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# Primary perianal extramammary Paget's disease: Case report with review

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**Abstract**

Extramammary Paget's disease (EMPD) is a rare cancer which involves the skin and apocrine glands. It involves the vulva, perianal region, scrotum, penis and axilla. Primary disease originates from intraepidermal cells and secondary originates from underlying neoplasm. The disease presents with thickened plaque like lesion with erythema or white scaly appearance. The cancers of urinary tract like the bladder, urethra and prostrate are associated with EPMD involving the genitalia and rectal tumours are associated with perianal disease. The disease affects females more than males with the median age being 72 years. Primary perianal EMPD is even rare in presentation. It is a slow growing tumour and the prognosis is usually favourable other than advanced stage and old age. The surgical resection with clear margins is considered to be the standard of care and provides good outcomes. However, if surgery is not feasible other treatment options like imiquimod 5% topical cream and radiation therapy may be offered. We present a young male with perianal EMPD who was managed with surgical resection with clear margins with split skin graft leading to a favourable outcome.


*Keywords:* Perianal Paget's disease; Surgical resection; Clear margins; Skin grafting

## 1. Case report

A 39 year old male was referred to us with a history of a perianal non healing lesion since 3 years. It started as a small patch and was managed by dermatologists and surgeon elsewhere as fissure in ano, eczema, fungal infection etc. It had increased in size and involved both sides of the perineal lesion. The patient had pruritus with mild pain in the perianal area with the skin having a leathery feel (Fig. 1). There was no discharge and there was no history of constipation, or any problems in continence. There was no bleeding per rectum. The patient consumed alcohol occasionally and he was not a smoker. General physical examination was normal without any peripheral lymphadenopathy. On perianal examination there was a thickened plaque like area in the perineal region which involved left side more than the right as like a butterfly sparing the external anal opening. On per rectal examination the resting anal tone, squeeze was normal. Colonoscopy was performed till ileocaecal junction and was normal. An ultrasound examination of abdomen was normal and it did not reveal any lymph nodes in the inguinal region.

There was a clinical suspicion of EMPD or a non healing ulcer. We performed a punch biopsy which was suggestive of primary perianal EMPD. The patient was counselled for a sphincter saving surgical resection with per operative frozen section for the margin status (Figs. 2 and 3). The frozen section revealed clear margins and intraepidermal disease. Split thickness skin graft was taken from the thigh and applied with pressure at the perianal area (Fig. 4). The post operative period was uneventful and the patient had around 40% graft uptake. The final histopathology confirmed primary perianal EMPD with

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negative margins confirmed on immunohistochemistry as CK 7 and GCCDFP-15 positive (Fig. 5).

The patient is doing well and is completely relieved of his symptoms. There is no evidence of any residual disease or recurrence on more than 2 years of follow up which includes clinical examination and short colonoscopy on an annual basis (Fig 6)

## 2. Discussion

EMPD was first described by Crocker as a rare disease of scrotum and penis (Crocker, 1889). Darier and Coulillaud first reported perianal Paget's disease (Darier and Coulillaud, 1893). Perianal EMPD is a rare disease as published in experience of high volume centres. Disease presents in elderly patients with a mean age of 60 years. In contrast our patient is a young male (Rajendran et al., 2014; Isik et al., 2016; Perez et al., 2014). Because of its rarity the disease is often misdiagnosed. The patients usually present late after multiple treatment protocols by various clinicians. Presentation includes single or multifocal lesions that are dry, erythematous area which gradually progress to eczematoid, crusted, ulcerated or papillary lesions. Pruritus may precede these skin lesions (Rao and Henry, 2004). It has to be differentiated from superficial spreading melanoma, Bowen's disease, neuroendocrine carcinoma, mycosis fungoides, psoriasis, leucoplakia, eczema and fungal infection (Jones et al., 1979). Another differential diagnosis is pagetoid effect which is intra epidermal spread of visceral carcinoma (Minicozzi et al., 2010). The disease which involves the vulva, perianal region, scrotum, penis and axilla may be primary if it arises from the skin or secondary in case arising from an underlying neoplasm, which is reported in 24% of patients with a worse prognosis (Chanda, 1985). Rectal adenocarcinoma involving the perianal area or the tumour of urinary tract involving the genitalia is usually associated.

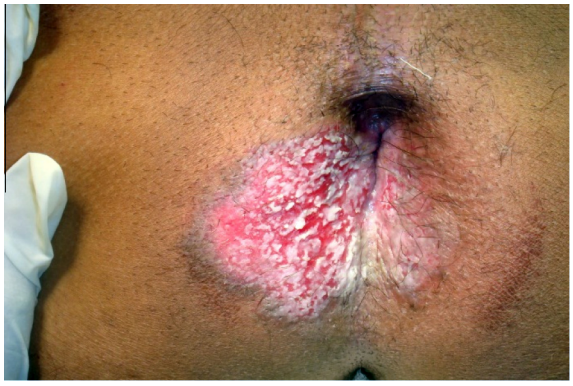

Figure 1. Perianal Paget's disease.

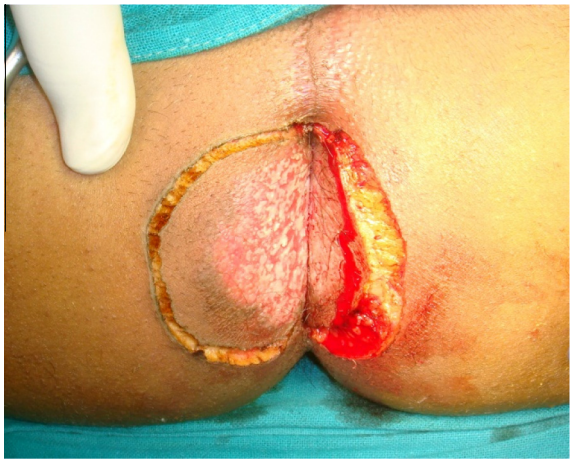

Figure 2. Excision of involved perianal skin.

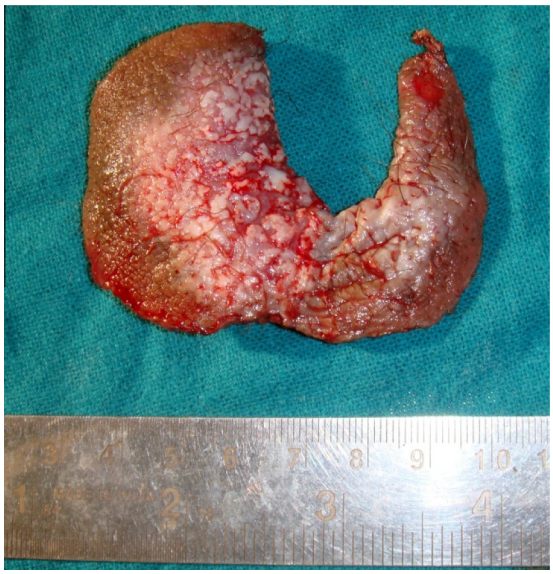

Figure 3. Excised Skin of paget's disease.

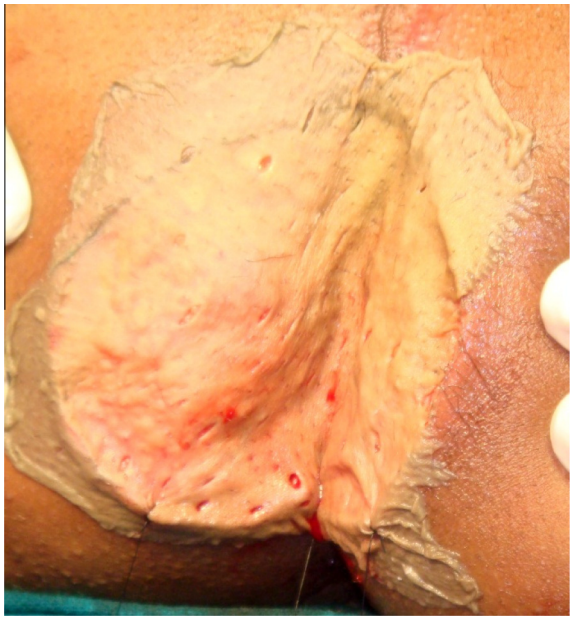

Figure 4. Skin grafting over raw wound.