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Case Report

Extramammary Paget's Disease of Axilla-A Rare Case Report

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Abstract

The incidence of Extramammary Paget Disease is rare. It is difficult to diagnose Extramammary Paget Disease, as there are many benign and malignant mimickers. We report a case of Axillary Extramammary Paget's Disease in a 40-year-old female who presented with a single violaceous plaque in the right axilla. Emphasis is given to clinical and histopathological correlation to narrow down the spectrum, and furthermore, immunohistochemical markers play a major role in reaching the Final diagnosis.

Keywords: Extramammary Paget's Disease, Paget's Disease, Breast carcinoma.

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INTRODUCTION

Extramammary Paget's disease is an uncommon neoplasm that usually develops in the areas having numerous apocrine glands. It is classified into two types- primary and secondary. Primary Extramammary Paget's Disease develops as an intraepithelial neoplasm of the epidermis. At the same time, secondary Extramammary Paget's Disease develops from the epidermotropic spread of malignant cells or direct extension from an underlying internal neoplasm [1]. Paget's Disease was first described by Sir James Paget in 1874 and in 1889 extramammary Paget disease was defined by Crocker. Paget disease can be Mammary type and Extramammary type. Mammary Paget disease is a special form of ductal carcinoma, which arises in the main excretory ducts of the breast, and may extend to the skin of the nipple and areola. Extramammary Paget's disease can be at any site, other than the breast [2].

Case Summary

A 40-year-old female patient presented to the Out-Patient Department with the complaint of a single violaceous plaque of size 11X5cm in the right axillary region (Figure 1). Plaque was not associated with pain, discharge, and itching. A biopsy was taken from the site of plaque and sent for histopathological examination. Microscopic examination revealed keratinized stratified squamous epithelium with basal layer of epidermis showing melanocytic hyperplasia and proliferation of atypical cells exhibiting vesicular chromatin and prominent nucleoli and abundant cytoplasm (Figure 2). Based on histomorphological features malignant melanoma and Paget's disease, Bowen's disease was kept in the list of differential diagnosis. To confirm the diagnosis IHC panel including EMA, CK7, P63 and HMB45 were applied. EMA and CK7 were positive in atypical cells favoring Extramammary Paget's disease (Figure 3, 4) while HMB45 was negative in atypical cells, ruling out melanoma. P63 was positive only in the basal layer of the epidermis (Figure 5), after which Bowen's disease was ruled out. The final impression was given as Extramammary Paget's Disease. Later on, the whole axillary plaque was excised along with lymph nodes and sent for histopathological examination. On microscopic evaluation, all the margins were uninvolved by tumor cells with no involvement of lymph nodes. The final Impression was Extramammary Paget's Disease pT1s N0 Mx.

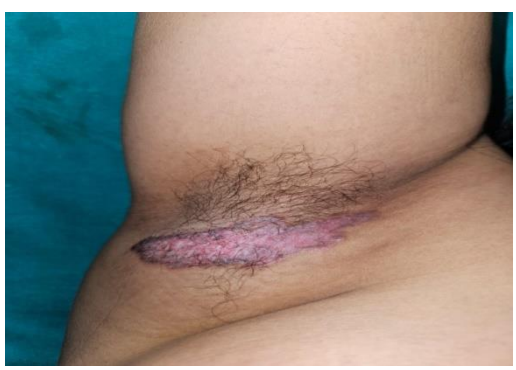

Figure 1: Single violaceous plaque of size 11x5cm in the right axillary region.

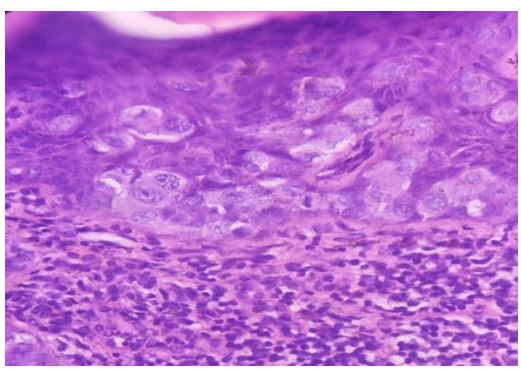

Figure 2: Microscopic examination shows keratinized stratified squamous epithelium with basal layer of epidermis showing proliferation of atypical cells exhibiting vesicular chromatin and prominent nucleoli and abundant cytoplasm.

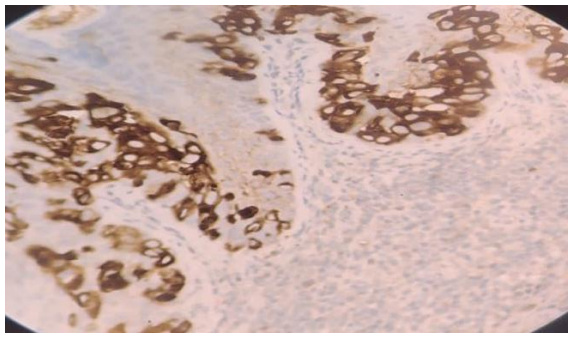

Figure 3: EMA positive in atypical cells.

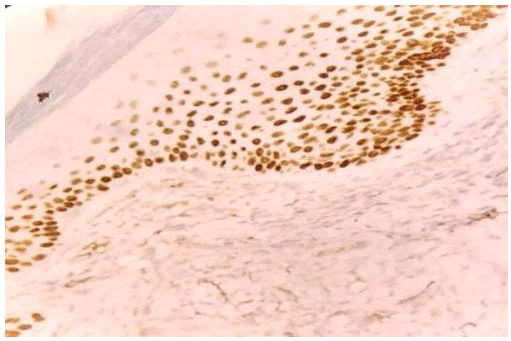

Figure 4:CK7 positive in large atypical cells.

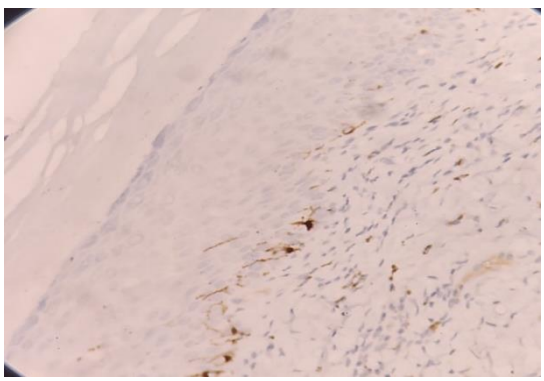

Figure 5: P63 is negative in atypical cells, and shows positivity in the basal layer of the epidermis only.

DISCUSSION

Extramammary Paget's Disease (EMPD) presents with diverse clinical features, making its diagnosis challenging. The condition commonly manifests as an erythematous plaque, scaling, ulceration, or a hypopigmented macule. Other presentations include hyperpigmentation associated with variable-sized nodules and regional lymphadenopathy. While lesions are predominantly solitary, they may occasionally appear as multiple lesions with normal skin intervening between them [3,4]. Pruritus is the most frequent symptom, reported in the majority of cases, followed by burning, irritation, pain, and bleeding. EMPD primarily affects the vulva, perianal region, scrotal, and penile skin. Less commonly involved areas include the axilla, buttocks, thighs, eyelids, and external auditory canal [5]. EMPD often presents with nonspecific clinical features, making it difficult to distinguish from other dermatological conditions such as contact dermatitis, atopic dermatitis, psoriasis, Bowen's disease, lactiferous duct ectasia, intraductal papilloma, and superficial spreading melanoma [6,7]. Immunohistochemical studies are critical in diagnosing EMPD. Positive staining for CK7 and EMA is characteristic of Paget cells. Conversely, p63 is typically positive in atypical cells of Bowen's disease but negative in Paget cells. HMB45, a marker for melanoma, is positive in atypical melanoma cells but negative in Paget cells [8,9]. Surgical excision remains the cornerstone of treatment for EMPD, with wide local excision being the preferred approach. Mohs micrographic surgery is effective in reducing recurrence rates. Despite appropriate surgical intervention, the risk of recurrence remains significant. Poor prognostic factors include dermal invasion and lymphatic metastasis [10].

CONCLUSION

Axillary EMPD is an exceptionally rare presentation. Diagnosis relies on a combination of clinical examination, histopathology, and immunohistochemical analysis. Close follow-up is crucial due to the high recurrence risk associated with this condition.

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Nil

Conflicts of interest
There are no conflicts of interest

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