Successful treatment of a primary uterine B-cell lymphoma with rituximab-CHOP immunochemotherapy

We report the case of a 26 year old patient with a primary, Ann Arbor stage IE, diffuse large B-cell lymphoma of the uterine corpus. The patient was admitted to our hospital because of a one year history of vaginal contact bleeding. Final diagnosis was based on the histological and immunohistochemical evaluation of a specimen obtained by fractionated cervical and uterine curettage. Further staging has not revealed any other localization of the disease. Antilymphoma treatment was started with CHOP combined with rituximab and resulted complete remission. This is the first reported case of an uterine lymphoma treated by rituximab-based immunochemotherapy.

Extranodal involvement in non-Hodgkin lymphoma (NHL) is usual, but the primary uterine occurrence is extremely rare. An extensive histopathological study reviewing 1766 nodal and extranodal cases of lymphomas depicted only 7 cases of uterine origin. Moreover, most of the diagnosed uterine lymphomas have cervical localization with approximately 100 cases reported. In contrast, the overall number of uterine corpus lymphomas in the literature not exceeds 25 cases. It is accordance with the result of a study on 1467 extranodal NHL cases where the frequency of primary uterine corpus lymphoma was 0.002%. Most of the reported cases were diffuse large B-cell lymphoma (DLBCL), but rarely other histological types was also diagnosed.

In our case, a 26 years old Hungarian woman visited the gynecological unit of our hospital complaining to atypical contact vaginal bleeding during the last one year. A spontaneous delivery of a healthy child and later an artificial abortion was reported in her medical history. Her Papanicolaou smear was P II. The performed fractionated curettage revealed smooth internal surface of the uterus with measured cavity length of 7 cm. The histopathological assessment of the obtained tissue sample showed cervical and endometrial gland parts surrounded by proliferating diffuse cellular infiltrate. The infiltrate is heterogenous, the main elements are blastic cells with roundish nucleus and several nucleoli resembling centroblasts. There are reactive cells as well, small lymphocytes, plasma cells and eosinophils. During the immunohistochemical analysis these cellular elements were negative to myeloid markers and cytokeratine but showed massive positivity to CD45 and B-cell markers including CD20. These findings established the diagnosis of a diffuse large B-cell lymphoma which was confirmed by the analysis of new specimens from a second curettage done a month later. Additional immunohistochemical analysis was done later, the tumor cells were CD10 and MUM-1 negative and bcl-6 positive, this phenotype suggest a germinal center (GC) derivation for the tumor cells, the tumor is a GC-type DLBCL. There was no palpable spleno-, hepato- or lymphadenomegaly. The transvaginal ultrasonography showed a normal sized uterus (80x47 mm) with normal myometrial structure and a 4 mm thick endometrial layer. The complete staging including CT-scans of the neck, chest and abdominal regions, bone marrow biopsy, complete and detailed pelvic MR-scans does not indicated any myometrial, nodal or other organ involvement (Ann Arbor stage IE, IPI: 0). The oncological team decided the administration of 6 cycles of 375 mg/m² rituximab combined standard CHOP chemotherapy (R-CHOP). During the chemotherapy there was not any unexpected complications, menstruation kept its regularity with less bleeding than it was before. After completion of chemotherapy, transvaginal ultrasonography showed again a normal sized uterus (77x52 mm) with a 6.2 mm thick endometrium. As a control, a new fractionated curettage was done, and the subsequent histopathological review of the obtained endometrial sample showed normal secretory endome-
Abnormal uterine bleeding is the most common presenting symptom beside a pelvic mass in advanced state. On the basis of recent studies, the standard and effective DLBCL therapy, regardless of age, localization, stage or IPI, is the anti-CD20 immunotherapy (rituximab-CHOP), proper histopathological assessment is vital to distinguish this disease from other malignancies or benign lymphoproliferative lesions. Because of the very few cases reported and in the light of results usefulness of described treatment options including total abdominal hysterectomy, salpingo-oophorectomy, pelvic node dissection, radio- and/or chemotherapy are controversial.

In the therapeutic decision regarding this patient our oncological team considered the fact that she was in reproductive age. Preserving the ovarian functions and avoiding sterility induced by local irradiation or hysterectomy was a weighty issue. Moreover, local curative intents do not control micrometastases, and patient can further die from distant recurrences despite adequate local control. Knowing that R-CHOP became the new treatment standard of DLBCL, choosing this regimen was quite obvious even it was never reportedly done before our group.

This is the first reported case of a uterine lymphoma treated by rituximab-based immunotherapy.

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References