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Letters to the Editor

Paget disease presenting as a perianal papilla: Report of a unique case

Key words: Paget disease. Perianal papilla. Mucosectomy.

Dear Editor,

A 54-year-old man with no relevant personal history, was referred to our department because of anal bleeding. The abdomen was normal and there were neither inguinal lymphadenopathy nor lesions in the perianal region. Digital rectal examination revealed a large polypoid mass.

Laboratory data were normal, including complete blood count, liver function and carcinoembryonic antigen. Colonoscopy showed, close to the dentate line, a sessile polyp of 4 cm in size (Fig. 1A). Biopsy specimens revealed a villous adenoma with high-grade dysplasia.

Ultrasound endoscopy and pelvic MRI showed a mucosal lesion. CT scan excluded abdominal metastasis.

The patient underwent endoscopic resection of the lesion in retroflexion (Fig. 1B). Histological examination revealed a well differentiated adenocarcinoma, without invasion of the submucosa.

One month later, an endoscopic tattooing of the mucosectomy scar was done, because the surgeons wanted to know exactly the location of the scar, in order to perform the most conservative transanal resection. During this procedure, a perianal papilla was observed (Fig. 1C).

Transanal resection of the tumor and removal of the anal papilla were performed. Histological examination excluded residual adenocarcinoma in the rectal wall flap, but, in the papilla, there were signet ring cells with a "pagetoid" spread to the glandular structures (Fig. 1D) –diagnostic with perianal Paget disease. The immunohistochemical stains were positive for cytokeratin 20 (CK20+) and negative for cytokeratin 7 (CK7-).

He was submitted to a widening of the anal margin of the lesion. Biopsy specimens of the frozen sections excluded intraepithelial neoplastic structures.

Discussion

Perianal Paget disease (PPD) is a rare carcinoma, characterized by the presence of atypical Paget´s cells in the anal or perianal epidermis. In 33 % of the cases, the patients have an associated anorectal carcinoma, however, less than 30 cases have been reported in the literature (1).

The pathogenesis is not well defined, but PPD can be classified as primary (majority of the cases), arising as a primary adenocarcinoma in the epidermis, or secondary, resulting from epidermotropic spread or metastasis of an underlying cancer, mainly gastrointestinal (2). The capacity of the tumor cells to move from their place of onset to skin is known as pagetoid diffusion, as observed in our patient.

Immunohistochemical stains for CK7 and CK20 are useful in differentiating the two types of PPD: primary PPD has an immunophenotype CK20+/CK7 –while the secondary is CK7+/CK20– (3). The CK20 is a marker of anorectal carcinoma with pagetoid diffusion. In our patient, the histological examination and the positivity of the signet ring cells for CK20 were important to confirm the origin of PPD in a synchronous anorectal carcinoma with pagetoid diffusion.

Usually, the PPD presents as an erythematous lesion and patients describe eczema-like symptoms. In contrast, our patient was asymptomatic. The only perianal lesion present was a papilla, observed after the endoscopic resection of the rectal carcinoma. The most reasonable explanation is that the PPD was already presented, but it manifested weeks later as an anal papilla that became more prominent with some degree of inflammation caused by the mucosectomy performed in that area.

Wide local excision is recommended in PPD when invasiveness is absent. For invasive cancer, the abdominoperineal resection is the treatment of choice (4).

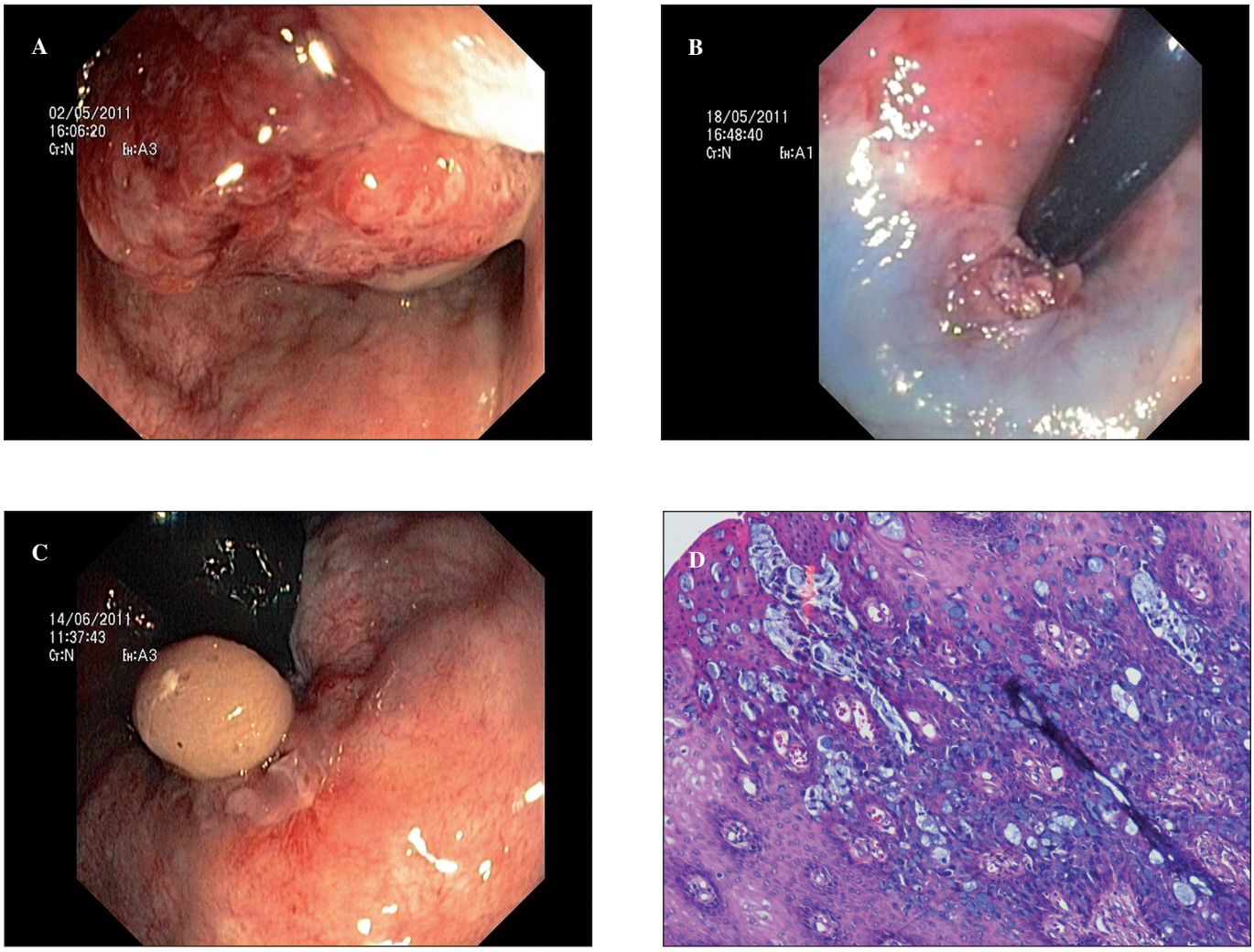

Fig. 1. A. Sessile polyp close to the dentate line. B. Endoscopic resection of sessile polyp, seen in retroflexion (arrow). C. Anal papilla (arrow). D. Histological examination of anal papilla showing pagetoid diffusion (arrow).

Although the optimal follow-up remains controversial, the long term follow-up is needed because it has an increased tendency for local recurrence.

This case emphasizes that, even in the absence of clinical signs of PPD, pathologic examination of the perianal area is import in patients with anorectal carcinoma. Besides, we should value all the anal findings because papilla can be a form of presentation of PPD. To the best of our knowledge, this is the first reported case of PPD manifested as a perianal papilla, in a patient with synchronous rectal cancer.

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