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Case Report

Psoriasis with extramammary paget disease in a male: a case report

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Abstract: Psoriasis is a chronic inflammatory skin disease that is characterized by erythematous, sharply demarcated papules and plaques covered by scales. Extramammary Paget disease (EMPD) is a uncommon neoplastic condition of apocrine gland-bearing skin and its occurrence in combination with psoriasis is very rare. We report an interesting case of a 61-year-old male with extensive psoriasis presented with penoscrotal EMPD, which was confirmed by histopathological stain.

Keywords: Extramammary paget disease, psoriasis, male genitalia

Introduction

Psoriasis is a chronic inflammatory skin disease that is characterized by erythematous, sharply demarcated papules and plaques covered by scales [1]. Extramammary Paget disease (EMPD) is a rare neoplastic condition of the skin or its underlying appendages commonly found in the vulva, perianal region, scrotum, penis, and axilla [2]. Clinically the condition presents as a well-demarcated, thickened, pruritic, erythematous, or white scaly plaque with irregular borders [3, 4]. Microscopically EMPD involves large cells with vacuolated cytoplasm and centrally located nuclei characterized as Paget cells [5]. The disease is categorized into primary or secondary EMPD with primary EMPD originating from intraepidermal cells and secondary EMPD coming from an underlying neoplasm [2, 5]. However, there is no report suggesting the association of psoriasis with EMPD, or psoriasis admixed with EMPD. In this paper, we present a case of extensive psoriasis in a male combined with penoscrotal EMPD.

Case description

A 61-year-old man was referred to us with a two-year history of a pruritic, painless erythematous skin rash on the penis and scrotum. Because of progressive pruritus, erythema, and thickening of the affected area, he sought medical attention. Treatment was topical steroid creams therapy, with an empiric diagnosis of psoriasis. Despite conventional treatment, the lesion had gradually increased in size and extended to the scrotum and penis. The patient initially underwent a punch biopsy and the pathology report came back as EMPD.

The patient had a history of psoriasis for about 30 years and a chronic erythematous dermatological condition, which had been controlled with long-term methotrexate treatment, systemic narrow band ultraviolet (UV)-B phototherapy and topical steroid therapy. The patient had several previous records and was required clinic appointments and adjustment of medication. His family history was negative for melanoma, colorectal cancer, or prostate cancer.

Examination was remarkable for extensive bilateral erytherodermic nodular lesions with plaques involving the extensor surfaces of elbows, forearms, thighs and knees. He also had scattered involvement over the abdomen, lower back and scalp. Approximately 15~25% of his skin surface was covered with lesions. As shown in **Figure 1**, focal examination revealed an erythematous plaque extending over the base of the penis and 2-cm round peripheral rim onto the scrotum. The involved area con-

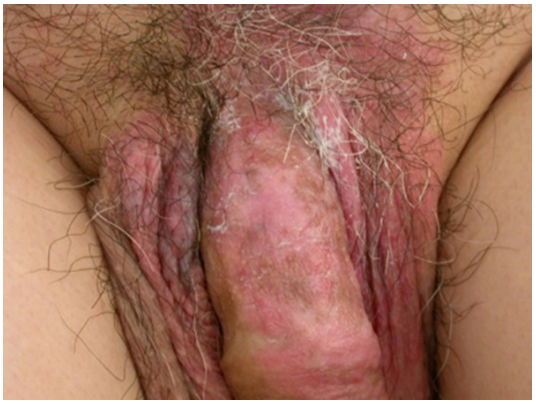

Figure 1. Scaly, crusting, erythematous patch on the pens and scrotum.

tained whitish superficial exudates, and was scaly. No inguinal lymph nodes were palpated on physical examination.

The patient was admitted to the department of oncology and had an extensive cancer work-up. The tumor markers, routine colonoscopy and esophagogastroduodenoscopy were performed to check for internal malignancy of the lower gastrointestinal track. However, there was no evidence of an underlying internal malignancy. A whole-body positron emission tomography (PET)/CT scan did not find other abnormal hypermetabolic lesion.

After the thorough physical check-up, the patient underwent surgical wide excision of an involved lesion of the scrotum and penis and the specimens were sent to the department of pathology for analysis. The skin defect was covered with a scrotal flap and split-thickness skin graft for the penile shaft. All of the surgical safety margins were clear, and there was no evidence of metastasis in the abdomen or pelvis. The patient was followed up in the outpatient department for 10 years and has had no evidence of recurrence or other complications so far.

As shown in **Figure 2A**, the histopathological examination showed large, round cells in the epidermis, which were scattered discretely or grouped forming nests in the epidermis. As shown in **Figure 2B**, Immunohistochemical tests showed that the tumor cells were positive for cytokeratin 7 (CK7), epithelial membrane antigen (EMA), and C-erbB2. The cytoplasm of the tumor cells was positive for D-PAS staining. The tumor cells were negative for S-100, HMB-45, and Melan-A staining.

Discussion

Psoriasis is a common T helper type-1 and -17-mediated chronic inflammatory disease, affecting 2~3% of the general population [1, 6]. Multiple observational studies have demonstrated that patients with psoriasis, particularly those receiving systemic treatments or phototherapy, have higher incidences of systemic disease mortality independent of traditional risk factors for these outcomes [1, 6].

EMPD was first described by James Paget, and usually occurs at apocrine glands-rich areas, such as the penis, scrotum, vulva, perineum, and axillae of middle-aged and older individuals [2, 3]. It presents as reddish patches or gray-white plaques, which are eczematoid, crusty scaling, or papillomatous. It can be easily misdiagnosed as psoriasis, eczema, contact dermatitis, Bowen's disease, or tineacruris before histological examination, owing to their similar appearance and symptoms. For this reason, proper treatment may be delayed [2-5].

Presently, we report a rare case of psoriasis combined with EMPD. The clinical appearance was psoriasis, but the biopsy specimen showed Paget cells combined with EMPD. Unfortunately, even after two years of treatment our patient did not show significant signs of improvement from topical application of steroid cream and long-term methotrexate treatment. Thus, dermatologists should avoid the pitfalls through appropriate histopathological examination when the diagnosis is ambiguous.

The pathogenesis of EMPD has been debated. One theory postulates that EMPD results from an underlying sweat gland adenocarcinoma that spreads into the epidermis [2]. Another theory suggests malignant transformation of pluripotent dermocytes that are capable of glandular differentiation [3]. However, the exact mechanisms of psoriasis combined with EMPD are still unknown, like in our case. Those who support the multipotent stem-cell theory also suggest that chronic inflammatory skin disease may play an important role in the pathogenesis of EMPD. Therefore, further study about the connection between EMPD and psoriasis may be necessary.

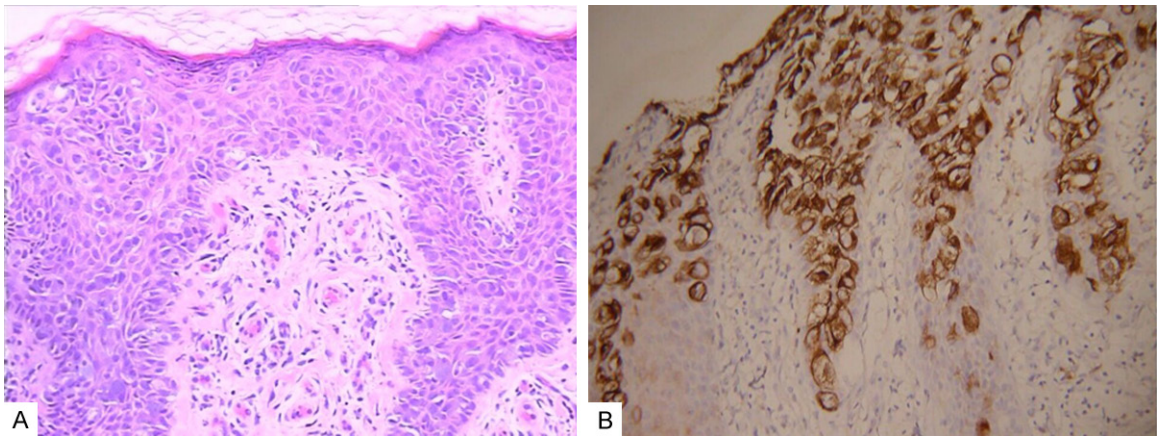

Figure 2. A. Histopathologic findings showed a tumor cell nest in the dermis and the epidermal cells with hyperchromatic and polymorphic nuclei, intraepithelial gland cells. HE, ×200. B. The tumor cells were positive for C-erbB2. SP, ×200.

Currently, surgical resection is the standard of care for EMPD and if selected it would be likely to involve wide local resection and reconstruction via various modalities such as a scrotal skin flap or a gracilis muscle flap [7]. However, there is a high rate of reoccurrence, the use of Mohs microsurgery or the modified peripheral Mohs technique have helped reduce the rate of reoccurrence [8].

Disclosure of conflict of interest

None.

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