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CASE REPORT

A Rare Case Of Vulvar Paget's Disease Associated With Squamous Cell Carcinoma

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ABSTRACT

Primary cutaneous Paget's disease is an uncommon neoplasm usually of post-menopausal white women. In approximately 10-20% of women with vulvar Paget disease, there is an invasive component or an underlying skin appendage adenocarcinoma. Only a very few case reports in literature have specifically identified proliferative squamous cell component in Paget's disease. This histological change may be misinterpreted as a primary squamous pathology if the Paget's component is not identified and vice-versa. We are reporting one such rare case of vulvar Paget's disease with squamous invasion in an Asian woman.

Key Message: This rare association may lead to the misinterpretation of a squamous associated Paget's disease as Paget's disease alone if the squamous component is not identified, leading to mismanagement of a case.

Key Words: vulvar Paget's disease, Squamous carcinoma, Radical vulvectomy

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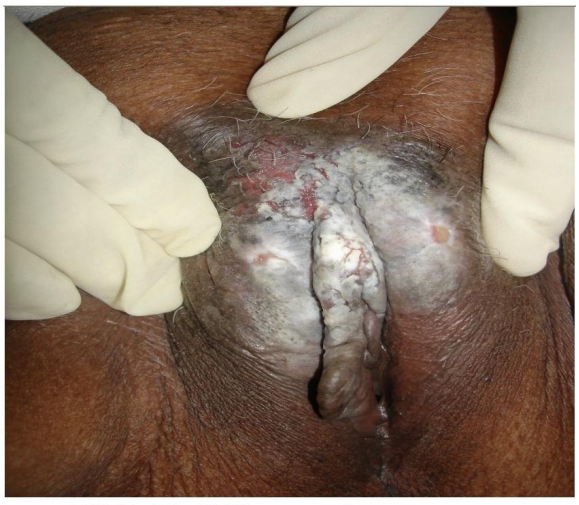
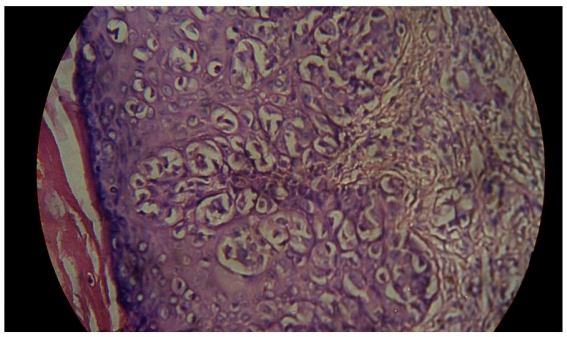
Case Report

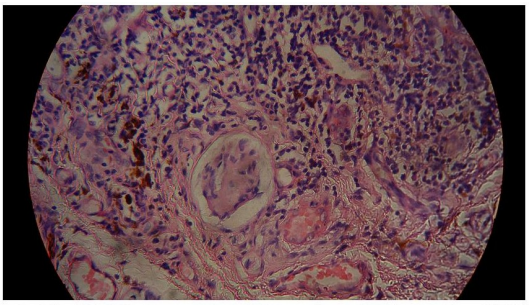
A 60 year old postmenopausal woman presented to Department of Dermatology with complaints of vulvar pruritis of 2 years duration. It was diagnosed as lichen simplex chronicus. She was treated with topical steroids. As there was no improvement with the above treatment, she was referred to Department of Obstetrics and Gynaecology . On examination, patient had hyperpigmented lichenified skin lesion of more than 2 cms in diameter with exaggerated skin markings over the medial aspect of labia majora. Curdy white precipitate was seen over the external genitalia. Few erosions and excoriations were seen [Table/Fig 1].Vulvar biopsy was taken which showed features suggestive of squamous cell carcinoma. After complete gynecological and staging work-up, radical vulvectomy with bilateral groin dissection was done. Histopathological examination of the specimen showed Paget's disease of vulva with focus of squamous invasion with lymphovascular tumour emboli [Table/Fig 2],[Table/Fig 3].



(Table/Fig 1) Macroscopic appearance



(Table/Fig 2) Microscopic picture of Paget's cells (400X)



(Table/Fig 3) Microscopic picture showing invasion (400X)

Discussion

Radcliffe Crocker first recognized and reported extra mammary Paget disease (EMPD) as a distinct clinical entity in 1889. Vulvar Paget's disease is an intraepithelial neoplasm of cutaneous origin expressing apocrine or eccrine glandular-like feature. It is characterized by distinctive large cells with prominent cytoplasm referred to as Paget cells .The subtypes of vulvar Paget disease include primary (cutaneous) Paget disease arising from a pluripotent stem cell within the epithelium of the vulva, and secondary (extra cutaneous) Paget disease of the vulva. Primary vulvar Paget's is divided into primary intraepithelial neoplasm, intraepithelial neoplasm with invasion and as a manifestation of an underlying adenocarcinoma of skin appendage or vulvar glands. Secondary vulvar Paget's arises from non vulvar sites but secondarily involves the vulva presenting as Paget's disease. This may be observed in patients with anorectal adenocarcinoma, urothelial carcinoma of the bladder or urethra, carcinoma of cervix, ovary or endometrium [1]. Vulvar Paget's disease is seen in isolation with an underlying invasive component of adenocarcinoma in 10-20% of cases [1],[2]. This is unlike Paget's disease of the breast which is almost always associated with underlying invasive malignancy. Clinically, it may have an appearance varying from moist, oozing ulcerations to an eczematoid lesion with scaling and crusting to a greyish lesion [3]. Differential diagnosis include Bowen's disease , Superficial fungal infection, Psoriasis, Leukoplakia and Eczematous Dermatosis, Lichen simplex, Mycosis fungoides [4]. A biopsy will establish the diagnosis. Very rarely, Paget's disease may be associated with squamous proliferation. A series of 35 cases of Paget's disease of the vulva and the perianal areas were studied by Brainard et al [5] . They studied the spectrum of proliferative epidermal lesions that occur in this rare disease, that generally have been overlooked. Only 2 patients were found to have malignant squamous cell carcinoma with Paget's disease in their study . We are therefore reporting one such rare case. When an underlying invasive carcinoma is present , it should be treated in the same manner as a squamous vulvar carcinoma [6].

Treatment depends on type of Paget's disease. Wide, deep local excision with 1-3 cms clinically clear margins of excision should be accomplished in primary Paget's intraepithelial neoplasm. In primary Paget's with evidence of invasion, wide partial or total vulvectomy with inguinal-femoral nodal assessment followed by chemotherapy or radiotherapy is advised.In cases of secondary Paget's disease, after treating the primary, either superficial excision, laser ablation; application of topical 5-FU or imiquimod cream can be done [7]. Follow up is mandatory. The prognosis depends on whether the disease extends beyond the epidermis and the adenexal epithelium. If it is associated with subjacent adenexal carcinoma or regional visceral carcinoma, the prognosis is poor, with Helwig and Graham reporting a death rate of 83% in one series [4].

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