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Perianal Paget's Disease: Case Report

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ABSTRACT

Introduction: *Perianal Paget's disease (PPD) is considered a subgroup of extramammary Paget's disease (EMPD). It represents less than 1% of all anal diseases and 6.5% of all EMPDs. It is characterized by the presence of Paget cells in the epidermis, representing an intraepithelial adenocarcinoma. Clinically it presents as an erythematous plaque in areas of apocrine glands. There is an association between Paget's disease and other carcinomas of the gastrointestinal and genitourinary tract and remote areas of the skin. As treatment, resection with wide margins is recommended.*

Case: *A 63-year-old female consulted with a 4-year history of a perianal pruritic lesion without response to topical corticosteroids. Examination revealed a perianal erythematous lesion measuring 5 cm in diameter. Biopsy reported PPD. Local excision with wide margins and primary closure of the defect was performed. Pathological examination reported EMPD with disease-free skin adnexae and apocrine glands, and clear resection margins. No recurrence occurred after a two-year follow-up.*

Discussion: *PPD is a rare disease. Clinical presentation is characterized by non-specific symptoms, making diagnosis difficult. Faced with failure to treat eczema or dermatitis, PPD should be suspected and the lesion should be biopsied. Some authors recommend surgical treatment, from local excision with wide margins to abdominoperineal resection. Follow-up of patients with PPE is essential and should focus on controlling local recurrence and the development of associated carcinoma.*

Keywords: *Extramammary Paget's Disease; Anal Paget's Disease; Anal Neoplasia*

INTRODUCTION

In 1874 James Paget described Paget's disease of the breast (MPD), in 1889 extramammary disease (EMPD) of the scrotum and penis was identified[1] and in 1893 Darier and Couillaud[2] reported perianal Paget's disease (PPD), considered a subgroup of the EMPD. The most common location is the vulva.

PPD represents less than 1% of all anal diseases and 6.5% of all EMPD cases.[3] MPD and EMPD are characterized by the presence of Paget cells in the epidermis, representing an intraepithelial adenocarcinoma and share a similar clinical presentation, although the difference lies in location and histogenesis, which continues to be a matter of debate.[4] EMPD presents as an erythematous plaque in areas with apocrine glands, suggesting that these could be the site of origin. The association between Paget's disease and other carcinomas of the gastrointestinal tract, genitourinary tract, and remote areas of the skin has been recorded in previous publications.[4] Due to the low frequency of the disease, there are no studies to investigate the optimal treatment which remains controversial. Wide local excision of skin and perianal subcutaneous cellular tissue with reconstruction of the defect is recommended, for which multiple techniques have been described.[2]

The authors declare the absence of conflicts of interest.

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CASE

A 63-year-old woman arrived to the consultation with a 4-year history of a perianal pruritic lesion and no other pathological medical history. She had received treatment at another medical center with topical corticosteroids without improvement. Physical examination revealed an erythematous lesion of approximately 5 cm in diameter that affected the two posterior quadrants and the midline (Fig. 1). The rectal examination was normal and no inguinal adenopathies were palpated. The biopsy reported PPD. Colonoscopy, gynecological ultrasound, and chest, abdomen, and pelvic tomography were performed, all normal. Local excision with wide margins and primary closure of the defect was performed (Fig. 2, Fig. 3 and Fig. 4). Histopathology report showed intraepidermal EMPD that spares the dermis, adnexae and apocrine glands, with clear resection margins. The patient evolved favorably with hospital discharge at 24 hours. She had no recurrence after two years of follow-up.

DISCUSSION

PPD is a rare disease, less than 200 cases have been reported and its incidence is difficult to estimate.[5] In Argentina there are few published cases.[6] The clinical presentation is characterized by non-specific symptoms such as pruritus, pain, erythematous plaque and even bleeding, making the diagnosis difficult.[2,4] A differential diagnosis should

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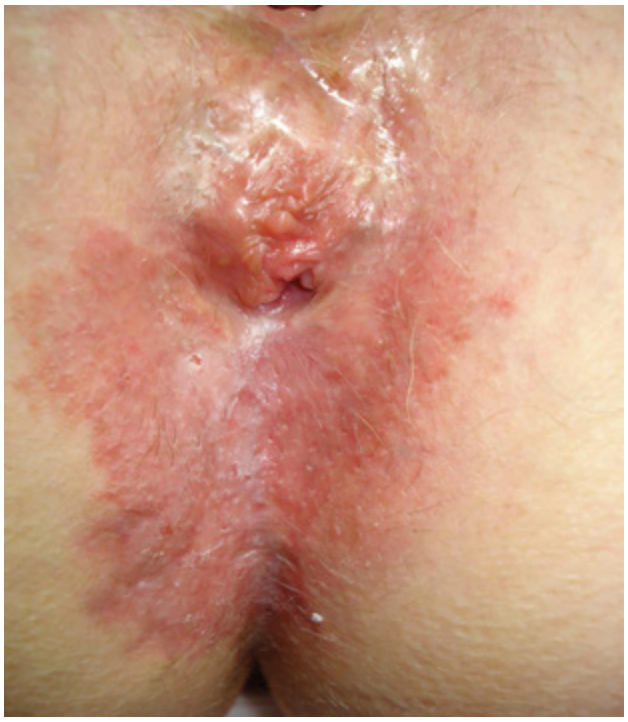

Figure 1: Perianal erythematous lesion with partially defined borders, involving both posterior quadrants and the midline and extending approximately 5 cm towards the intergluteal region.

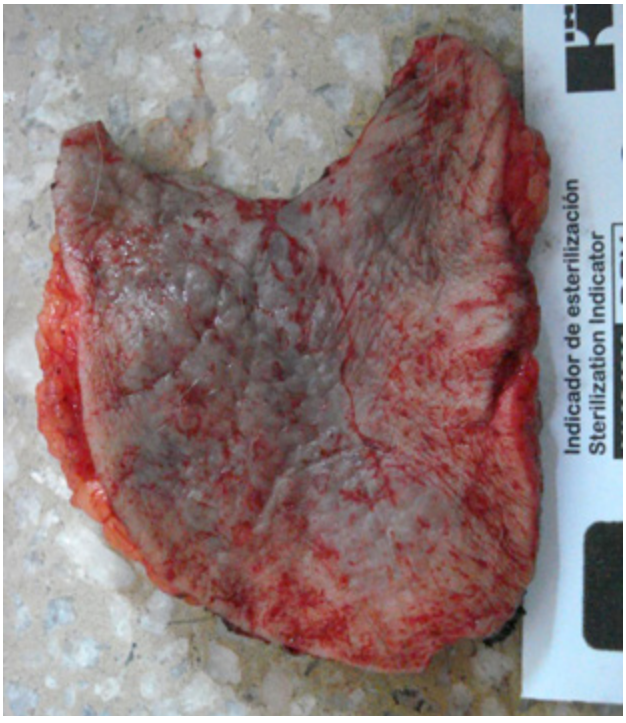

Figure 2: Biopsy specimen with negative margins.

be made with dermatitis, eczema, hemorrhoids, anal fissure, condylomata, Bowen's disease, melanoma and anal carcinoma.[3] In the present case, the patient had been treated with topical corticosteroids without a favorable response. Faced with failure to treat eczema or dermatitis, PPE

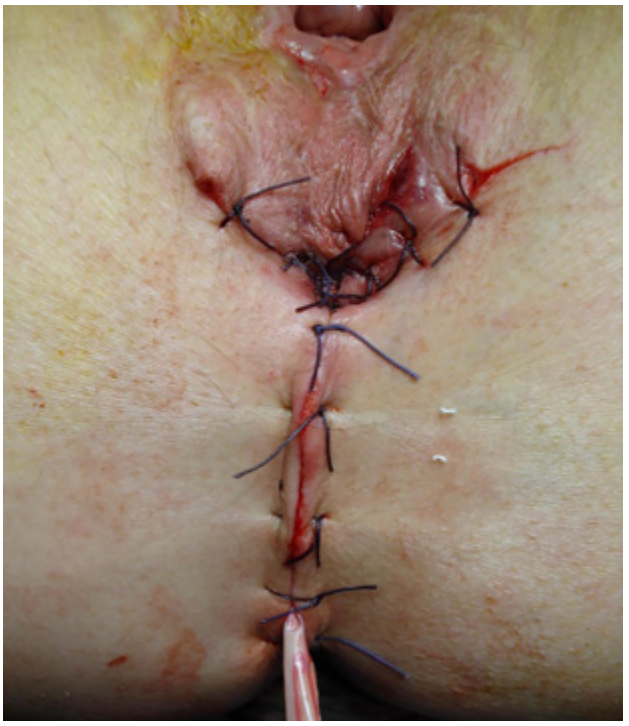

Figure 3: Primary closure of the defect with interrupted resorbable sutures.

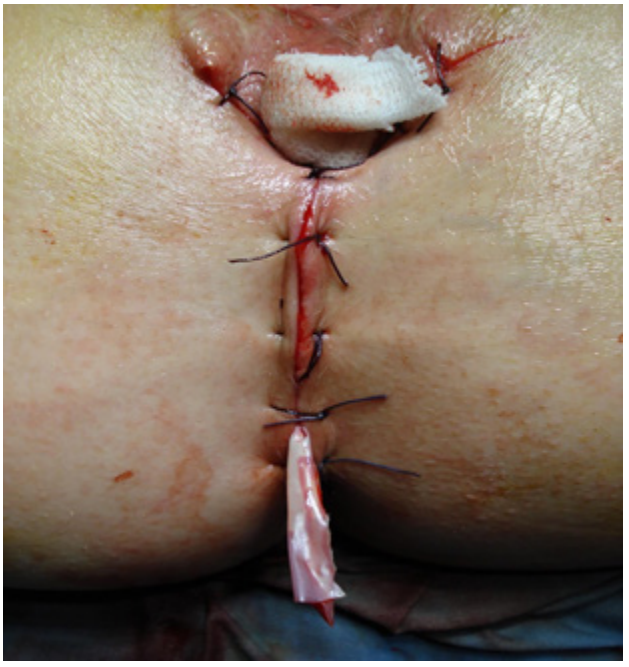

Figure 4: Gauze indicates the location of the anal opening.

should be suspected and a biopsy of the lesion performed. It is difficult to reach a consensus on treatment since most of the literature deals with case reports. However, some authors recommend surgical treatment, ranging from local excision with wide margins to abdominoperineal amputation.[4,5] Shutze and Gleysteen[7] established a classification by stages and recommended some options for treatment.

The present case corresponds to stage I of the classification: Paget cells in the perianal epidermis and adnexae without primary carcinoma; the recommendation for this stage is local excision with wide margins. Finally, follow-up is critical and should focus on controlling the recurrence of the disease and the development of an associated carcinoma, performing biopsies of the margin of the old lesion, digital rectal examination, physical examination of the inguinal region, colonoscopy and imaging studies of the chest and abdomen.[4]

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COMMENT

Perianal Paget´s disease corresponds to 20% of the cases of extramammary Paget´s disease, however it is the location with the highest incidence of malignancy.

The contribution made by this article to the local bibliography is an illustration of the typical symptoms of these skin lesions, which are usually treated for long periods of time with corticosteroids or topical antifungals, without good results. Simonds et al.[1] report a delay in diagnosis of between 2 and 10 years.

Although it is a skin lesion, in up to 30% of cases there is an underlying neoplasm. According to a recent publication by Hutchings et al., from the Johns Hopkins Hospital Department of Pathology,[2] secondary extramammary Paget's disease can present in association with an invasive and/or in situ colorectal lesion. Although the latter is an uncommon presentation of a recognized rare disease, knowledge of this phenomenon is important to prevent overdiagnosis of invasion and possible overtreatment. To rule out an associated anal or rectocolonic neoplasm, a prior colonic study should always be performed, as outlined in this article.

As it is a recurrent pathology, a 2019 review proposes the Mohs microsurgical technique in order to perform an oncologic procedure that allows tissue to be preserved in the event of multiple interventions. Wide surgical resection has been the gold standard of treatment for years, however recurrence rates of 30-60% and recent studies have shown that the Mohs technique would allow 100% control of the margins, becoming the technique of choice in multiple skin neoplasms. However, as it is such a rare disease, more studies are required to support this technique for its treatment.

There is no consensus on the margins, described as 1 to 5 cm. Hendi et al.,[3] in 2004 presented a casuistry of 27 patients treated with the Mohs technique, describing that excising 5 cm of apparently healthy skin would achieve negative margins in 97% of cases, while excising only 2 cm would achieve a clear margin in 59%, recommending a surgical margin of 5 cm in case of not performing the microsurgical technique described.

In those cases not amenable to surgery, there are publications that evaluate non-operative treatment with immunomodulators such as imiquimod,[4] photodynamic therapy and even radiotherapy. Despite the chosen treatment method, long-term follow-up, both local and of the underlying disease, must be strict.

As a last comment, quoting Goethe: "You only find what you are looking for, you only look for what you know", hence the relevance of this clinical case.

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