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Perianal Eruption in a 40-Year-Old Woman: An Exceptional Case of Extramammary Paget's Disease

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Abstract **Case Report**

Extramammary Paget's disease (EMP) is a rare form of intraepithelial adenocarcinoma that primarily affects older women. This case report describes a 40-year-old patient diagnosed with EMP who presented with a rapidly growing, painful, and bleeding perianal erosion that did not respond to antifungal treatment. The patient underwent wide excision of the lesion and the postoperative course was uneventful. The case highlights the importance of considering EMP in the differential diagnosis of perianal eruptions, particularly in older women, and emphasizes the need for careful histological examination and appropriate management.

Keywords: Extramammary Paget's disease, perianal erosion, adenocarcinoma, histological examination, treatment.

INTRODUCTION

Extramammary Paget's disease (EMP) is a rare form of intraepithelial adenocarcinoma that primarily affects older women [1]. It typically develops in apocrine-rich areas such as the genital, anal, and axillary regions. Although the relationship between EMP and associated neoplasms is not well understood, surgical excision remains the preferred treatment approach [2]. This case report describes a 40-year-old patient diagnosed with EMP who presented with a rapidly growing, painful, and bleeding perianal erosion that did not respond to antifungal treatment. The patient underwent wide excision of the lesion and the postoperative course was uneventful. The case highlights the importance of considering EMP in the differential diagnosis of perianal eruptions, particularly in older women, and emphasizes the need for careful histological examination and appropriate management.

CASE PRESENTATION

This is a 40-year-old patient who has been followed for 1 year for squamous cell carcinoma of the cervix treated with radiotherapy for 6 months. The patient has since developed a painful, bleeding, rapidly growing perianal erosion on the right buttock, which has been treated with antifungal medications for several months without improvement. Our dermatological examination found an erosion in the intergluteal fold, starting at the anus and spreading out as an oil stain on the right buttock, oozing with an irregular border (Figure 1). There were no genital or pubic lesions or lymphadenopathy. Dermoscopic examination revealed glomerular vessels, a red-white zone, bright white lines, gray/black dots, and white scales (Figure 2). A skin biopsy was performed and showed that the epidermis was widely infiltrated by an in situ adenocarcinoma, consisting of tumor cell clusters with abundant pale cytoplasm and large irregular nuclei with prominent nucleoli, while the dermis was free of any tumor proliferation (Figure 3). Immunohistochemical analysis showed positive results for CK7/MUC5A/P53/HER2 antibodies, but negative results for CK20/RE/RP/PAX8/CDX2 antibodies. The histological appearance and immunohistochemical profile were consistent with extramammary Paget's disease. Further testing, such as cervical and vaginal smears, fibroscopy, and colonoscopy, were unremarkable. Pelvic MRI showed no tumor recurrence in the cervix, which was sclerotic. So, after these tests, we have retained the diagnosis of a primary extramammary Paget's disease given its histological characteristics (limited to the epidermis) and the absence of involvement of visceral organs. The patient underwent wide excision of the lesion with a 2 cm safety margin. The postoperative

course was uneventful. The patient is currently undergoing a program of 10 LED sessions to ensure proper healing (Figure 4), she will be reviewed once complete healing is achieved and then reviewed at 3 months, 6 months, 1 year, and then annually to detect a frequent relapse rate.

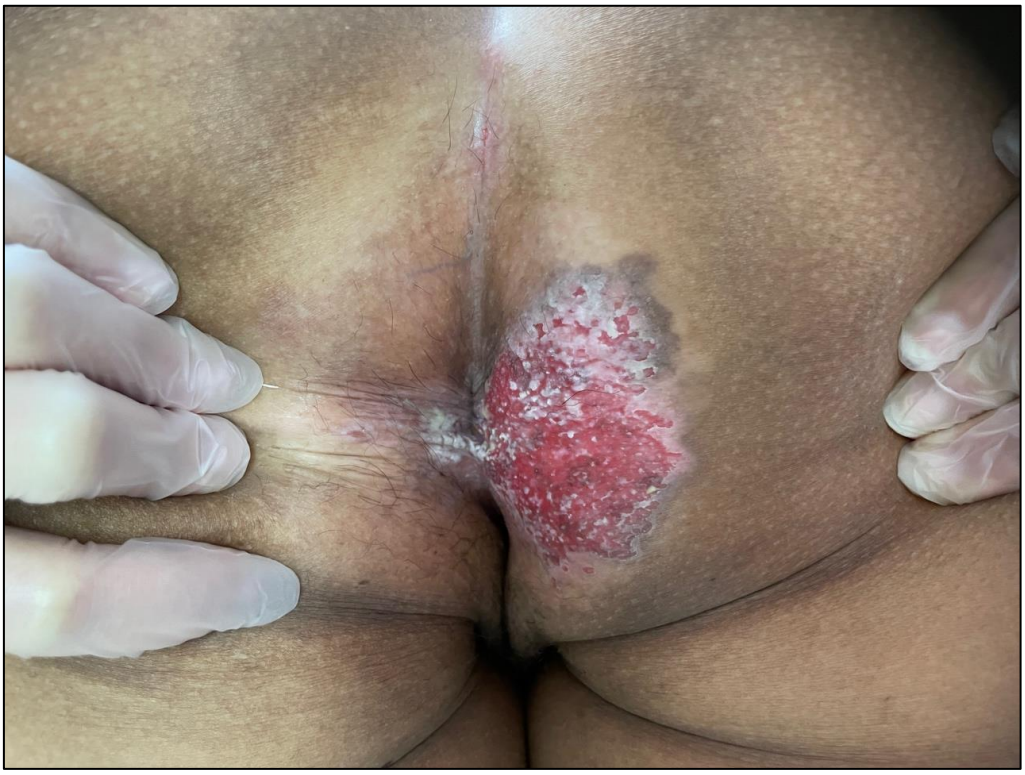

Figure 1: Clinical image showing erosion in the intergluteal fold, starting at the anus and spreading out as an oil stain on the right buttock, oozing with an irregular border

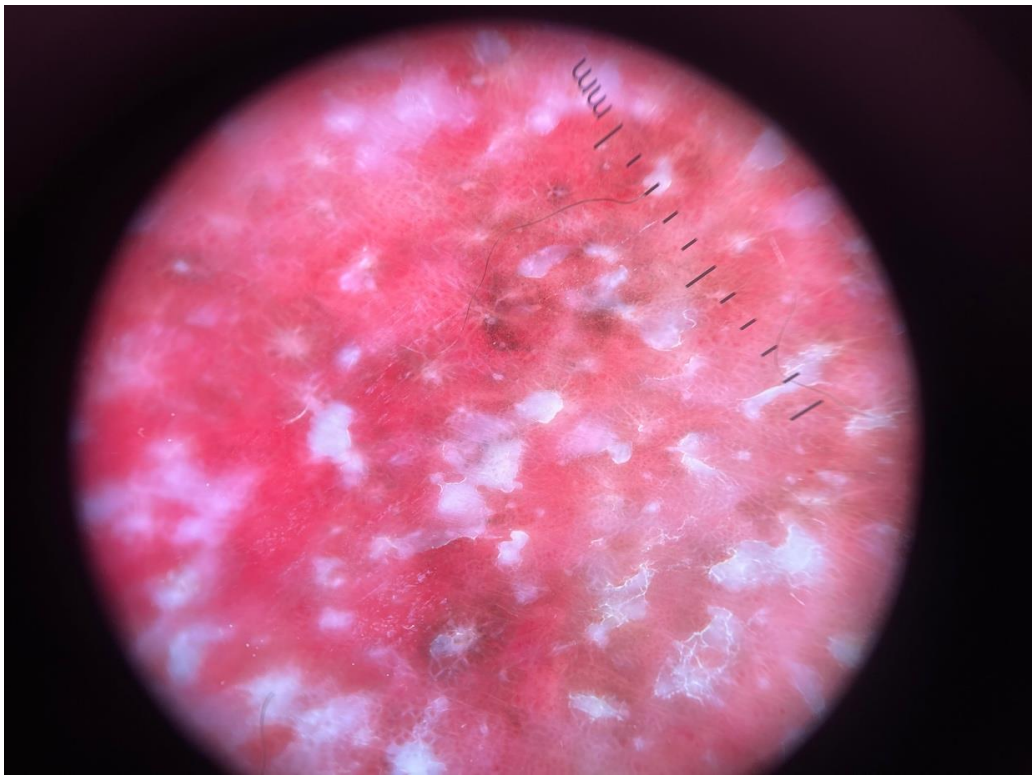

Figure 2: Dermoscopic examination revealing glomerular vessels, a red-white zone, bright white lines, gray/black dots, and white scales

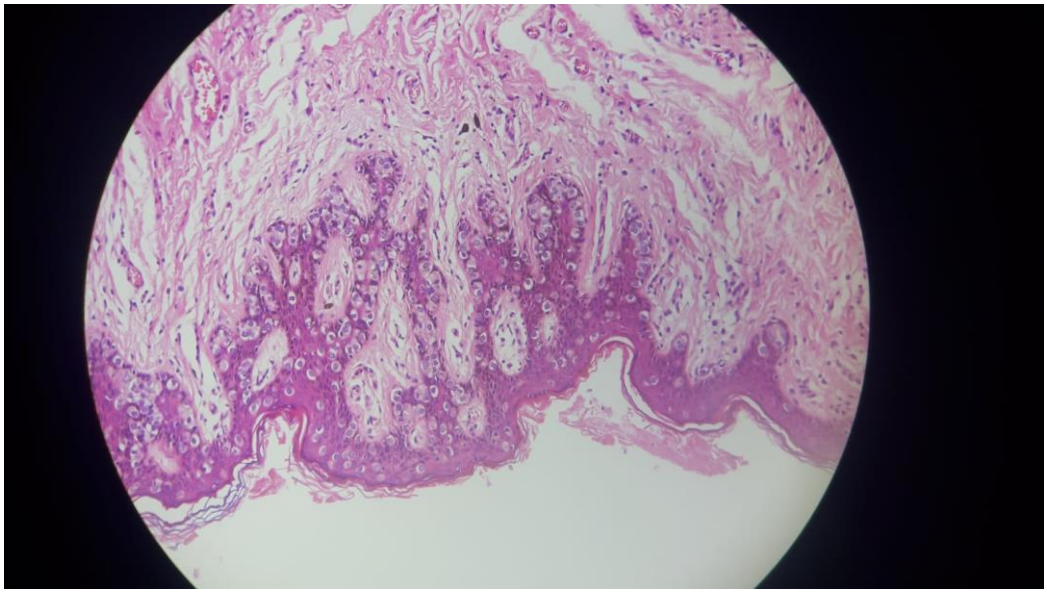

Figure 3: Skin biopsy showing that the epidermis was widely infiltrated by an in situ adenocarcinoma, consisting of tumor cell clusters with abundant pale cytoplasm and large irregular nuclei with prominent nucleoli, while the dermis was free of any tumor proliferation

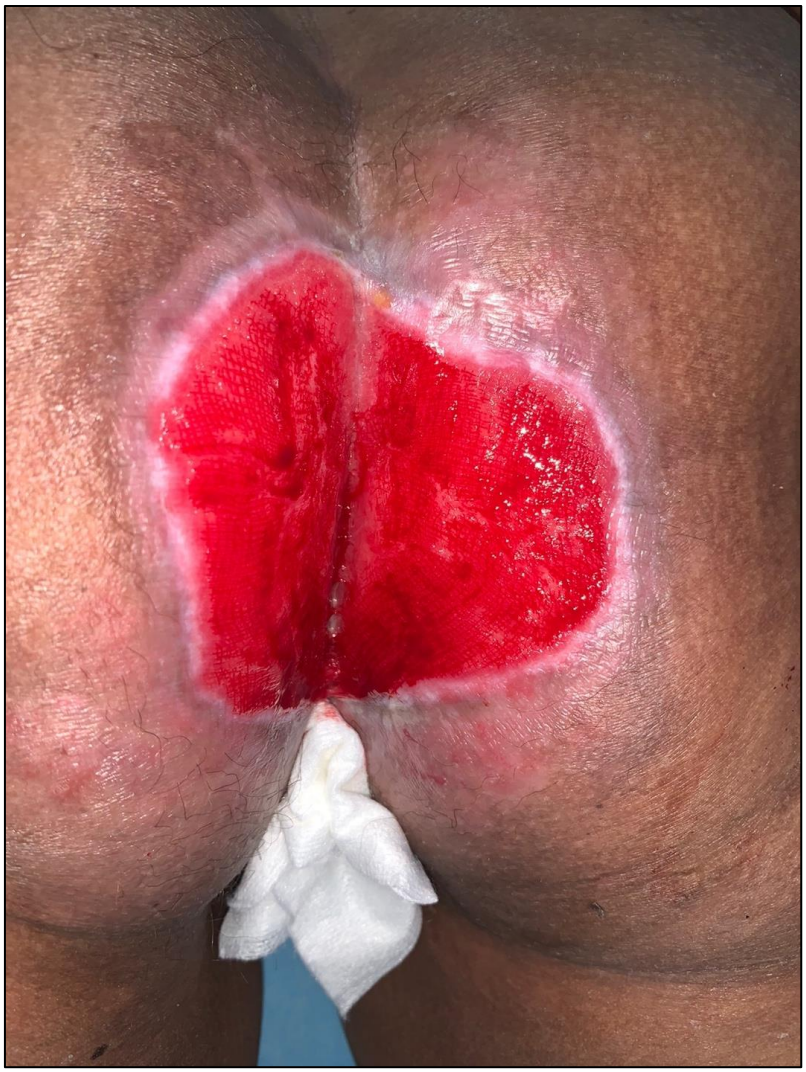

Figure 4: The patient is currently undergoing a program of 10 LED sessions to ensure proper healing

DISCUSSION

Extramammary Paget's disease (EMP) is a rare form of intraepithelial adenocarcinoma that primarily affects older women, with an average age of 74 years and a male to female ratio ranging from 1/4 to 1/7 [3]. Our case is unique because of the young age of the patient (40 years) which is exceptional for a primary EMP. He developed in perianal area, which is one of the typically apocrine-rich areas such as the genital, anal, and axillary regions, with the vulvar region being the most common site, followed by the perianal region. Other sites that may be affected include the perineal area, inguinal fold, scrotum/penis, and axillary areas. In rare cases, EMP may occur in areas devoid of apocrine glands, such as the scalp, cheeks, fingers, knee, back, arms, thorax, and abdomen [4-5]. EMP can be classified into primary (confined to the epidermis and dermis) and secondary (involvement of visceral organs). Primary EMP, as in our case, initially limited to the epidermis (in situ), can then progressively invade the underlying dermis (invasive EMP) and even lead to lymph node or visceral metastases at advanced stages. Secondary EMP would be related to invasion of the epidermis by an underlying tumor, as in most cases of mammary Paget's disease. The tumor can then be located in the underlying dermal appendages or in a neighboring organ whose epithelium is contiguous with the epidermis affected by Paget's disease. [2]. Clinically, the lesions are clearly defined, more or less infiltrated, erythematous with gray-white plaques, with chronic evolution, can take eczematous, crusty, papillomatous or, rarely, ulcerated forms [6]. Histologically, acantholytic forms would favor recurrence and immunohistochemistry is useful as it can provide information on its primary or secondary character, and can also constitute a prognostic marker [5]. EMP is a pathology that occurs most often in subjects over 65 years of age; this is also the age at which the incidence of cancers is highest, hence the need for a comprehensive and hierarchical assessment according to the location of the lesion [6], but in our case, our patient was young, but we searched for an associated cancer. In terms of treatment, surgical treatment remains the first-line treatment. There are two types of surgery: standard surgery, which consists of a wide excision with a lateral margin of 2-5 cm, depending on the authors [7, 8], and micrographic surgery, developed by Fréderic E. Mohs [4], which is intended to treat severe forms of cutaneous carcinomas. By performing a microscopic and topographic analysis of the entire excision specimen, it is possible to reduce the excision margins while preserving healthy tissue and reducing the risk of recurrence, which is very high in standard surgery, varying between 33% to 61%, even if the excision margins are healthy, as the lesions evolve in a jump [5]. Local immunotherapy can be used as neoadjuvant therapy before surgery, or in cases where surgery is contraindicated [9]. CO2 laser therapy can be used in place of surgery for vulvar locations in young women as it preserves sexual function and anatomy. Unfortunately, recurrences remain high and early [10]. Radiotherapy is an alternative to surgery in cases where the patient refuses or is contraindicated for surgery. It is used exclusively in the context of invasive EMP, with doses of more than 60 Gy and margins of 2 to 5 cm around the lesions, with recurrences that can reach 50% [11]. Dynamic phototherapy selectively destroys pathological cells that have accumulated a photosensitizing substance. The rate of relapse remains high, but the treatment can be repeated without aesthetic or functional consequences, unlike surgery [12]. Isolated extramammary Paget's disease has a favorable prognosis, but with frequent local recurrences after treatment. The prognosis is related to the presence or absence of an associated carcinoma, ancillary or distant [2].

CONCLUSION

This case report presents a rare and unique case of extramammary Paget's disease in a younger patient. EMP is a rare form of intraepithelial adenocarcinoma that primarily affects older women. It typically develops in apocrine-rich areas such as the genital, anal, and axillary regions. This case emphasizes the importance of considering EMP in the differential diagnosis of perianal eruptions, particularly in older women, and the need for careful histological examination and appropriate management. Although surgical excision remains the preferred treatment approach, other treatment options such as local immunotherapy, laser CO2, radiotherapy, and photodynamic therapy are also available. The patient outcome was good with no recurrence after the surgery but the risk of recurrence is always high.

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