

Surgical Treatment of a Perianal Paget's Disease: A Case Report and Short Review of the Literature

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Abstract

Extra-mammary Paget's disease (EMPD) is a rare and slow progressive skin lesion located in the perineum, perianal region, groin, scrotum or vulva. Perianal Paget may be associated with synchronous or metachronous visceral malignancies which makes necessary a complete study of the patient. Clinical presentation varies and there has been proposed many types of treatments. This article presents a 71-year-old man with no medical history of relevance referred to our department with a perianal lesion that was diagnosed afterwards as a Perianal Paget Disease. Surgical excision was performed. The patient underwent uneventful recovery and the long term follow up is asymptomatic so far.

Keywords: Paget's disease; Perianal; Surgical Treatment

Introduction

Paget's disease is an intraepithelial adenocarcinoma of the mammary glands. Its extra-mammary location was described afterwards. Extra-mammary Paget's disease (EMPD) is a rare and slow progressive skin lesion arising from the apocrine glands located in the perineum, perianal region, groin, scrotum and vulva. Its incidence and prevalence are not well known.

The main histologic feature are the Paget cells; large, basophil, vacuolated, PAS+ cells. As noted in previous publications Perianal Paget's disease can be associated with synchronous or metachronous visceral malignancies [1]. This makes necessary to complete the study of the patient with a colonoscopy and imagery, such as CT scan [2]. And also imposes a long-term follow-up. Symptoms are nonspecific sometimes leading to a delay diagnosis [3]. Clinical presentation includes exfoliated, exudative, and warty or hypopigmented patches associated with perianal pruritus or pain. Dermatologists generally diagnose it after the lack of response to both local and systemic treatments.

There are several non-surgical approaches described, such as radiotherapy, local application of imiquimod or interferon [2,4]. However, surgical treatment with negative margins is preferred when possible. To ensure free margins it is necessary to perform biopsies beyond the macroscopically affected area. The aim of this article was to review the literature and report our short experience with this rare disease.

Case Report

A 71 year-old-man with irrelevant medical history was referred to us by the Department of Dermatology due to perianal pruritus. He was under local and systemic corticosteroid therapy with no response. We scheduled a perianal biopsy and the results showed findings compatible with Paget disease (Figures 1A and 1B).

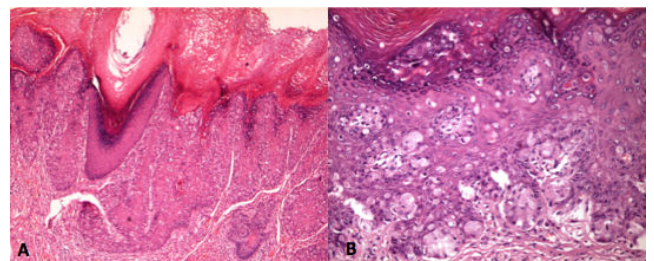


Figure 1: Results of Perianal biopsy.

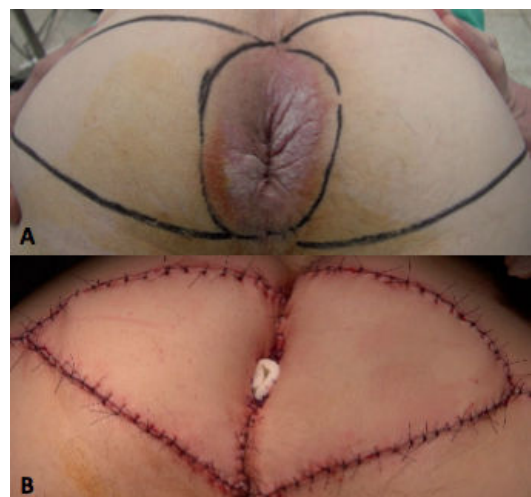


Figure 2: (A) Perianal region showed a painless, hypopigmented and exfoliative lesion (B) Figure showing V-Y advancement flap reconstruction.

The patient was in good health. Physical exam of the perianal region showed a painless, hypopigmented and exfoliative lesion (Figure 2A). Full colonoscopy was normal.

A surgical excision with 1-cm margin was performed on June 30 2015. Intraoperative frozen analysis of margins was done in order to ensure free margins. The defect was covered with a bilateral V-Y advancement flap reconstruction (Figure 2B). Immediate postoperative course was uneventful. Hospital discharge was on postoperative day 5.

Pathological analysis confirmed the initial diagnosis. Microscopic examination showed a warty pattern with clusters of Paget cells located above the basement membrane associated with acantosis, papillomatosis and hyperkeratosis. Negative margins were informed.

After 60 days he suffered a minor complication consisting in mild anal stricture that was treated with anal dilations. Now his long term follow up is asymptomatic (Figure 3).



Figure 3: After 60 days of follow up.

Discussion

Paget disease is observed in less than 5% of cases of breast cancer. Extramammary Paget's disease (EMPD) is a skin tumor arising in apocrine glands and represents nearly 6% of skin cancers. This adenocarcinoma is a rare condition and its etiology is not completely elucidated. Perianal location is infrequently reported.

Different types of EMPD have been described. Primary EMPD is originated in the Paget's cells of the apocrine glands of the skin and secondary EMPD is originated from epidermotropic spread of malignant cells from an underlying carcinoma (pagetoid phenomenon). The underlying mechanism of this phenomenon remains unclear and it is not frequently reported [5]. Gastrointestinal tract, adnexal organs and genitourinary area are the most frequent origins.

Perianal Paget's disease represents nearly 20% of all EMPD and 6% of all cases of Paget's disease [5,6]. Clinically, it shows a slowly expanding erythematous plaque frequently associated with pruritus and pain with occasional bleeding from ulcerated areas [2]. Minicozzi et al. [5] found that 58% of cases were not associated with other neoplasia while 10% were associated with another synchronic or metachronic neoplasia, 1% was associated with adnexal neoplasia, and in nearly 30% of cases an intradermal spread of rectal carcinoma was found. For this reason, clinical evaluation should include sigmoidoscopy, cystoscopy, colposcopy and mammography [2].

EMPD is a disease with high rates of local recurrence even after surgical resection with negative margins. The nodal spread is a

possibility and ilioinguinal or retroperitoneal nodes are the most frequently affected. Distant metastases can compromise liver, lung, bones and adrenal glands [5].

The most important characteristic of the pathologic exam is the presence of Paget cells –enlarged cells with clear PAS + cytoplasm and large nuclei- located in the epidermis as solitary cells or in clusters.

The gold standard of treatment is complete surgical resection with negative margins. Previously, a satisfactory biopsy should be performed. Surgical resection can be easily performed in the majority of extramammary location but in genital or perianal regions it may be difficult. The treatment of EMPD in perianal location has particular features. Frequently, the perianal skin is compromised in a circular way and so the reconstruction after surgical resection may be complex and skin grafts or local flaps are usually used. Some authors suggest that temporary colostomy may be an option to avoid wound complications. Local recurrence may be as high as 60% and is frequently due to incomplete resection. For this reason, resection needs to be performed with wide margins, usually 1-2 cm of healthy skin beyond the clinical tumor limits although some authors suggests a more aggressive resection (Ali Hendi). The use of Mohs micrographic surgery was postulated as an attempt to reduce tumor recurrence. This technique may be used as described by Mohs with intraoperative frozen evaluation or using the called "slow Mohs" that represents a staged resection. The reported recurrence rates with this technique seem to be lower (8% to 26%). As commented previously, the post-resection defect in the perianal region usually needs a complex reconstruction. Wide specters of reconstructive options have been described. If a skin graft is not an option because of the size of the perianal defect the use of flaps should be taken into account. The most popular flap used for reconstruction of large perianal defects is the V-Y advancement posterior thigh fasciocutaneous flap.

Lymph node dissection is not routinely recommended. In cases of nodal involvement, therapeutic lymphadenectomy may be performed. The use of radiotherapy is recommended in case of unrespectable disease or as adjuvant treatment. Chemotherapy may be used in inoperable patients, in cases of metastatic disease or before surgery with the objective of tumor reduction.

Finally, several alternative treatments in perianal Paget's disease have been described. The most reported are the use of photodynamic therapy, imiquimod 5% cream for topic use and even radiotherapy [2,4,7].

Conclusion

In conclusion, perianal Paget's disease represents a rare entity with specific associated conditions that should be investigated. Surgical resection remains the treatment of choice. We consider of vital importance to report these cases in order to contribute to the knowledge of this disease.

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