Non-Hodgkin's Lymphoma Involving the Vulva and Vagina: A Clinicopathologic and Immunophenotypic Feature of a Case

VULVA-VAGİNAL NON-HODGKİN LENFOMA: KLİNİKOPATOLOJİK VE İMMUNOFENOTİPİK ÖZELLİKLERİ

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Abstract

A 65 year old woman presented with vaginal bleeding and with history of a mass in clitoris since 6 months. Biopsy of the mass which in clitoris was performed and then, the definitive histological diagnosis showed that the tumor was an extranodal diffüse large B cell non-Hodgkin lymphoma.

A primary vulvar localization of malignant lymphomas is quite unusual. There was no difference in clinical manifestations compared between cancer of the vulva and vagina. Because of this non Hodgking's lymphoma should be considered in patient with vulvo-vaginal mass.

Key Words: Vulva, vagina, lymphoma, non-Hodgkin

Özet

65 yaşında kadın hasta 6 aydır var olan klitoris bölgesinde kitle ve vaginal kanama şikayeti ile hastanemize başvurdu. Klitoris bölgesindeki kitleden yapılan biyopsi sonucu diffüz büyük B hücreli non-Hodgkin lenfoma olarak değerlendirildi.

Malign lenfomaların primer vulvar lokalizasyonu oldukça nadirdir. Vulva/vagina kanserleri ile karşılaştırıldığında klinik olarak herhangi bir fark yoktur. Bu nedenle vulva-vaginal kitlesi olan hastalarda non-Hodgkin lenfoma düşünülmelidir.

Anahtar Kelimeler: Non-Hodgkin lenfoma, vulva, vagina

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Malignant lymphoma is a rare entity in the neoplasms of the female genital tract. It can be seen in female genital tract during advanced stage of systemic disease but a primary localization is quite unusual.[1] Also the distinction between primary extranodal lymphoma and secondary involvement of an extranodal site by a lymphoma originating from lymphoid organ is not always possible. non-Hodgkin's lymphomas (NHL) represent 70-80% of all lymphomas in female genital tract.[2] The most common genital localizations are the cervix, the uterine corpus and the ovary. The vulva seems to be involved least often. There are very few cases of NHL involving the vulva in the literature.[3] We describe the clinicopathologic and immunohistochemical features of a case of NHL involving the vulva.

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Case Report

A 65 year old, gravida 5 para 3, woman presented with history of a mass in clitoris since 6 months, and with vaginal bleeding. On vaginal examination there was a 2 cm mass around clitoris with pathologic involvement of left vaginal

wall and periüretral tissue. There was also an ulceratıon on left vaginal wall. Cervix was multipara and uterus and adnexes were normal. Biopsy of the mass which in clitoris was performed and firstly reported to be a indifferentiated carcinom. Then unexpectedly, the definitive histological diagnosis showed an extranodal diffüse large B cell non-Hodgkin lymphoma, immünoblastic variant according to the that the tumor was REAL Classification. The patient underwent several diagnostic procedures consisting of a ultrasound scan of the abdomen, bone marrow biopsy, all body syntigrafi, CT (computed tomography) scan of abdomen and thorax. In ultrasound scan there was a 4 cm mass under the bladder and in abdomen CT, there were 2 hypodens nodul between liver and kidney. Blood examination and thorax CT were normal. Patient didn't showed systemic symptoms such as fever, night sweats and weight loss. The stage of the disease according to the Ann Arbor Classification was stage 2 E. Lymphoma cells were negative for citokeratin, aktin, desmin, kromogranin and positive for LCA. Immunoblast like cells in lymphoma were positive for CD 10, CD 20, CD 38 and CD 79; they were negative for CD3, CD 30, HMB45. The patient received 6 cycles of CHOP (cyclophosphamide, doxorubicin, vincristine and prednisone) polychemotherapy. Unfortunately her condition has rapidly deteriorated during the 5 months since her admission. She died at 5 months after her admission, just after her sixth cycle of chemotherapy.

Discussion

Malignant lymphomas involve the female genital tract in 30% of cases. The majority (more than 90%) are non-Hodgkin lymphoma, involving in order of frequency, ovary (49%), uterus (29%), fallopian tubes (11%), vagina (7%), vulva (4%).[4] So a primary vulvar localization seems to be quite unusual. There was no difference in clinical manifestation compared betwieen cancer of the vulva and vagina. The diagnosis was made using histological and immunohistochemical methods. The histologic differential diagnosis of vulvar NHL includes Langerhans'cell histiocytosis, lympho epithelioma like carcinoma, granulocytic sarcoma or chloroma, small cell neuroendocrine carcinoma, malignant mullerian mixed tumor, small cell variant of melanoma, extraosseous Ewing's sarcoma/primitive neuroectodermal tumor, poorly differentiated squamous cell carcinoma, Merkel cell carcinoma, epithelioid leiomyosarcoma, small round cell type of malignant peripheral nerve sheath tumor and Bartholin's gland adenocarcinoma. Due to their infrequency, pathologists who are unfamiliar with its clinical and pathologic features may misdiagnose NHL.[3] Delayed diagnosis and inappropriate management of lymphomas in the vulva may lead to widely destructive disease of the lower female genital tract. All female patients with pelvic lymphoma should undergo careful staging with conventional chest radiographs, CT scans, and bone marrow biopsy.[5] Primary vulvar NHLs most often present as a vulvar mass, are clinically aggressive, and pathologically most often diffuse large B-cell lymphoma (DLBCL). Most patients are in their third to sixth decade of life.[6] Because of the rarity of this cancer, there are no consensus in management of extranodal genital lymphomas. Chemotherapy, radiotherapy and surgery are the treatment options. In this case which was stage 2E, we performed chemotherapy treatment. Because according to the literature, radiotherapy plays an important role in the treatment of patients in stages IE but it is quite ineffective in the case of more advanced stages.[7] Recently, neoadjuvant chemotherapy has been proposed instead of radiotherapy in genital lymphomas.[2] A complete response to chemotherapy does not require additional therapies and radiotherapy and surgery could be restricted as a second line treatment for incomplete responses and local relapses.[2]

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