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SCIENTIFIC LETTER

Perianal Paget's disease: A rare disorder masking an underlying anorectal carcinoma☆

Enfermedad de Paget perianal: un trastorno raro que enmascara un carcinoma anorrectal subyacente

Perianal Paget's disease (PPD) is a rare type of cancer, with less than 200 cases reported in the literature. PPD generally affects Caucasian people, most often female, with a peak incidence between the ages of 50 and 80.[1]

PPD can present as a primary skin disorder or be associated with different synchronous or metachronous cancers, becoming a complex disorder due to the different pathological processes. Primary PPD has relatively favourable prognosis, with both overall and disease-free survival being approximately 60% at five years.[2]

In the secondary form, however, with synchronous or metachronous carcinoma of the anal region, based on limited reports, the prognosis is poor.[3]

We present the case report of a 61-year-old man who was referred to a proctology clinic for lesions in the perianal area associated with local discomfort and pruritus. A first biopsy showed extramammary Paget's disease. The patient was started on conservative treatment based on imiquimod cream and local corticosteroid therapy. Three years later, physical examination detected significant growth of the perianal lesions, with a fungating appearance and spread to the anal canal (Fig. 1). The protocol to rule out association with an underlying visceral cancer was applied to exclude rectal carcinoma with spread to the anus. A complete colonoscopy revealed, inside the anal canal and in the surrounding skin, an ulcerated fungating cancer reaching as far as the rectal mucosa, but without invading the rectal ampulla. The report from the new histological examination described an adenocarcinoma with mucinous differentiation and with the same immunohistochemical pattern as the first biopsy. Imaging tests of chest and abdomen for spread of the cancer revealed pathological involvement of the inguinal lymph nodes and iliac chains bilaterally. The first-line treatment of choice consisted of the surgical approach, involving abdominoperineal resection with end colostomy on the left flank. The definitive histological examination of the surgical specimen was positive for a moderately differentiated mucosecretory adenocarcinoma (TNM-pT3, AJCC 8th edition). To control the residual disease, it was decided to apply locoregional adjuvant radiotherapy to the iliac and inguinal lymphadenopathy. Three months after the intervention, a total body CT was performed which showed no evidence of distant visceral disease.

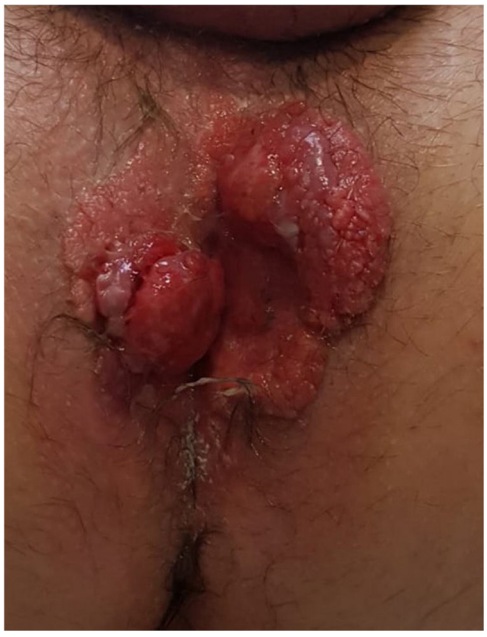

Figure 1 Hard, painful, fungating tumour in the perianal region on dystrophic skin, approximately 10 cm long, with small superficial erosions.

Extramammary Paget's disease is a clinicopathological entity with a complex diagnostic classification. The location in the perianal region is important from a prognostic point of view, because of the possibility, in up to 50% of cases, of finding an anorectal carcinoma, the most important prognostic factor for which is the depth of invasion. Tumours with a depth of >1 mm are associated with a worse prognosis when they spread outside the skin to the lymph nodes or other surrounding tissues.

PPD initially presents as an erythematous plaque with scaly and eczematous areas, causing local itching and/or burning. It is important to stress that, when local therapy

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(corticosteroids or antifungals) is not effective after a certain amount of time, PPD should be suspected and a biopsy of the lesion performed. In late stages it can progress towards the formation of a bleeding, fungating mass, as a harbinger of an underlying cancer.

From a therapeutic point of view, there is no unified treatment protocol available at present, so the choice of management in each case will depend on the type (primary vs. secondary) and extent of the lesion, and on clinical experience.

In cases of secondary PPD, therapeutic success will depend on the oncological approach to the associated carcinoma, combining surgery, radiotherapy and/or chemotherapy. Although as yet there have been no randomised clinical trials comparing it to surgery, radiotherapy can be considered in selected cases as an effective treatment modality and a suitable alternative to extensive surgical excision.[4]

In conclusion, the significance of PPD means that it is essential not only to make the differential diagnosis with other skin cancers, but also to ensure early management of a condition which may be the first metastatic manifestation of an underlying visceral carcinoma.

References

1. Carbotta G, Sallustio P, Prestera A, Laforgia R, Lobascio P, Palasciano N. Perineal Paget's disease: a rare disorder and review of literatura. Ann Med Surg (Lond). 2016;9:50–2.
2. Marchesa P, Fazio VW, Oliart S, Goldblum JR, Lavery IC, Milsom JW. Long-term outcome of patients with Perianal Paget's disease. Ann Surg Oncol. 1997;4:475–80.
3. William SL, Rogers LW, Quan SH. Perianal Paget's disease: report of seven cases. Dis Colon Rectum. 1976;19:30–40.
4. Liu CF, Wang Q, Kong YY, Tu X, Wang J, Zhu X. A clinicopathological study of perianal Paget's disease associated with internal rectal adenocarcinoma. Zhonghua Bing Li Xue Za Zhi. 2004;33:11–5.

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Metastasis of lobular breast carcinoma in the bowel

Metástasis de carcinoma lobular de mama en el intestino

Breast cancer is the most common malignancy in women, but only 6% of newly diagnosed patients have distant metastasis at diagnosis.[1] Metastatic spreading disease to the gastrointestinal tract is very rare.[2] Here we report a case of bowel metastasis as the initial presentation of lobular breast carcinoma.

A 67-year-old female presented with six months of diffuse abdominal pain, diarrhea and weight loss. She had a past history of dyslipidemia, currently medicated with simvastatin. Abdominal examination was unremarkable. Laboratory investigation and upper endoscopy were normal. Colonoscopy revealed eodema and loss of vascular pattern in the ascending colon and cecum; the ileocecal valve presented marked erythema and friability, preventing ileoscopy (Fig. 1A). Pathologic evaluation of biopsies from the ileocecal valve showed poorly cohesive tumor cells in lamina propria and some signet ring cells (Fig. 1B); immunohistochemistry was positive for estrogen receptor (Fig. 1C), cytokeratin 7 and AE1/AE3. These findings were consistent with metastatic lobular breast carcinoma to the colon.

The patient was sent to Ginecology clinic for examination and, in the upper outer quadrant of the left breast, it was palpable a hard, immovable, painless mass with irregular borders, associated with ipsilateral retracted nipple and axillary adenopathy. Breast ultrasound demonstrated a hypoechoic nodule with posterior acoustic shadowing, measuring 29 mm × 17 mm (Fig. 1D). Histology of nodule biopsy confirmed invasive lobular carcinoma (estrogen receptor positive, progesterone receptor negative, Her2 negative, Ki-67 10%). Further staging imaging also revealed axillary lymph nodes and bone metastasis. The patient was proposed for palliative hormonal therapy.

Metastatic breast cancer is uncommon at the time of diagnosis,[1] usually spreading to the bone, lung, liver and brain.[3] Gastrointestinal metastasis from breast cancer are rare and usually associated with lobular histology.[2] The most frequent site of metastasis on the gastrointestinal tract is the stomach, followed by the small bowel and the colon.[2] Clinical manifestations are non-specific and include abdominal pain, bowel obstruction or inflammatory diarrhea.[2] Endoscopic features include mucosal nodularity and rigidity, or lobulation and deformity caused by the presence of a mass. Therefore, diagnosis is challenging as metastasis may mimick inflammatory bowel disease or primary colon cancer.[2] In conclusion, we report a rare case of bowel metastasis as the initial presentation of lobular breast carcinoma, that requires a high index of suspiction for the diagnosis from the pathologist and the gastroenterologist.