A Notorious Plaque over the Groin

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Question

A 54-year-old male patient presented with an itchy raised lesion over the right groin for 5 years. The lesion was around the size of a peanut to begin and gradually extended to involve the groin and the medial aspect of the right thigh.

The physical examination revealed a single well-defined irregularly shaped erythematous plaque with maceration and hyperpigmented border over the right groin extending on to the medial aspect of the right thigh without any associated regional lymphadenopathy [Figure 1]. Histopathological examination of hematoxylin and eosin-stained tissue revealed hyperkeratosis, focal parakeratosis, irregular acanthosis with severe dysplastic changes in the epidermis. The clusters of large vacuolated cells having foamy cytoplasm with a pleomorphic hyperchromatic nucleus, focally forming tubules were noted [Figure 2]. Upper dermis showed dense lichenoid infiltration with melanin incontinence. Mild periadnexal inflammatory infiltration and severe dysplastic change with cytoplasmic vacuolation in the underlying eccrine glands were noticed [Figure 3].

Immunohistochemical evaluation of skin biopsy suggested cytokeratin (CK)-7, CK-20 positivity. Contrast-enhanced computed tomography scan of chest, abdomen, and pelvis showed no remarkable findings.

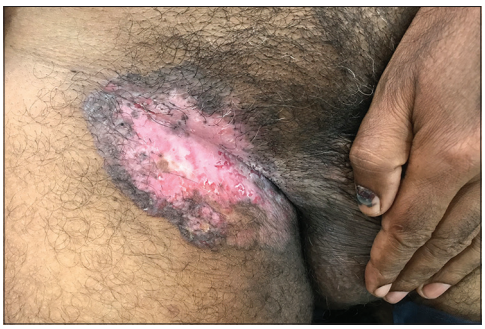

Figure 1: Irregularly shaped well-demarcated erythematous plaque with grayish-white crust over the right groin

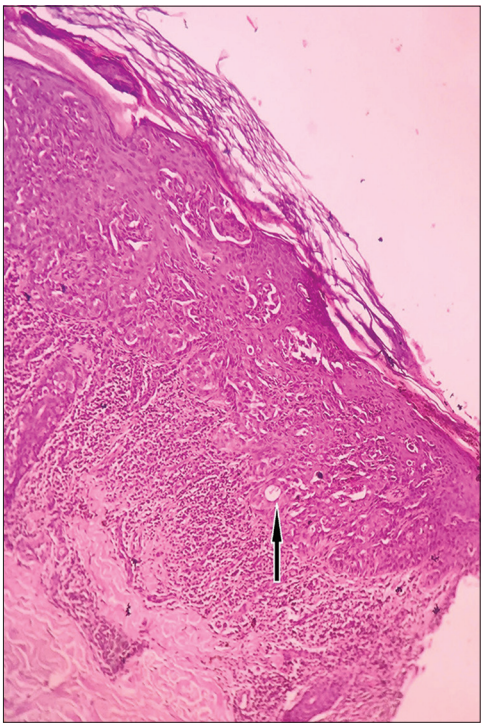

Figure 2: Clusters of large vacuolated cells with a pleomorphic hyperchromatic nucleus, focally forming tubules as shown by black arrow [H and E 100X]

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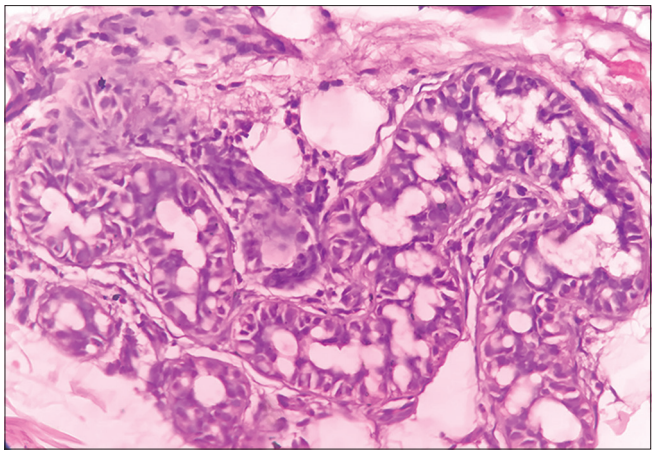

Figure 3: Severe dysplastic change with cytoplasmic vacuolation in the underlying eccrine glands [H and E 400X]

Answer

Extramammary Paget's Disease.

Discussion

Sir James Paget first described Paget's disease in 1874.[1] Extramammary Paget's disease (EMPD) is a rare cutaneous intraepithelial adenocarcinoma with a predilection to the apocrine gland bearing area. It typically involves the vulvar, perianal, perineal, scrotal, and penile regions.[2] Clinically, it presents as an erythematous to grayish-white asymptomatic, sharply demarcated plaques with secondary crusting, eczematous, scaly, or ulcerated changes.[3]

The diagnosis of EMPD is based on the identification of Paget's cells with prominent nuclei and abundant lightly stained cytoplasm on hematoxylin-eosin staining in skin biopsy.[2] The disease can arise from two major pathological mechanisms:[4] as an *in situ* intraepithelial adenocarcinoma which has the potential for local invasion and subsequent metastasis; and as a pagetoid spread of visceral malignancy.

Two distinct immunohistochemical subtypes have been identified: type I (endodermal) expresses an endodermal phenotype CK 7+, CK 20+, gross cystic disease fluid protein (GCDFP) 15− and may be associated with distant carcinomas whereas type II (cutaneous or ectodermal) expresses sweat gland markers CK 7+, CK 20−, GCDFP15+ and would be of cutaneous origin.[2]

EMPD is generally considered a disease with a good prognosis. Poor prognostic factors include dermal invasion, nodular skin lesions, lymph node involvement, and distant metastasis.[5]

Surgery is the standard treatment for EMPD, with an emphasis on obtaining negative margins to control the disease. Recent evidence suggests successful treatment with topical imiquimod 5% cream immunotherapy.[3]

Conclusion

In summary, patients with pruritic plaques in groin unresponsive to topical therapies should undergo biopsy for definitive diagnosis. Even though immunohistochemistry can help in establishing the risk of associated malignancy in EMPD, a detailed evaluation for internal malignancy is a must along with regular follow-up.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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