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Letter to the Editor

Primary malignant lymphoma of the uterus: a report of four cases

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Primary lymphomas are extremely rare in the uterus. Primary lymphomas of the uterus comprise only 0.002% of all extra-nodal lymphomas. Among uterine lymphomas, the most common seem diffuse large B-cell lymphoma (DLBCL), followed by other types of B-cell neoplasmas [1]. The prevalence of uterine ones is not known. Extra-nodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT, MALToma) is an indolent B-cell neoplasms mainly found in the stomach, where Helicobactor Pylori (HP) infection-related-gastritis is very common, followed in order by ocular area, thyroid, skin, and other rare organs. MALT lymphoma is once regarded as inflammation, but the fact that the inflammation can give rise to DLBCL and that there are light-chain restriction suggesting B-cell monoclonality and even IgHc rearrangement have suggested that such lymphoid infiltrates as MALT lymphoma are lymphomas. However, gastric MALT lymphoma can resolve after removal of HP. In recent years, it was suggested that chronic long-standing inflammation can lead to the development of carcinomas and lymphomas, and that such inflammatory lymphoid infiltrates are preneoplastic or early neoplastic lesions, for example Hashimoto thyroiditis.

The author recently encountered MALT lymphoma of the uterine endometrium, which facilitated the author to survey lymphomas of uterus samples (both cervix and corpus) in the computed data base during the past 13 years, which included 9,529 specimens. Four cases of primary lymphomas of uterus were found; the prevalence was 0.04%. All four cases occurred in endometrium. There were no cases whose involvements were limited to the cervix or myometrium. Myometrial and cervical involvements were seen in 4/4 and 1/4, respectively. All four cases were local diseases and no nodal involvements were noted. The stages were Ann-Arbor Stage I in all cases. The age of the four cases was as follows: 40, 49, 71, and 77 years (range 40-77, median 60, and mean 59). The initial manifestations were blood discharge in two cases, uterine prolapse in one, and CIN3 in one. Diagnosis was made by biopsy in three and by hysterectomy in one. Three cases were indolent B-cell neoplasms consisting of MALT lymphoma (two cases) and lympho-plasmacytic B-cell lymphoma (LPL) (one case), and the remaining one was DLBCL. All the cases showed microscopical tumorous proliferation of atypical small lymphocytes. The treatment was R-CHOP in all cases. CR was obtained in each case, which was clinically suggested in three cases and confirmed by biopsy in one case. No relapse was noted.

The MALT lymphoma cases (Case 1 and Case 2) showed tumorous severe lymphocytic infiltration consisting of predominantly centrocyte-like cells (CLC) and small B cells, formation of lympho-epithelial lesions (LEL), focal plasma cell differentiation, presence of monocytoid B cells, and formation of germinal center-like follicles (Figures 1A and 1B). Follicular colonization was not seen. The LPL case (Case 3) showed tumorous proliferation of small atypical lymphocytes and plasma cells (Figure 1C): no features of MALT lymphoma were seen. The DLBCL case (Case 4) showed monotonous tumorous proliferation of large atypical lymphocytes (Figure 1D).

Immunohistochemically, immunophenotype of the MALT lymphomas was as follows: Case 1: CD45+++, CD20+++ (Figure 2A), CD79a++, CD138++, CD10 -, light chain restriction + (kappa>>lamda), CD3+, CD 45RO+, CD56-, p53+, Ki-67 21%, cytokeratins-; Case 2: CD45+++, CD20+++, CD79a++, CD138++, light chain restriction + (lamda>kappa), CD3+, CD 45RO-, CD56-, p53++, Ki-67 19%. The immunophenotype of LPL (Case 3) was as follows: CD45+++, CD20+++, CD79a+++, CD138+++, light chain restriction ++ (kappa>>lamda), CD3+, CD 45RO-, CD56-, p53+, Ki-67 23%. The immunophenotype of DLBCL (Case 4) was as follows CD45+++, CD20+++, CD79a+, CD138+,

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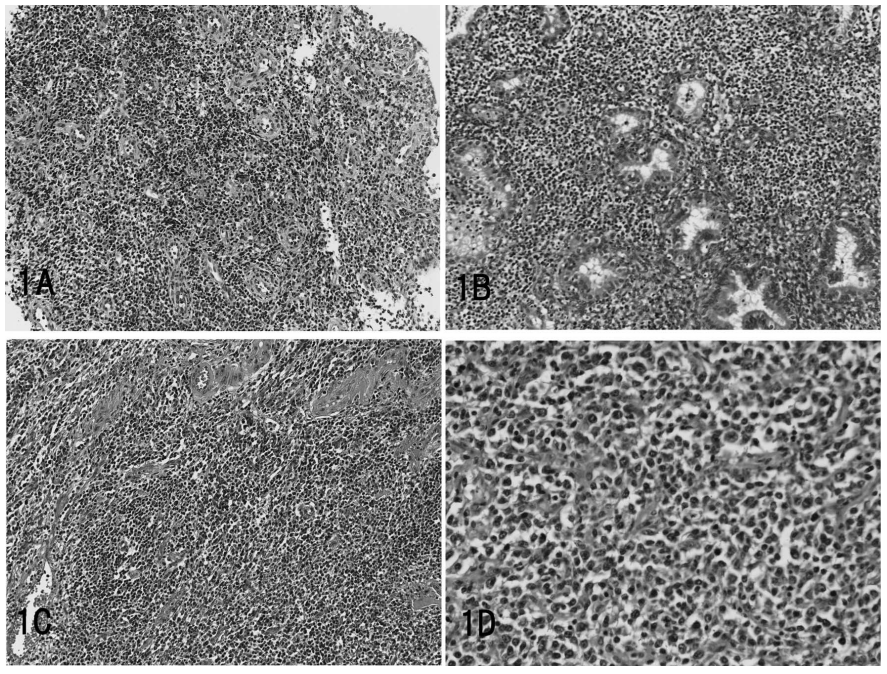

Figure 1. — Histology of the four cases of uterus lymphoma. A) and B) (Cases 1 and 2, both MALT lymphoma): MALT lymphoma of endometrium. Proliferation of small atypical lymphocytes are seen. Focal areas show lymphoepithelial lesions. A), B); HE, x40. C) (Case 3): lympho-plasmacytic lymphoma. Proliferation of atypical small lymphocytes and plasma cells is recognized. HE, 40. D) (Case 4): diffuse large B-cell lymphoma non-GC. Diffuse proliferation of atypical lymphocytes is noted. HE, x100.

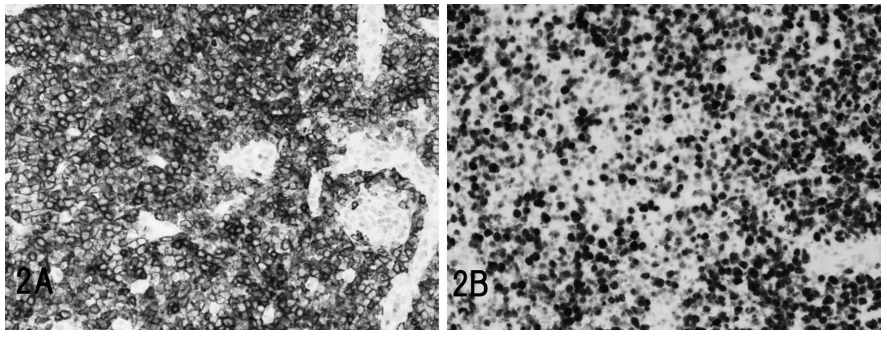

Figure 2. — Immunohistochemical findings. A) (Case 1): MALT lymphoma. The atypical cells are positive for CD20. x200. B) (Case 4): Diffuse large B-cell lymphoma. The Ki-67 labeling is very high (in this field circa 50%, x200).

CD10 -, MUM1 +, bcl-6 +, CD3-, CD 45RO-, CD56-, p53+++, Ki-67 53% (Figure 2B), cytokeratins-. Thus, the DLBCL was non-germinal B-cell type (DLBCL, non-GB).

Conclusions

Lymphoma of the uterus is very rare; the incidence being around 0.04% of all uterine lesions. Uterine lymphoma is mostly seen in endometrium with frequent myometrial and cervical involvements. Most uterine lymphomas are of B-cell lymphoma: T-cell, NK/T cell lymphomas were not found. Low-grade indolent B-cell lymphoma predominated over high-grade B-cell neoplasmas.

References

[1] Sugimoto K., Imai H., Shimada A., Wakabayashi M., Sekiguchi Y., Nakamura N., *et al.*: "Diffuse large B-cell lymphoma of the uterus suspected of having transformed from a marginal zone B-cell lymphoma harboring trisomy 18: a case report and review of the literature". *Int. J. Clin. Exp. Pathol.*, 2013, *6*, 2979.

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