

## A Case of Extramammary Paget's Disease Resistant to Photodynamic Therapy and Surgery Successfully Treated with Radiotherapy

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Dear Editor:

Although surgery is widely accepted as the first-line treatment for extramammary Paget's disease (EMPD), it cannot always be accomplished due to its frequent involvement of areas with important function<sup>1</sup>. Nonsurgical treatments including radiotherapy (RTx) and photodynamic therapy (PDT) are considered in such cases. We report a case of EMPD refractory to PDT and surgery that was successfully treated with RTx.

A 68-year-old man presented with a well-defined erythematous patch with ulceration on the left side of scrotum,

which enlarged during the last 7 months (Fig. 1A). Skin biopsy revealed numerous Paget cells (Fig. 2). Possible accompanying internal neoplasms were discounted through multiple means, ranging from physical examination to positron emission tomography/computed tomography. Diagnosed as localized EMPD, the patient was primarily treated with PDT because he strongly requested preservation of his genitourinary functions. Methyl aminolevulinate cream (Metvix; Galderma, Fortworth, TX, USA) was applied occlusively and was maintained for 3 h. The lesion was then irradiated with a 630-nm light-emitting di-

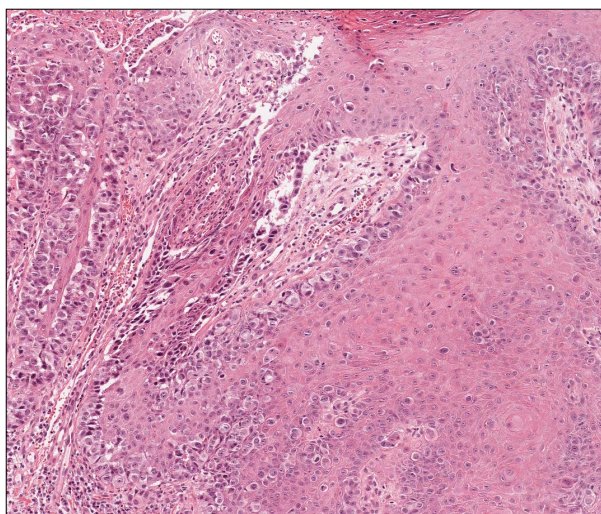


**Fig. 1.** (A) Patient image at initial visit. (B) The patient revisited our clinic suspecting of recurrence. The biopsy of skin lesion did not show any tumor cells.

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**Fig. 2.** Skin biopsy showing acanthotic epidermis with numerous Paget cells of large nucleus and eosinophilic cytoplasm (H&E,  $\times 200$ ).

ode (Aktilite CL128; Photocure, Oslo, Norway). After two sessions, the patient showed a partial response and underwent local excision. He was treated with 15 sessions of PDT for possible remnants. As Paget cells were still seen on follow-up biopsy of the surgical margin, he underwent a three-stage Mohs surgery and received 11 additional sessions of PDT. Nonetheless, the lesion persisted clinically. We referred him to the radiation-oncology clinic. Radiation was administered by using an electron beam of 6 MeV, at 200~250 cGy per day, 4 days a week, until a total dose of 37 Gy was reached. He received subsequent RTx with a total dose of 30 Gy in same manner. The lesion disappeared completely. After 18 months, the patient revisited our clinic with an erythematous patch on the scrotum (Fig. 1B). Biopsy only revealed telangiectatic dermal vessels.

A systematic review of clinical studies examining the use of PDT for EMPD showed that of 133 EMPD lesions, 77 exhibited complete response and 52 exhibited partial response with satisfactory cosmetic outcome in all patients<sup>2</sup>. However, the durability of this response remains unclear,

as the follow-up periods were typically less than a year<sup>2</sup>. In a long-term follow-up study of EMPD *in situ* for 24 months, the recurrence rate was 50%. The authors stated that PDT should be recommended only for cases of scrotal lesions <4 cm in diameter<sup>3</sup>. Larger lesions showed an unfavorable response presumably because of the denser infiltration of Paget cells and more adnexal involvement, followed by disproportionate protoporphyrin IX accumulation in the glands resulting in photodynamic oxygen depletion.

In the present case, the lesion was much larger than 4 cm, which may have been responsible for the ineffectiveness of PDT. The optimal RTx dose is not standardized; however, a total of 40~50 Gy has been considered suitable for primary treatment<sup>4</sup>. The lesion of our case was refractory to other therapies; hence, a higher dose of 67 Gy was chosen.

In conclusion, EMPD lesions resistant to PDT and surgery may be significantly resolved with RTx; thus, it should be considered a treatment option in cases of repeatedly unsuccessful PDT and surgery.

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