Uncommon disease:
A case report of Stage III invasive Paget's disease of the vulva

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This patient is a 57-year-old female who underwent radical vulvectomy and bilateral inguinal lymph node dissection followed by radiotherapy for Stage III invasive the Paget's disease of vulvar cancer. She was free of disease 2 years after primary therapy.

Key Words: *Vulvar cancer, Paget's disease,*

Nadir bir olgu: Vulvanın stage III invazif Paget hastalığı

Paget hastalığı, stage III vulva kanseri olup radikal vulvektomi ve bilateral inguinal lenf nodu diseksiyonunu takiben radyoterapi uygulanan ve primer tedavisinden sonra 2 yıldır hiçbir yakınması olmayan 57 yaşındaki bir olgu sunuldu.

Anahtar kelimeler: *Vulva kanseri, Paget hastalığı*

Paget's disease of the vulva is an uncommon entity that appears as a sharply demarcated, edematous area, hyperemic areas with a superficial white coating to give the impression of "cake-icing effect".(1) This finding is almost pathognomonic for Paget's disease. The first case of Paget's disease of the vulva was reported by W. Dubrewilh (2). To date approximately 200 additional the Paget's disease of the vulva have been added to the world's literature. Invasive the Paget's disease of the vulva has been seen much less than intraepithelial lesions. In this report, we present a patient with stage III Paget's disease of the vulva who underwent radical vulvectomy and bilateral inguinal lymphadenectomy followed by radiotherapy. She had no evidence of disease 2 years after primary therapy.

CASE: A 57-year-old nullipar patient who had vulvar lesion was referred to M.D. Anderson Cancer Center. On admission time to hospital, she was alert, well-developed, obese, white female in no acute distress. Pelvic examination revealed an extensive beefy, red and white lesion involving the entire right labium majus and minus with extension around the clitoris. The margin of this lesion extended to the hymeneal rim medially and to the edge of the perineal body, inferiorly. Colposcopic examination of this lesion revealed coarse punctation with markedly abnormal vasculature, and islands of white epithelium with an irregular topography. The urethral meatus and vagina were clear without lesions or nodularity. The cervix was grossly normal appearance. Rectovaginal examination revealed normal uterus and no pelvic masses. Vulvar biopsies were taken. Pathologic report revealed invasive Paget's disease of the vulva. Radical vulvectomy and bilateral superficial and deep inguinal lymph node dissection was performed. The final pathology report revealed invasive Paget's

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disease with right inguinal lymph node metastases.

She received postoperative 45 Gy tumor dose through anterior and posterior portals, anteriorly with cobalt 60, posteriorly with 25 MEV photons over 5 weeks. (total dose was 60 Gy). The patients has no evidence of disease 2 years after primary therapy.

DISCUSSION

The Paget's disease of the vulva is extremely indolent form of epithelial neoplasia which occurs in women in the seventh decade of life, but can be seen young women.(1) Symptoms may have been present from months to years. The most common site of lesion is the labium majus. Histologically the lesion is characterized by a nest of clear cells (Paget's cells) in adenomatoid patterns noted initially in the basal layers of the epidermis but later found about skin appendages as well as throughout the entire thickness of the epithelium.(3) The lesion can be differentiated from squamous cell cancer and melanoma by differential staining techniques.

Treatment of the Paget's disease of the vulva is usually wide local excision, because the disease almost always extends well beyond the gross lesion.(1,4) After surgery, recurrences are common when the surgical margin contain neoplastic cells. These new lesions were treated by wide local excision and if required full-thickness skin graft is used. However, proper therapy for the Paget's disease of the vulva with invasion is a more difficult problem. Patients in whom an underlying adenocarcinoma is present should be treated with more radical procedure such as radical vulvectomy and inguinal lymph node dissection. (4) If the lymph nodes are positive for cancer, the prognosis is very poor. According to limited experience, the role of radiotherapy is unclear. Metastatic lesions in the pelvis of some patients showed little objective response to a dose of 3000 rads given as palliation for pain., and some invasive recurrences regressed after radiation therapy.(4)

Figure 1. Invasive Paget's disease of the vulva

Our patient had inguinal nodes positive for metastatic cancer, therefore received radiotherapy 60 Gy, and had no evidence disease for 2 years. In our experience, underlying extensive invasive adenocarcinoma with positive groin lymph nodes, radical wide excision and appropriate lymph node dissection followed by irradiation therapy is excellent therapeutic alternative.

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