Extranodal Burkitt’s Lymphoma with Uterine Involvement: A Case Report

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Extranodal Burkitt’s lymphoma with uterine involvement is uncommon. We present the case of a 71-year-old woman with bilateral edema of the legs and a decreased amount of urine for one month. A CT scan revealed enlargement of the uterus with a rather homogenous infiltrative mass lesion, and a Pap smear exhibited atypical lymphocytes. Endocervical curettage revealed NHL of the uterine cervix. The right inguinal lymph nodes are also involved. Both the morphologic features and the immunophenotype indicate Burkitt’s lymphoma, a highly malignant, aggressive and rapidly growing B-cell neoplasm. The patient was treated with a combination of multiple drug therapy and high-dose chemotherapy, but died 6 months after her initial presentation at our hospital.

Key words: non-Hodgkin’s lymphoma, Burkitt’s lymphoma, uterus

Introduction

The majority of extranodal lymphomas are instances of secondary involvement. The genital organs are infrequently involved with lymphomas, and therefore the disease is rarely associated with gynecologic symptoms. Primary non-Hodgkin’s lymphoma (NHL) of the female genital organs is an uncommon condition, accounting for approximately 1% of primary extranodal lymphomas [1]. Burkitt’s lymphoma originating in the cervix or uterus is extremely rare, with only 4 cases reported so far in the English literature [2–5]. Non-Hodgkin’s lymphoma (NHL) can involve extranodal sites, the most common being the gastrointestinal tract and the skin; however, uterine involvement with non-Hodgkin’s lymphoma (NHL) is uncommon. We present a case of Burkitt’s lymphoma that involved the uterus and the right inguinal lymph nodes. Burkitt’s lymphoma is a highly malignant, aggressive and rapidly growing B-cell neoplasm that has low long-term survival rates.

Case History

A 71-year-old woman was suffering from bilateral edema of the legs and a decreased amount of urine for one month. There was history of CAD (coronary artery disease) with regular follow up and medical control at a local medical department for 25 years. The patient presented at Chi-Mei Medical Center Hospital and clinical examination revealed the following results: WBC, 11.3 ×10³/uL; RBC, 3.67 ×10⁶/uL; Hb, 10.4 mg/dL; PLT, 291 ×10³/uL; creatinine, 1.61 mg/dL; CA-125, 41.8 IU/mL; and other levels within normal limits. A CT scan of the pelvis showed the following: hydronephrosis of the right kidney with poor perfusion, enlargement of the uterus with a rather homogenous infiltrative mass lesion, which invaded into the right adexa and extended to the surrounding fatty planes of the pelvic floor, and lymphadenopathy over the right inguinal region. On transvaginal sonography, one cervical mass of about 5.3 ×5.1 cm with myometrial extension was shown. A Pap smear was performed, followed by cervical curettage and a biopsy...
of the right inguinal lymph node.

The cytology smear revealed normal-looking epithelial cells and many atypical lymphocytes with slight variation in size but no maturation. These atypical lymphoid cells had round to oval, notched nuclei, coarse chromatin and distinct nucleoli; no tangible body macrophage was found (Fig.1A). The section of endocervical curettage showed mainly blood clots and fragmented endocervical glands, as well as a fragment of stroma tissue infiltrated by atypical lymphoid cells. The architecture of the lymph node was almost totally effaced by monotonous infiltrating tumor cells with a starry-sky pattern, the infiltration being composed of medium-sized atypical lymphocytes with frequent mitosis. The surrounding adipose tissue was also involved. Immunohistochemically, these atypical lymphocytes express CD10, CD20, Bcl-6 and IgM (Fig.1B–E). The proliferation fraction as determined by Ki-67 immunostaining was almost at 100% (Fig.1F). Both the morphologic features and immunophenotype indicated a Burkitt’s lymphoma. The patient was treated with a combination of multiple drug therapy and high-dose chemotherapy, but the patient died 6 months after her initial presentation at our hospital.

Discussion

Uterine involvement in non-Hodgkin’s lymphoma (NHL) is uncommon. Most patients experience abnormal bleeding and a large, bulky cervix. Cervical cytology is usually negative [6]. Cytology is not a sensitive tool for detecting lymphoma, because lymphoma typically originates from the stroma, and the superficial squamous epithelium is often present leading to a false negative result. The diagnosis of lymphoma in the cervix is based mainly on the results of a cervical biopsy [7–8]. In the present case, a Pap smear revealed only normal squamous epithelial cells and many atypical lymphoid cells.

So-called lymphoma-like lesions of the cervix were first described in 1985 [9], differing from typical cervical lymphoma in superficial distribution, heterogeneity of lymphoid cells, and presence of inflammatory cells. Our case showed diffuse infiltration of monotonous lymphocytes with coarse chromatin in the cervix and uterine corpus. Therefore, the diagnosis of a lymphoma-like lesion was excluded. In the present case, positive results for CD10, CD20, Bcl-6, a nearly 100% proliferate index and the starry-sky pattern of the H&E slide are all indicative of a Burkitt’s lymphoma.

Most NHL of the female genital organs are aggressive and treatment is delayed due to late diagnosis. The management of NHL in this location has not been well-defined due to the low incidence of the disease and limited discussion in the literature. Primary NHL involvement of the female genital organs may be successfully treated by pelvic irradiation, but in younger women cytotoxic chemotherapy should be considered first to preserve fertility. The most effective treatment regimens are a combination of radiotherapy and chemotherapy [10]. Neoadjuvant chemotherapy followed by surgery was emphasized by Kuo et al. [11].

The prognosis for patients with extranodal lymphomas is usually poorer than for those with nodal lymphomas due to inaccurate or delayed diagnosis. If diagnosed in the earlier stages, prognosis may be excellent compared to other gynecologic malignancies.

References

Extranodal Burkitt’s lymphoma with uterine involvement

Fig. 1. (A) The cytology smear revealed normal-looking epithelial cells and many atypical lymphocytes with slight variation in size but no maturation. These atypical cells had round to oval, notched nuclei, coarse chromatin and distinct nucleoli. No tangible body macrophage was found (Pap stain 400×). (B) Tumor cells express membrane IgM with light chain restriction and (C) B-cell associated antigens (CD20). (D) The expression of CD10 and (E) Bcl-6 point towards a germininal center of origin for the tumor cells. (F) Ki-67 labeling index, associated with high mitotic activity is at 100% in the tumor cells. (immunostain 400×).
淋巴結外子宮 Burkitt’s 淋巴癌：一案例報告

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淋巴結外 Burkitt’s 淋巴癌發生在子宮是很少見的，我們報告了一位 71 歲的婦人，他的雙下肢腫大而且尿量減少約一個月，而在骨盆腔的斷層攝影中，發現此病人的子宮腫大，腫大的因素是含有均質浸潤的腫瘤所造成，在細胞學抹片(Pap smear)檢查發現細胞含有非典型的淋巴球，追蹤子宮頸病理切片報告的結果，發現是非賀金傑氏淋巴癌，而且也轉移到右腹股溝的淋巴結。在病理細胞型態及免疫染色分類兩者的診斷，我們判定了這是一例罕見的女性生殖器官 Burkitt’s 淋巴癌。Burkitt’s 淋巴癌是一個高惡性侵犯及生長快速的 B 細胞腫瘤，僅管臨床給予病人多種藥及高劑量的化學治療，病人還是在 6 個月後就死亡。

關鍵詞: 非賀金傑氏淋巴癌、Burkitt’s 淋巴癌、子宮