



Penoscrotal Extramammary Paget's Disease - an Exceedingly Rare Entity

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Case Study

We are reporting a rare case of extra mammary Paget's disease involving genital region in a 62 year old male. Patient complained of an erythematous lesion over penoscrotal area with relentless progression for last 15 years, associated with intermittent sticky discharge and mild pruritus (Figure 1). History of application of topical steroid and antibiotic without any improvement. Other significant history includes adenocarcinoma in mother and hepatic cell carcinoma in late father. On examination, there was a 10 cm × 2 cm erythematous plaque with eroded surface, fungating near the root of the penis where it had started as an asymptomatic papule, extending into scrotum and shaft of the penis, with gray crusts hanging on to the eroded surface. Vitals and systemic examination was uneventful. Routine examinations were within normal limits. Histopathology View 10x revealed Intra-epidermal pagetoid proliferation of atypical cells in nests and also in continuous fashion. Dense chronic inflammatory infiltrate is present in subepidermal stroma in band like fashion (Figure 2). View 40x demonstrated tumor cells having abundant vacuolated pale staining cytoplasm with moderately pleomorphic nuclei and mitotic activity. PAS positive, diastase resistant material was also noted in cytoplasm of some of the atypical cells. On the basis of Clinical and histopathological analysis our differentials were Extramammary Paget's disease, Pagetoid Bowen's disease and malignant melanoma (superficial spreading). For further clarification, Immunohistochemistry was performed with CK7, CK20, S100 and HMB-45. Result was strong positive for CK7, weakly positive for CK-20 and negative for both S100, HMB-45 (Figures 3-6). Thus we arrived at the final diagnosis of extramammary Paget's disease. Surgical management was planned and the patient was treated with wide local excision followed by skin grafting, margins declared free histopathologically.

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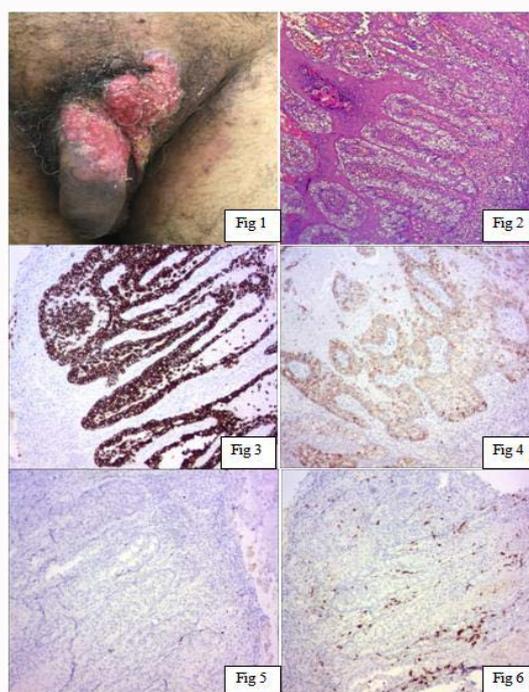
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Figures:

Extramammary Paget's disease (EMPD) was originally described by Crocker in 1989 [1]. It is a rare clinical entity of unknown etiology and mostly affects elderly individuals with a mean age of 67.9 years [2]. Apocrine regions are the main site of involvement with females more frequently affected than males [3]. EMPD usually presents as an erythematous, skin colored or even hypo pigmented plaque, which may be pruritic (73%) or asymptomatic [4]. Lesions may resemble benign dermatoses, squamous cell carcinoma *in situ* or melanoma. The characteristic diagnostic point is unremitting progression despite topical therapy. Three usual patterns of EMPD are seen: (1) *in situ* epithelial form without associated carcinoma (2) epithelial form with adnexal carcinoma (3) those associated with visceral malignancy of either genitourinary or the gastrointestinal tract. The typical histology of EMPD is Paget's cells which are intraepithelial neoplastic cells with glandular differentiation having abundant pale staining cytoplasm and large atypical nuclei. The cells invade epidermis and rarely the dermis. Lymphocytic infiltration is noted in upper dermis. Histological differentials include pagetoid Bowen's disease and malignant melanoma *in situ* [5]. Although treatment of choice is surgical excision, other remedy includes topical 5-FU, bleomycin, 5% imiquimod, CO₂ laser ablation, curettage, radiation and cryosurgery. Local recurrence is a major issue mainly because of multifocal involvement and trouble in clinical demarcation of tumour margins [6].

According to some radical surgery should be reserved for invasive disease or relapse cases [7].

Prognosis depends on the disease extent and is poor in cases where the involvement is beyond the epidermis and adnexal epithelium. Poor prognostic factors include dermal or lymphatic infiltration, high levels of CEA or internal malignancy. Overall survival similar to general population in early cases [8].

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