

PRIMARY LYMPHOMA OF THE VAGINA. A CASE REPORT

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ABSTRACT

Primary vaginal non-Hodgkin lymphoma is really uncommon and may be misdiagnosed as inflammatory disease or solid cancer, so careful diagnostic procedures are needed, particularly as far as pathological and immunocytochemical evaluation is concerned. Most of these lymphomas present with follicular patterns and limited stage disease, so high cure rates are possible with surgery and/or radiotherapy, but chemotherapy has to be considered on the basis of clinical presentation and pathologic features. We report a case of vaginal lymphoma with primary bulky ulcerated mass that was treated with chemoradiotherapy.

Key words: non-Hodgkin lymphoma, vagina

Primitive extranodal malignant lymphomas may occur in the female genital tract in about 30% of cases. Most of them are non-Hodgkin lymphomas involving, in order of frequency, ovary (50%), uterus (30%), tubes (10%), vagina (6%) and vulva (4%). Primary appearance of a malignant lymphoma in the vagina is really uncommon in that no more than 21 cases have been described so far.¹⁻⁵ There usually are no significant differences in clinical manifestations as compared with vaginal cancer. The most frequent complaint in these patients is bleeding; pain and discharge are more likely to be associated with an ulcerated mass presentation, which is uncommon. Therefore it seems worth reporting a case of vaginal non-Hodgkin lymphoma with primary bulky ulcerated mass recently observed in our center.

Case report

A 55-year-old female presented in November, 1991, after 2 months of vaginal discharge and bleeding. Medical history did not reveal any sig-

nificant abnormalities, except hypertension (still on therapy and well controlled) and total hysterectomy plus bilateral salpingo-oophorectomy 8 years earlier, owing to multiple uterine fibromas.

Baseline physical examination did not show any tumor-related sign outside the genital tract. Local examination, palpation and colposcopy showed a circular, ulcerated, exophytic bleeding mass measuring about 5 cm in maximum diameter, located in the upper third of the vaginal stump and protruding in the canal. The vaginal walls appeared to be rigid and infiltrated. Multiple biopsies were performed and pathologic evaluation showed wide spread infiltration of the vaginal wall by large lymphoid cells with irregular and vesicular nuclei. A considerable amount of macrophages and plasma cells were also present.

These morphologic findings were in keeping with the hypothesis of a high-grade non-Hodgkin lymphoma, so immunocytochemical staining was promptly performed.

The neoplastic cells displayed a CD45⁺, CD20⁺, CAM5.2⁻, S100⁻, CDw75⁺, CD45R/MT2⁺

phenotype. A definitive diagnosis of centrocytic-follicular, predominantly large cell malignant lymphoma with rather high proliferation rate (PCNA 30-40%), group D according to the Working Formulation was established.

Genotypic examination confirmed the presence of a monoclonal B-cell lymphoid population. As far as laboratory tests are concerned, increased ESR (43 mm/h) and IgM (342 mg/dL) values emerged.

Clinical staging of the tumor consisted of physical examination, chest X-rays, CT scans of the thorax, abdomen and pelvis, abdominal and pelvic echography and bone marrow biopsies. CT scans did not show retroperitoneal nodal involvement nor any extension of the lymphoma beyond the vaginal wall. There were no signs of supradiaphragmatic disease. Therefore the final staging was IE.

Despite the limited clinical stage, the intermediate grading and the local bulky presentation and extension led us to choose aggressive chemotherapy as first-line treatment, to be followed by radiation therapy.

The ProMACE/CytaBOM schedule was adopted and at least six cycles planned. After 3 cycles CT scans and colposcopy showed partial remission of the disease. After 6 cycles a complete clinical response was achieved. Random vaginal biopsies confirmed local pathological remission. Treatment was completed with radiation therapy, which consisted of 3600 cGy to the whole pelvis and the para-aortic area through anterior-posterior opposite fields, plus a 1000 cGy moving technique boost to the vaginal stump. Both courses were given with standard fractionation.

No hematologic toxicity was observed during chemotherapy, thus 100% delivery of the planned dose without any delay was possible. Grade II leukopenia occurred during irradiation, but treatment was never discontinued. Non hematologic toxicity from chemotherapy was represented by grade III hair loss and grade II stomatitis. Nausea and grade II vaginitis and proctitis were experienced during irradiation.

The patient is still in complete clinical remission 11 months after the end of treatment.

Discussion

Primitive lymphomas are unexpected diseases in the female genital tract, particularly in the uterus and vagina; therefore they are likely to be misdiagnosed as either inflammatory diseases or other types of malignancies.

The histology of these lymphomas is similar to that observed in primary nodal presentation, but subtypes with a partially or predominantly follicular pattern, thought to be rare in extranodal sites, represent a substantial number of all the case series in the literature.^{6,7}

The prognosis of uterine and vaginal lymphomas is relatively good, particularly when compared to ovarian lymphoma: overall survival is over 70%, which is fully comparable with other extranodal presentations and far better than that of ovarian localization (30%).

In particular, stage IE vaginal lymphomas reach 80% 5-year survival. Treatment must be selected after carefully managed staging procedures. Most of the cases present with localized disease, thus high cure rates are achievable with surgery±radiotherapy.

Nevertheless, chemotherapy has to be considered on the basis of the pathological features and clinical presentation (local "bulky" extension, etc.) or when the extent of surgery and radiotherapy has to be minimized.

References

1. Bagella MP, Fadda G, Cherchi PL. Non Hodgkin lymphoma: a rare primary vulvar localization. *Eur J Gynaecol Oncol* 1990; 11(Suppl 2):153-6.
2. Canevari A, Brusamolino E, Inverardi D, et al. Linfomi maligni extralinfonodali primari. *Progressi in Ematologia Clinica* 1990; 9:303-10.
3. Liang R, Chiu E, Loke SL. Non Hodgkin's lymphomas involving the female genital tract. *Hematol Oncol* 1990; 8(Suppl 5):295-9.
4. Prevot S, Hugol D, Audouin J, et al. Primary non Hodgkin's lymphoma of the vagina. Report of 3 cases with review of the literature. *Pathol Res Pract* 1992; 188(Suppl 1-2):78-85.
5. Perren T, Farrant M, McCarthy K, Harper P, Wiltshaw E. Lymphomas of the cervix and upper vagina: a report of five cases and a review of the literature. *Gynaecol Oncol* 1992; 44(Suppl 1):87-95.
6. Harris NL, Scully RE. Malignant lymphoma and granulocytic sarcoma of the uterus and vagina. A clinicopathologic analysis of 27 cases. *Cancer* 1984; 53:2530-45.
7. Aguiar RC, Pozzi DH, Chamone DA. Granulocytic sarcoma of the ovary in a nonleukemic patient. *Haematologica* 1993; 78:53-5.