Case Report



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A Very Rare Case of Primary Non-Hodgkin's Lymphoma of the Vagina: Diagnose and Survivability

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Abstract

Non-Hodgkin's lymphomas are with high mortality rate and usually affect elderly patients. It's very rare for those lymphomas to originate from the female reproductive system and especially the vagina. We present a case of a 71 years old patient with genital bleeding, diagnosed with non-Hodgkin's lymphoma, who underwent standard treatment for that disease.

Keywords

Primary non-hodgkin's lymphoma; Vagina; Diagnosis; Treatment

Introduction

Non-Hodgkin's lymphomas (NHL) are a group of malignant blood diseases with unclear etiology. Risk factors include: episodical or persistent immunosuppressive condition, defects in normal cell proliferation, chronic antigenic stimulation, leading to an autoimmune condition: viral infection, alergic or inflammatory agent. A non-Hodgkin's lymphoma is considered primary when it is found in one or more organs only in the female genital tract; no atypical cells are found in the peripheral blood and bone marrow and for a period of 6 months after diagnosing no localizations in other organs are found [1].

Clinical Case

Patient is a 71 years old woman, in menopause for 20 years. Accompanying diseases include arterial hypertension and emphysema. Patient has never been operated and has given birth twice. Hospitalized in the department of oncogynecology because of a genital bleeding for 10 days. Patient reports of a light weight reduction during the last few months.

During the gynecology exam we found a normal gynecological statusexcept few polypose lesions on the anterior, lateral and posterior wall of the vagina, sizes from 1/1 to 1/2 cm (Figure 1).

After a standart preoperative preparation a dilatation and curettage (D&C) was performed and biopsy from the vagina was taken with histological results: cavum uteri – endometrial polyp with atrophic and cystic changes, vagina – malignant lymphoma.

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Figure 1: Lesion in the vagina.

Immunophenotyping with immunohistochemistryproved, that it is a difuse B-cell lymphoma with a high rate of malignancy.

After receiving the histology results, the patient was sent to the clinic of Haematology, where it was discovered, that it is indeed a primary non-Hodgkin's lymphoma of the vagina (after performing whole body CT scan and revision of the histology results) and were performed 7 courses of polychemotherapy with cyclophosphamide, pharmorubicin, vincristine and methylprednisolone; MabThera was not applied because of positive markers for hepatitis (HBsAg⁺). After finishing the treatment, the patient has no signs of the disease for 1 year; then a persistent cough is reported. The performed full body CT scan shows mediastinal lymphadenopathy, connected to the primary disease. No local recidive is found by the gynecology exam. Four courses of CVP were performed (cyclophosphamide, vincristine and prednisone). Despite the treatment the patient passed away due to progression of the primary disease 28 months after diagnose, with no data of a local recidive.

Discussion

Non-Hodgkin's lymphomas are a heterogeneous group of malignant diseases and originate from a different group of cells in the lymphoid tissue. They are ranked 5thin incidence among different tumors – 6/100000. They affect people with average age of 64. NHL are separated in 2 groups B and T-cell, and B-cell are more common – 35% of all NHL. They are aggressive, but 50% of cases can be treated with chemotherapy. In 2/3 of the cases they are with nodal presentation, 1/3 with extranodal presentation [2]; with increase of age the percentage of extranodal presentations increase, most commonly: stomach, tonsils, central nervous system, skin, bone, mammary gland. The Ann Arbor system is used for staging, while for risk stratification we use the International Prognostic Index.

Stage I

Involvement of a single lymphatic site (i.e., nodal region, Waldeyer's ring, thymus, or spleen) (I); or localized involvement of a single extralymphatic organ or site in the absence of any lymph node involvement (IE).

Stage II

Involvement of two or more lymph node regions on the same side of the diaphragm (II); or localized involvement of a single

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extralymphatic organ or site in association with regional lymph node involvement with or without involvement of other lymph node regions on the same side of the diaphragm (IIE).

Stage III

Involvement of lymph node regions on both sides of the diaphragm (III), which also may be accompanied by extralymphatic extension in association with adjacent lymph node involvement (IIIE) or by involvement of the spleen (IIIS) or both (IIIE,S).

Stage IV

Diffuse or disseminated involvement of one or more extralymphatic organs, with or without associated lymph node involvement; or isolated extralymphatic organ involvement in the absence of adjacent regional lymph node involvement, but in conjunction with disease in distant site(s). Stage IV includes any involvement of the liver or bone marrow, lungs (other than by direct extension from another site), or cerebrospinal fluid.

The presence or lack of clinical symptoms must be marked with the following signs after each stage:

A - No symptoms.

B - Fever (temperature >38°C), drenching night sweats, unexplained loss of >10% of body weight within the preceding 6 months.

E - Involvement of a single extranodal site that is contiguous or proximal to the known nodal site.

S - Splenic involvement.

In our case there was only weight reduction, but it was insignificant.

In around 40% of cases of NHL the female reproductive system is affected through themetastasis mechanism [3]. Primary extranodal lymphomas of the female genitalia are around 2% of all cases, the ovaries being the most common localization, while the vagina, cervix and uterus are affected in 1 out of 175 cases [4].

Despite the extremely low incidence of that disease, we can say, that the clinical presentation is non-specific and includes the presence of a tumor formation, genital bleeding, vaginal discomfort, dyspareunia, pelvic pain [5].

In the differential diagnosis we have to consider a sarcoma, low differentiated carcinoma, neuroendocrine tumors and chronical infection [6]. The survivability of those patients depends on the disease stage.

Conclusion

Despite the rarity of primary NHL of the female reproductive system, we can expect their incidence to increase, based on the increase of incidence of extranodal NHL in general. That's why clinicists and pathologists will have to keep that in mind when they encounter patients with abnormal genital bleeding.

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