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Case report

HDR-plesiotherapy for the treatment of anogenital extramammary Paget's disease

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ABSTRACT

Background: Extramammary Paget disease (EMPD) is a rare condition that most commonly affects the anogenital region in the elderly. It may be associated with an underlying invasive carcinoma. Surgical excision is nowadays considered a standard treatment for extramammary Paget's disease, although this approach might not be suitable for all patients. Good rates of local control and cosmetic outcome have been achieved by using high-dose-rate (HDR) plesiotherapy in the treatment of skin tumors arising in different locations.

Material and methods: We present the results observed in a patient with EMPD treated by HDR plesiotherapy with a custom-made mold up to a final dose of 54 Gy in 12 fractions of 450 cGy over 4 weeks.

Results: After a follow-up of 18 months, the patient is alive and without evidence of local or distant relapse. Acute toxicity was acceptable and cosmetic result was considered as excellent.

Conclusion: This technique represents an attractive alternative for those patients who are not candidates for surgical procedures because of unacceptable risk of disfigurements or functional impairment, medical contraindications or patient preference.

1. Background

Paget's disease (PD) is a rare type of intraepithelial adenocarcinoma characterized by the presence of malignant Paget's cells lying within the epidermis of the skin, first described by Sir James Paget in 1874.[1] Extramammary Paget's disease (EMPD) expresses the occurrence of this entity in areas containing apocrine glands such as the perianal region, vulva, penis, scrotum, perineum and axilla, different from the breast nipple. The first case of EMPD of the anogenital area was reported by Crocker in 1888, and until 2010 almost 300 cases have been published.[2–4] The disease sites present as patchy and well-circumscribed, erythematous eczema or erosions together with itching, bleeding or burning.[5] The prognosis of simple EMPD is relatively favorable, reaching disease-free and overall survival rates of above 70% at 5-year after curative intention treatment.[6] Wide surgical excision remains to be the mainstream of therapy for vulvar Paget's disease. However, local recurrence rates range from 12% to 58% of treated

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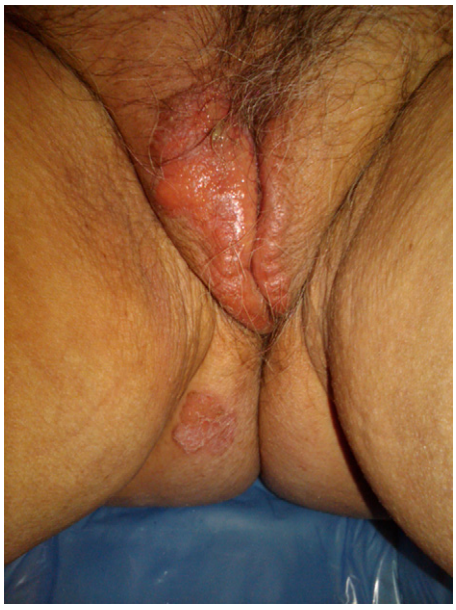

Fig. 1 – Patient with two biopsied lesions of EMPD located in the right labium majus and on perianal skin.

Fig. 2 – Patient with the custom-made mold in place for plesiotherapy during irradiation.

women during the follow-up time, even after extensive local resections.[7–9] On the other hand, radical surgery morbidity can be high, especially in the elderly. Radiotherapy has been used only occasionally in this condition. Primary radiotherapy was indicated in patients not suitable for surgery because of the presence of irresectable tumors or co-morbidities precluding radical surgery. Radiotherapy was also used occasionally in the adjuvant setting following surgery or with palliative intention after recurrent disease.[10,11] Recently, a curative role for radiation therapy of EMPD has been proposed.[12] We present a case of vulvar and perianal Paget's disease treated by HDR brachytherapy with a custom made mold (plesiotherapy) with curative intention.

2. Case report

A 79-year-old woman presented with a 2-year history of redness and pruritus in the vulvar and perianal region. Clinical examination revealed two well-circumscribed erythematous plaques with some scattered whitish points on her right labium majus and on perianal skin (Fig. 1). No enlarged inguinal lymph nodes were detected. Both areas were biopsied evidencing the presence of multiple Paget cells, forming irregular nests and scattering individually in a pagetoid pattern in the epidermis, leading to the diagnoses of extramammary Paget's disease. Additional exams, including whole-body CT, colonoscopy and gynecological examination did not reveal an underlying malignancy. The patient was treated initially by means of topical applications of imiquimod cream but in the absence of response she was referred to the Radiation Oncology Department. A treatment by means of HDR superficial brachytherapy (plesiotherapy) with a custom-made mold encompassing both lesions with a security margin of at least 2 cm surrounding the visible disease was proposed and the patient gave her consent to the treatment.

Our technique of HDR plesiotherapy based upon custom-made molds has been previously described.[13] Briefly, personalized molds consist of a flexible pad of silicone rubber, 10 mm thick, with a customizable array of 6F plastic catheters embedded separated by 1 cm that easily conforms to the shape of surfaces. The number of plastic tubes and the separation between them depended on the size of the area to be treated. This area was defined by the radiation oncologist being wide enough to include an adequate margin of uninvolved skin of at least 1 cm around the tumor or the surgical bed. A CT scan carrying the mold with wire dummies inserted on the plastic tubes were obtained from each patient at 1-mm-thin intervals. The macroscopic tumor/surgical tumor bed with a margin (planning target volume (PTV)), along with the surrounding organs at risk were contoured on these CT slices before calculation. PTV dose distribution and additional treatment parameters were calculated by using the 3D treatment planning software PLATO developed by Nucletron (Nucletron BV, Veenendaal, the Netherlands). Treatment dose was prescribed at 5 mm depth from the surface of the mold. At the moment of treatment, the personalized mold was applied to the tumor lesion, placed in direct apposition to the skin of the involved area, and kept in place by adhesive tape (Fig. 2). The mold was then connected to a HDR equipment containing a 192-iridium radioactive source (Microselectron Nucletron BV, Veenendal, the Netherlands). Radiation treatment was administrated in 12 fractions of 450 cGy, 3 fractions a week over 4 weeks, until a total dose of 54 Gy (equivalent dose in 2-Gy fractions (EQD2): 65.3 Gy) was reached. Adverse effects of plesiotherapy included acute grade 3 epithelitis on the treated areas that resolved with topical measures 2 weeks after treatment cessation. Postradiotherapy evaluation revealed a complete disappearance of the Paget's disease areas 3 months after discharge. On the last follow-up at 18 months after plesiotherapy,