Coexistence of Invasive Inguinal Extramammary Paget Disease and Recurrent Intertrigo: A Case Report

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Introduction

An 89-year-old male presented with recurrent bilateral inguinal intertrigo that intermittently responded to topical antifungals over the past 2 years. A few months ago, a nodular lesion with ulceration developed in the right groin at the site of intertrigo. Subsequent shave biopsy confirmed the diagnosis of invasive Extramammary Paget Disease (EMPD). Further workups were negative for metastasis or internal malignancy. The lesion was improved after 6 weeks of topical imiquimod. We report this case of coexistence of invasive inguinal Extramammary Paget Disease and recurrent intertrigo and discuss related topics below.

Case Description

An 89-year-old Caucasian male with history of meningioma, benign prostate hyperplasia, visited our clinic 2 years ago, complaining about pruritus in the groin area. There were erythematous plaques spanning in the bilateral groin extending to the medial thighs with sharp demarcation without scaling, which was consistent with intertrigo. The rash improved with miconazole cream but recurred 6 months later. It resolved completely with another 2-month use of nystatin-triamcinolone cream.

Early this year, the patient visited us again with persistent bilateral rashes in the groin refractory to cream. Miconazole powder was prescribed, with instructions to keep the affected area clean and dry. At the follow up visit 3 months later, a new nodular lesion with ulceration developed in the right groin over the intertrigo. Physical exam revealed a well demarcated, erythematous, and eczematous plaque with a nodular lesion and ulceration on the right groin extending to the scrotum [Fig. 1], as well as a partially treated erythematous rash noted on the left side. Shave biopsy of the nodular lesion was performed. The pathology reports invasive EMPD extending to deep and peripheral edges. Both epidermis and dermis were involved by a nested, epithelioid neoplasm with single cells including pagetoid spread. Further, a subset of neoplastic cells exhibits vacuolated and mucinous features. Immunohistochemical stains are positive for cytokeratin CAM 5.2, CEA, CK7 and p63 (focal), GATA3 (diffuse), androgen receptor (multifocal), Nkx3.1 (focal), and p40 (focal), and negative for CK20, PSA, PSAP, and SOX10.

Cystoscopy and sigmoidoscopy were negative for malignancies. PET CT scan revealed curvilinear F-18 FDG activity measuring 2.8 cm x 0.6 cm with SUV max of 5.2 at the interface of the right scrotal sac and extreme medial right inguinal region and upper right thigh. No definite metastatic disease was identified. Surgery was recommended, but he would require a rather extensive skin graft. He opted for imiquimod 5% cream starting with 3 times weekly and titrated it himself as tolerated. After 6 weeks of treatment, improvement of the lesion was observed.

Discussion

EMPD is a rare dermatologic neoplastic disease arising from apocrine gland-rich skin. It tends to occur in Caucasian females or Asian males aged between 60 and 80 years old. The most common sites are the vulva and penis, while only 2.1% of the patients presents in groin. Primary EMPD can be classified as intraepidermal EMPD or invasive EMPD if the dermis is involved. Secondary EMPD is associated with underlying adenocarcinoma with epidermal invasion. EMPD has a variety of presentations including erythema, nodules, ulceration, dyspigmentation, eczematous appearance. Differential diagnosis includes contact dermatitis, intertrigo, candidiasis, tinea curis, seborrheic dermatitis, lichen sclerosus, bowen disease, etc.

EMPD is diagnosed by histopathological findings of vacuolated Paget cells in the epidermis, with nest-like patterns of gland-like structure. A diagnostic immunohistochemical panel for EMPD consisting of CK7-positive, CK20-positive or CK20-negative, p63-negative, SOX10-negative, and CEA-positive results is recommended. Pathology-confirmed EMPD requires age-appropriate and anatomical location-directed screening including colonoscopy, cystoscopy, to identify secondary EMPD.

Surgical removal is commonly performed for primary EMPD. Photodynamic, imiquimod or laser therapy may be options for nonsurgical candidates. Radiotherapy may be used postoperatively or used for nonsurgical candidates with curative intent. For metastatic disease, chemotherapy, targeted therapy, or immune checkpoint inhibitors may be considered. Post-surgery surveillance is recommended to monitor for recurrence and adverse effects, but there is no well-established method of surveillance.

In our case, the patient’s erythematous rash initially responded well to antifungals, which anchored our diagnosis to intertrigo. At follow-up visits, a newly developed malignancy was masked until it showed nodular lesions. He works at an auto body shop and denies family history of cancer or tobacco or alcohol use. It is unclear whether recurrent intertrigo, a chronic inflammatory skin condition, could increase the risk of EMPD. With the case we present above, we would like to emphasize the necessity of a careful skin examination and extra cautions for malignancy in the setting of recurrent intertrigo.

Conclusions

In conclusion, diagnosis of EMPD is challenging when it coexists with recurrent intertrigo. EMPD is commonly misdiagnosed due to its rarity and shared features with other dermatologic conditions. Chances of developing skin malignancy together with recurrent intertrigo should not be neglected. Biopsy should be performed as soon as possible for any refractory lesions or atypical features including nodularity.

References
