Primary Lymphoma of the Uterine Cervix: A Case Report

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Abstract

Primary lymphoma of uterine cervix is a rare disease. Diffuse large B-cell lymphoma is the most common subtype. We report the case of a 66-year-old woman who presented for post-menopausal metrorrhagia. Physical examination revealed a voluminous bleeding mass of the uterine cervix. The biopsy showed a non-Hodgkin's lymphoma with large B cells. The extension assessment was normal. The patient was put on RCHOP protocol (Rituximub, cyclophosphamide, doxorubicin, vincristine, prednisone) followed by radiotherapy with complete response.

Keywords: Non-Hodgkin's Lymphoma; Uterine Cervix; Metrorrhagia; Chemotherapy; Radiotherapy.

Introduction

Primary non-Hodgkin's lymphoma of the female genital tract is a rare entity that represents less than 1% of extra nodal lymphomas [1]. Primary lymphoma of the uterine cervix is extremely rare with an overall incidence of less than 1% among all malignant tumours of the uterine cervix[2] Cervical involvement in multi-organ disease is more common than in primary lymphoma[3]. The most common histological subtype of female genital lymphomas is diffuse large B-cell lymphoma [4]. The common clinical presentation is vaginal bleeding, although occasionally atypical lymphoid cells on a routine cervical smear have led to detection [5]. The cervical lymphomas may be misdiagnosed or delayed on Papanicolaou (Pap) smears by the pathologist because lymphomas are unexpected at this site. In fact, often the diagnosis of these tumours is not suspected clinically and it is set only after biopsy. We report the case of a 66-year-old female who presented for post-menopausal metrorrhagia, in which the cervico-vaginal smear was normal while the biopsy concluded a primary non-Hodgkin's lymphoma of the cervix. The aim of our work is to highlight the clinical and therapeutic particularities of this rare entity.

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Case Report

A 66 - year old female presented with postmenopausal metrorrhagia evolving for 5 months. The gynaecological examination objectified a voluminous bleeding mass of the uterine cervix infiltrating the two parameters. The rest of the somatic examination was normal. The pelvic CT scan revealed an increase in the volume of the cervix, measuring 9×9 cm taking contrast heterogeneously with significant infiltration of the parameters (figure 1). The morphology of the rest of uterus was normal. The cervico-vaginal smear was normal while the Cervix biopsy showed effaced architecture by a diffuse proliferation of atypical lymphoid cells, intermediate /large size, with small cytoplasm, and vesicular nuclei (Figure 2). Immunohistochemistry objectified the expression of CD20, BCL2, CD10, BCL6, CD45, CD23 and CD. The definitive diagnosis was

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Germinal centre non-Hodgkin's lymphoma with large B cells. Metastatic workup was normal. The tumour was classified IE according to the Ann Arbor classification. She received 8 cycles of chemotherapy (Doxorubicin, Cyclophosphamide, Vincristine, and Prednisolone) with Rituximab (R-CHOP). There was complete response after 8 cycles. She then received radiotherapy (45 Gray in 25 fractions). She has been followed up for the last 2 years, is symptom-free and there is no disease activity.

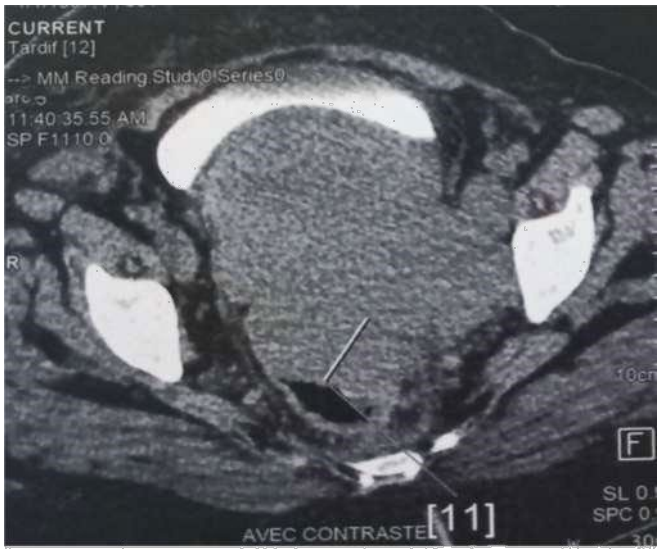

Figure 1: Pelvic CT scan: a voluminous mass of the cervix taking contrast heterogeneously.

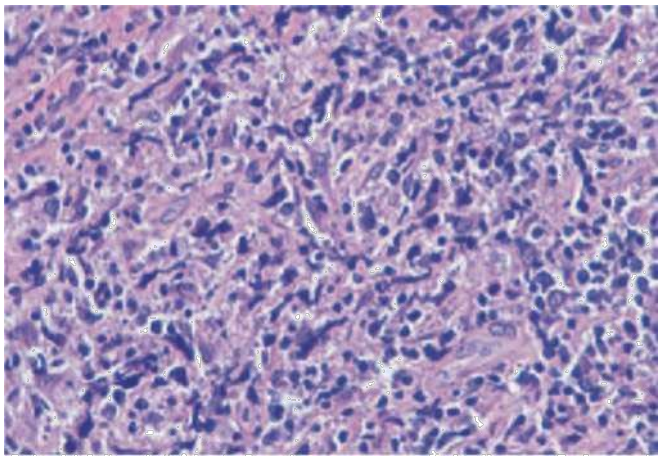

Figure 2: Atypical lymphoid cells, large - intermediate size, with small vesicular nuclei and cytoplasm

Discussion

Primary cervical lymphoma is an extremely rare entity representing 0.12% of all primary non-Hodgkin's lymphomas [6]. In a series of 147 primary non-Hodgkin's genital lymphomas, ovarian lymphoma was the most frequent (59%), followed by the uterine corpus (15.5%), uterine cervix (11.5%) and then the vulva and vagina [7]. The majority of lymphomas of the cervix are of the diffuse large B cell type, but other subtypes have been described, including Burkitt's lymphoma, marginal zone lymphoma and follicular lymphoma.

The Diagnosis of primary uterine cervix NHL is difficult because of its rarity and also because the clinical presentation mimics that of squamous cell carcinoma. The diagnosis of these tumours is not suspected clinically and it is histological. Most cases have been described in postmenopausal women like the case of our patient, but some studies also report cases of premenopause.The median age at presentation is around 44 years [8]. No sign is specific; but the symptomatology is dominated by a polyploid mass; metrorrhagia and more rarely pelvic pain and dyspareunia [1]. The etiological factors include infectious agents such as the human immunodeficiency virus (HIV); immunosuppressive therapies and exposure to pollutants and pesticides.

Screening for cervical cancer by systematic examination, using the cervico-vaginal smear, began in the 1950s and has proven its effectiveness in industrialized countries by reducing mortality of this cancer by more than 50% [9]. Primary cervical lymphoma is rarely suspected on the smear. In the case of our patient, the smear was normal while the biopsy revealed a diffuse NHL with large B cells. This is probably due to the fact that these tumours infiltrate the cervical stroma and that the squamous and glandular epithelial mucosa is initially preserved [2]. In a study by Dursun et al [10]. 41% of women with primary cervical lymphoma had abnormal cervical cytology.

The most common stage at diagnosis is stage IE according to the Ann Arbor classification [11]. Since cervical NHL is such a rare disease, no standard treatment has been defined and different treatment options are proposed from more aggressive therapies years ago, where surgery played a key role in treatment, to more conservative therapies, where chemotherapy combined with or without

radiotherapy seems be the mainstay of current treatment [8]. Combination chemotherapy regimens like CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisolone) have been shown to be effective treatment for this disease. Rituximab, a monoclonal antibody directed against the CD20 antigen of B cells, has been shown to that adding it to the CHOP chemotherapy regimen improved overall survival for these tumours [12].

Stroh et al [13] reported 16 cases of cervical lymphoma, achieved a 5-year disease-free survival of 90 % by the use of chemotherapy combined with radiotherapy. They concluded that combined modality treatment with CHOP chemotherapy and radiation is considered favoured treatment for cervical NHL. Autologous stem cell transplantation could be susceptible for relapsed or refractory lymphomas with complete or partial remission, but its use in first intention is not supported by any recent meta-analysis [14]. Although the prognosis of primary gynaecological lymphomas cannot be established given the low number of series, the estimated overall survival at 5 years varies between 73% [15] and 77% [13].

Conclusion

Primary lymphoma of the uterine cervix is a rare disease. Most patients present with stage IE. Treatment often includes a combination of immune-chemotherapy and radiotherapy with good results. Further studies are needed to standardize the treatment protocols.

Declaration:

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Authors' contributions

All authors have read and approved the final version of the manuscript

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Conflicts of interest

None.

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