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The Spanish Journal of Gastroenterology

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DOI: 10.17235/reed.2022.9304/2022
Link: PubMed (Epub ahead of print)

Please cite this article as:
Navajas Hernández Pilar, Valdés Delgado Teresa, Machuca Aguado Jesús , González-Cámpora Ricardo, Argüelles Arias Federico. Perianal Paget's disease. Rev Esp Enferm Dig 2022. doi: 10.17235/reed.2022.9304/2022.

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Perianal Paget's disease

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Keywords: Perianal disease. Perianal Paget disease. Ulcerative colitis.

Dear Editor,

We present the case of an 85-year-old man diagnosed 20 years ago with ulcerative colitis (UC) that received maintenance treatment with oral mesalazine 3 g/24 h and a correct control of his disease. The last colonoscopy was performed in 2017, and the biopsies were compatible with minimal inflammatory changes, ruling out dysplastic changes.

In 2019 he lost to follow-up due to the COVID-19 pandemic. Two years later he consulted for anal itching and pain, which he initially attributed to his UC and sometimes improved with topical ointments.

Physical examination revealed a 10cm hard and erythematous plaque with raised edges in the perianal area (image 1a). After biopsy, a diagnosis of primary Paget´s disease (PD) was made (image 1b, 1c). Colonoscopy and thoraco-abdominal CT scan were performed and found to be normal. Due to the invasive nature of the lesion and the age of the patient, treatment with radiotherapy was decided. Nevertheless, the

patient died after 9 months due to dissemination of the disease.

Discussion

Perianal Paget's Disease (PPD), a subset of extramammary PD, is a rare cutaneous neoplasm, with less than 200 published cases (1), which is often misdiagnosed and undertreated. Its incidence accounts for less than 1% of perianal disease and 1.3% of cases of PD. It occurs most commonly in women aged 60-70 years with the formation of erythematous, hard, well-demarcated, pruritic and sometimes painful erythematous plaques or macules.

PPD is classified as primary PPD, if the tumor arises from the intraepidermal cells themselves, and secondary PPD, when the primitive cancer is usually occult in adjacent organs, mainly digestive or genitourinary (2). Immunohistochemical findings allow us to differentiate between these two types. Typically, markers such as CK7 and 34ßE12 are positive in the primary PPD and negative for HMB45, CK20 and CDX2 (3).

Primary PPD has a better prognosis when it is intraepithelial, but can progress to invasive disease with metastases. When the disease is localised, surgical resection is the choice; in metastatic disease, radiotherapy and/or chemotherapy will be chosen (4). Whether the stage is early or advanced, recurrence is frequent and therefore long-term follow-up is essential.

There are no cases described in the literature that establish a relationship between UC and PPD. We do not know therefore, whether the presence of inflammatory bowel disease or its activity could be related to a carcinogenic stimulus (5) that pathophysiologically triggers PPD. More cases and studies would be needed to corroborate this hypothesis.

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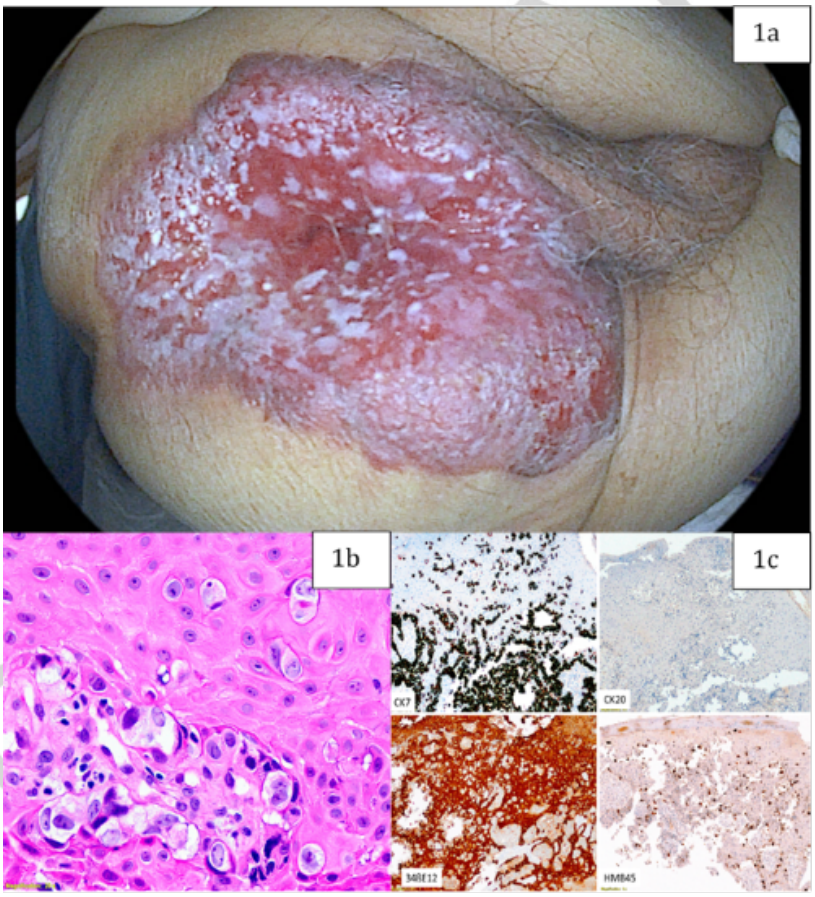

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1a. *Large, hard and erythematous plaque in perianal area*. **1b.** *Epidermal infiltration by cells with large and clear cytoplasm with large irregular nuclei and prominent nucleoli forming cluster, Paget cells.* **1c**. *Positivity for CK7 and 34βE12 markers and negativity for CK20 and HMB45 markers. These findings are compatible with primary Paget's disease.*