract. However, lymphomas are considered sensitive to chemotherapy and radiotherapy [4]. Chemotherapy regimen with R-CHOP is usually employed for lymphoma [2].

Case series reported that in most cases the management also includes surgery and/or radiation therapy. However combination of surgery and radiation therapy did not confer any survival benefit in most of cases. The role of surgery to improve overall survival is still unknown [1,3,5].

The prognosis may be excellent if the cervical lymphoma is diagnosed and treated correctly in an early stage. The 5-year survival rates have been reported to be 89% in early stage and 20% in II E-1V stage cases [1].

Conclusion

Primary cervical lymphoma is an uncommon disease that provides clinicians a diagnostic challenge. The diagnosis is often delayed due to non-specific clinical presentation and low index of suspicion. This condition has to be suspected in women with abnormal bleeding per vaginum, a suspicious cervix, and a normal Pap smear test. It is essential that the clinicians, radiologists, and pathologists have awareness for this rare disease.

Conflicts of interest

The author declares that they do not have conflict of interest.

References

1. Capsa C, Calustian LA, Antoniu SA, Bratucu E, Simion L, Prunoiu VM. Primary Non-Hodgkin Uterine Lymphoma of the Cervix: A Literature Review. Medicina (Kaunas). 2022 Jan 10;58(1):106. doi: 10.3390/medicina58010106.
2. Aminimoghaddam S, Salarifar E, Noei Teymoordash S. Primary large B-cell lymphoma of the cervix: A case report and review of literature. Clin Case Rep. 2022 Mar 24;10(3):e05639. doi: 10.1002/ccr3.5639.
3. Wang J, Zeng L, Chen S, Wu Q, Ma L, Wu S, Wang ZP, Tao Y, Chai D. Lymphoma of the female genital tract: a clinicopatholngical analysis of 25 cases. Am J Transl Res. 2019 Sep 15;11(9):5800-5811.
4. Del M, Angeles MA, Syrykh C, Martínez-Gómez C, Martínez A, Ferron G, Gabiache E, Oberic L. Primary B-Cell lymphoma of the uterine cervix presenting with right ureter

hydronephrosis: A case report. Gynecol Oncol Rep. 2020 Sep 11;34:100639. doi: 10.1016/j.gore.2020.100639.

5. Nasioudis D., Kampaktsis P.N., Frey M., Witkin S.S., Holcomb K. Primary lymphoma of the female genital tract: An analysis of 697 cases. Gynecol. Oncol. 2017;145:305–309. doi: 10.1016/j.ygyno.2017.02.043.