

Advanced Extramammary Paget's Disease Of Scrotum – An Uncommon Presentation

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Abstract

Extra Mammary Pagets Disease(EMPD) mostly involves areas rich in apocrine glands such as perianal area, perineum, vulva, axilla, mons pubis, glans penis and eyelids and rarely scrotum . EMPD is often seen in concurrence with an underlying visceral carcinoma but can also arise de novo. EMPD lesions may also be accompanied by adenocarcinoma in situ or invasive adenocarcinoma of the apocrine glands . The average age of presentation is usually between 58 and 67 and also has more preponderance to post menopausal women. Lesions clinically present as erosive plaques, scaly, eczematous and erythematous ulcers. Early identification is vital and requires high index of suspicion or else diagnosis is usually delayed. Here, we present a patient who had scrotal lesion with bilateral inguinal lymphnodal metastasis.

Key Words: Extramammary Paget's Disease (EMPD), Adenocarcinoma, inguinal lymphadenopathy

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I. INTRODUCTION

Paget's disease of the breast was first identified and described in association with underlying carcinoma breast by Paget in 1874 while Crocker first identified the first Extramammary Paget's disease in 1889(1). Most common area involved in EMPD is vulva which accounts to 65% of cases and later perianal region accounting for 20% of cases. Only 14 % of cases are contributed by EMPD of male genitalia that is penis and scrotum together (2). The clinical presentation is usually a non-specific erosive, erythematous and eczematous lesion of the skin mimicking other dermatological conditions. Almost an average of two years is being noted from the time of onset of symptoms to the actual appropriate diagnosis(3). Lesions in the penoscrotal area also has eczematous presentation presentation and often is associated with pruritis and burning sensation. Rarely, penoscrotal lesions might be associated with inguinal lymphadenopathy and ipsilateral limb edema.

The exact pathology of EMPD is not entirely clear . According to the current literature, EMPD has two forms : Primary and secondary. Primary EMPD arises from epithelium or underlying apocrine gland, initially limited but has the tendency to become invasive leading to lethal metastasis. Secondary EMPD is associated with underlying adenocarcinoma of adnexal glands or with genitourinary or gastrointestinal tract(2). According to the available literatures, 42% of patients have underlying carcinoma associated, nevertheless, penoscrotal EMPD is associated with less incidence of internal malignancy(4).

Here, we describe a case of primary and locally advanced EMPD which initially presented in the scrotum with bilateral inguinal lymphadenopathy. We have discussed about the diagnostic investigations conducted and the treatment offered. Despite the appropriate treatment offered patient developed multiple distant metastasis and succumbed to the disease.

II. CASE PRESENTATION

75 year old gentleman presented to the surgical oncology department with history of erythema and ulcer over left side of scrotum associated with pruritis for 3 months . Patient also had history of swelling in the bilateral groin but not associated with pain. An erythematous superficial ulcer with rolled out margins measuring 8x7cm was present in the left side of scrotum near the root . Bilateral multiple enlarged inguinal lymph nodes noted(L>R). Patient had no history of symptoms pertaining to other systems. Patient had no history of any other skin malignancies or positive family history. No history of any other addictions.



Figure 1 : Showing erythematous ulcer in the left scrotum

Investigations:

Patient presented with an outside wedge biopsy report of the scrotal lesion as squamous cell carcinoma. Ultrasonogram of the bilateral groin and abdomen revealed enlarged bilateral inguinal lymph nodes. Fine needle aspiration cytology was done from the enlarged inguinal lymph nodes and reported to be having carcinomatous deposits.

Treatment and Post op histopathology report:

Patient underwent wide local excision of the scrotal lesion with primary closure and bilateral ilio-inguinal dissection. Intra-operatively frozen section was sent to ascertain the free margins of the excised primary tumor.

The overlying skin shows infiltration in the form of pagetoid spread and also direct infiltration with ulceration and necrosis of the skin. Bilateral lymph nodal specimens showed deposits with extra nodal extension. Immunohistochemistry was done and cytokeratin and GATA 3 were diffusely positive in all lesional cells. Final diagnosis was reported as Extramammary paget's disease of scrotal skin associated with invasive adenocarcinoma with lymphovascular invasion.

After recovering from surgery, patient was referred for adjuvant radiotherapy as lymph nodal positivity was present. When investigated further with PETCT, patient was found to have multiple bone metastasis and was advised palliative chemotherapy. During the process of treatment, unfortunately, patient succumbed to the disease.

Figure 2: Positive mucicarmine.

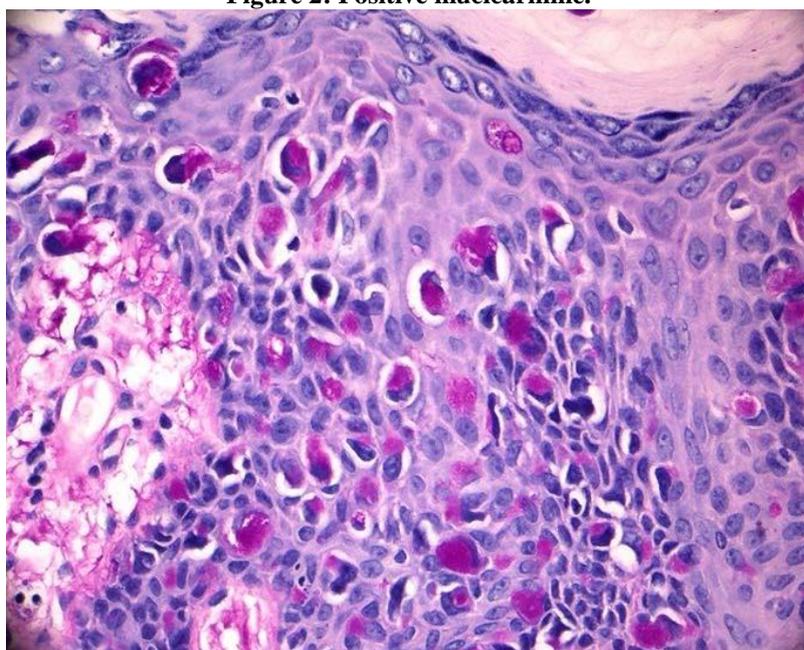


Figure 3 : Positive Periodic Acid Schiff stain

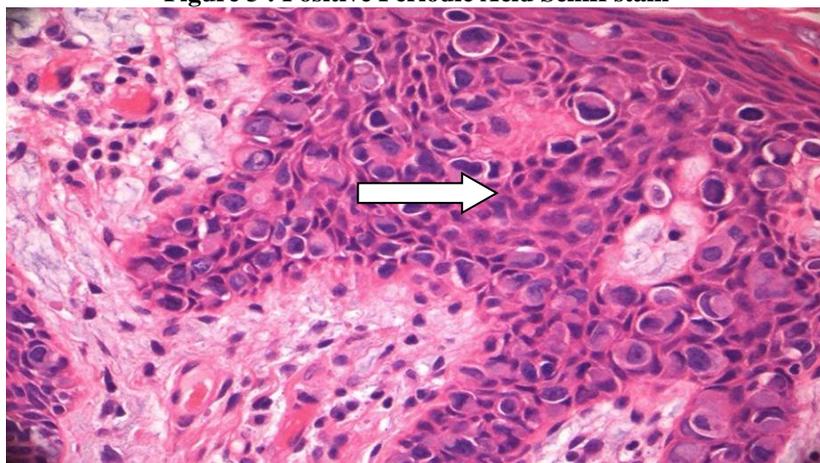
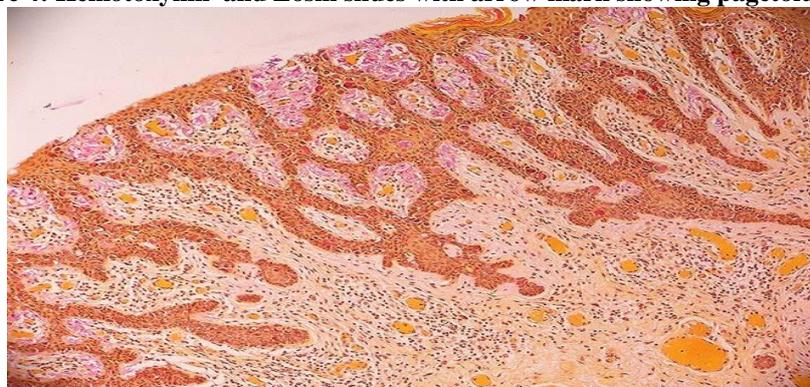


Figure 4: Hemotoxyllin and Eosin slides with arrow mark showing pagetoid cells.



III. DISCUSSION :

EMPD is remarkably rare and it has diverse clinical presentation from indolent to a locally advanced disease. Most common site of presentation being vulva followed by perianal region (2). EMPD has more female preponderance especially in post menopausal women. The actual pathogenesis of the disease is yet to be elucidated as there are various school of thoughts being proposed.

Most of the authors propose that it is caused by an underlying carcinoma from where the metastatic cells migrate to the epidermis and presents itself as EMPD(5) (6). Another school of thought is that these paget's cells are derived from apocrine gland cells and hence explaining the anatomical location of this disease where sweat glands are abundant(7). Many case reports and series have been reported of the EMPD in vulval and perianal regions but only a very few cases of EMPD of scrotum and especially with such advanced presentation as in our case have been reported overall.

Surgery with wide local excision and lymphadenectomy in a locally advanced disease is the primary modality of treatment in EMPD. Nevertheless, achieving a negative margin is crucial when associated with invasive carcinoma and are associated with recurrences if margins are involved(8). Various case reports have been published on the role of radiotherapy as therapeutic in patients who are not fit for surgery and also as adjuvant therapy to surgery(9). There is also role of chemotherapy in cases of locally advanced or metastatic diseases with agents like carboplatin, docetaxel, 5-FU, vincristine(10).

In our case report, patient presented initially with locally advanced disease with scrotal skin involvement and bilateral inguinal lymphadenopathy. Wide excision with negative margins as proven by frozen section along with bilateral ilio-inguinal dissection were done. Patient was referred for adjuvant radiotherapy and chemotherapy. During the evaluation, he also was found to have metastasis as detected by PETCT and patient succumbed to the disease three months later despite aggressive surgical intervention.

IV. CONCLUSION

As the EMPD of scrotum is very rare, it requires a high index of suspicion when the patient presents with any erythematous and eczematous lesion of scrotum. Histopathological examination of the biopsy from the lesion helps in the early intervention. As there are no approved treatment protocols in cases of locally advanced

and metastatic EMPD, combination of various modalities are being tried in an attempt to achieve near total resolution of the disease. Further studies and reporting are still required in order for the better understanding of the disease and its treatment.

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